A CLINICAL STUDY OF CONGENITAL MELANOCYTIC NAEVUS

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CERTIFICATE

Certified that this dissertation entitled **"A CLINICAL STUDY OF CONGENITAL MELANOCYTIC NAEVUS"** is a bonafide work done by **DR.K.Uma Maheswari ,** Post Graduate Student of the department of Dermatology, Venereology and Leprosy, Madras Medical College, Chennai – 600 003, during the academic year 2007 – 2010. This work has not previously formed the basis for the award of any degree.

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INTRODUCTION

Naevus is a Latin word which means maternal impression or birth mark. Naevus is a common skin lesion seen in patients of all ages and both sexes.It is usually present at birth or in early childhood. However, some may develop later in adulthood.

Hamartoma is a Greek word which means to err. Hamartoma is a tumour like, non neoplastic proliferation of abnormal mixtures of the normal components of a tissue. Previously naevus and hamartoma were considered to be synonymous but now they are regarded as distinct entities as there is no neoplastic proliferation in hamartoma whereas in naevus neoplastic proliferation occurs.

Naevi are of cosmetic significance if they are large and located on visible areas of the body.Their importance also lies in associated defects of organ systems and possible neoplastic potential, benign and malignant.

There are several types of naevi .Based on the tissue of origin, they are classified as epidermal naevi, melanocytic naevi, dermal and subcutaneous naevi.

Melanocytic naevi are benign neoplasms or hamartomas composed of nevomelanocytes. They are broadly classified based on their being derived from epidermal melalanocytes or from dermal melanocytes.

Congenital and acquired melanocytic naevi are derived from epidermal melanocytes while naevus of Ota, naevus of Ito and blue naevi are derived from dermal melanocytes.

Congenital melanocytic naevi are anomalies in embryogenesis. They could be considered as malformations or hamartomas, made up of nevomelanocytes which lack normal melanocytic differentiation and occur as ‘nests’ in the epidermis and /or dermis.

In this study the age and sex distribution of congenital melanocytic naevi, types of congenital melanocytic naevi, localization , their cutaneous or systemic associations if any, complications, skin biopsy, special staining and treatment of congenital melanocytic naevi were studied.

REVIEW OF LITERATURE

CONGENITAL MELANOCYTIC NAEVUS

Synonyms of congenital melanocytic naevus are

- Congenital Nevomelanocytic Naevus
- Garment Naevus
- Naevus Pigmentosus Et Pilosus
- Giant Naevus
- Verrucous Naevus
- Giant pigmented Naevus
- Bathing Trunk Naevus

Historical aspects:

1832 - Alibert described, a giant waist coat and drawer's naevus

1861 - Rokitansky described, a patient with a giant congenital melanocytic naevus and leptomeningeal hyperpigmentation.

1878 - Baker reported staged excisions of very large congenital melanocytic naevus

1879 - Jablokoff and Klein described, malignant potential of giant congenital melanocytic naevus [1]

1915 - Morestin
1925 - Roy
} Staged excisions of large congenital melanocytic naevus

1965 - Reed et al described histopathologic appearance of large congenital melanocytic naevus [2]

Incidence And Epidemiology: [3]

I in 100 new borns has one congenital Naevus[4]

I in 10 of these new borns has multiple lesions

I in 20,000 new borns has large congenital melanocytic naevus greater than 10 cms[5]

I in 500,000 new borns has Garment type naevus[5]

No gender predilection is seen in congenital melanocytic naevus.

Familial aggregation has been demonstrated for both large and small varieties of congenital melanocytic naevus.

Discordance for giant congenital melanocytic naevus has also been demonstrated in identical and non-identical twins.

Tardive congenital melanocytic Naevi:

Synonyms : Congenital – type Naevi or Early – onset Naevi

They are not present at birth. Appear in the first 2 yrs of life .Show clinical and pathological features of congenital melanocytic naevus rather than acquired naevus.

Aetiology and Pathogenesis

The aetiology of congenital melanocytic naevi remains unclear. The melanocytes of the skin originate in the neuroectoderm, although the specific cell type from which they derive remains unknown.

One hypothesis is that pluripotent nerve sheath precursor cells migrate from the neural crest to the skin along paraspinal ganglia and peripheral nerve sheaths and differentiate into melanocytes upon reaching the skin.[7]

One possible explanation for the presence of congenital melanocytic naevi is that an external insult results in a mutation that affects the morphogenesis of the embryonic neuroectoderm and migration of precursor cells to the skin.

Genetic Factors:

Congenital melanocytic naevus represent a developmental abnormality of normal

melanocyte development. This is presumably due to mutation of NRAS[6] that occurs in a progenitor cell that results in the abnormal extensive accumulation of melanocytic cells along migration pathways during normal development.

The high frequency of NRAS mutations in naevi that developed in utero demonstrates that UV light exposure is not required for NRAS mutations. In contrast, the complete absence of BRAF mutations in naevi present at birth, but their frequent occurrence in acquired naevi, parallels the finding in melanoma in which BRAF mutations are found in melanomas that occur on intermittently sun-exposed body sites but are rare or absent in melanomas developing in completely sun-protected mucosa-lined body cavities.[8,9,10] These striking patterns suggest a possible link between UV exposure and BRAF mutations in melanocytic neoplasia. However, any connection is likely to be complex as the mutations also occur in other cancers and do not show typical signatures of UV induction. [11]

Congenital melanocytic naevi develop independently of UV light exposure. So there is no BRAF mutations. In 81% of cases mutations in NRAS are found.[12]

Congenital pattern naevi showed inverse mutation pattern. BRAF mutations are found in 71% of cases where as NRAS mutations in 25% of cases. Thus naevi that develop in utero are genetically distinct from those that develop later.

Large congenital melanocytic naevi have so far always been considered to occur sporadically, and until now little has been written about a possible role of heredity as a cause of this disorder. With the occurance of familial cases of large congenital

melanocytic naevi, the concept of paradominant inheritance[13] was suggested as a possible genetic explanation.

The concept would imply that heterozygous individuals are phenotypically normal which is why the mutation would be transmitted unperceived through many generations. The trait would become manifest only when loss of heterozygosity occurred at an early developmental stage, giving rise to a patchy area of homozygous or hemizygous cells.

This would explain why the lesions of large congenital melanocytic naevi are always arranged in a mosaic pattern; why they occur virtually always sporadically; and why the exceptional cases of a familial aggregation of this trait do not show any consistent Mendelian pattern.

Time Of Development Of Congenital Melanocytic Naevi In Utero:

Clinical findings such as a congenital divided naevus of the eyelid [kissing naevus] can give us insight into when these events occur. The eyelids form at between 5 and 6 weeks in utero, begin to fuse at about 8 to 9 weeks, and reopen during the sixth uterine month.[14],[20]

Melanocytes appear in fetal skin around 50 days of gestational age. So congenital melanocytic naevus forms after 50 days but before the 6th month of intrauterine life, when the eyelids split.

Melanocytes present in the basal layer of the epidermis exhibit a certain degree of territoriality. Non neoplastic melanocytes typically exhibit contact inhibition to each other.So pigment cells are never found as contiguous cells.

Melanocytic naevi represent proliferations of melanocytes that are in contact with each other, forming small collections of cells known as nests. Congenital melanocytic naevi are lesion present since birth containing naevus cells.

In giant congenital melanocytic naevi, naevus cells may be found in regional lymph nodes.[15] They are found usually in capsules of lymph nodes, whereas malignant melanoma metastases are subcapsular. Rarely intraparenchymal aggregates of naevus cells may be present which causes diagnostic pitfalls with malignant melanoma and carcinoma.[16]

Immunostaining for the tumour suppressor gene p16 product is a useful marker to

differentiate melanoma metastasis from lymphnode naevus cells.[17]

Clinical Manifestations:

Morphological Characteristics:

Number: Small congenital melanocytic naevi present as single or multiple lesions. Giant congenital melanocytic naevi are usually present as single very large or multiple small lesions .

Shape : It may be round or oval in shape

Border : It may be regular,or irregular and usually sharply demarcated.

Surface : It may be smooth, pebbly, rugose, verrucous, cerebriform or grossly lobular surface . Skin markings distort the skin surface atleast slightly when assessed by oblique lighting.

Cutis Verticis Gyrata : Congenital melanocytic naevi of the scalp may present as cutis verticis gyrata. The term cutis verticis gyrata describes a morphological syndrome in which there is hypertrophy and folding of the skin of the scalp to present a gyrate or cerebriform appearance.Congenital melanocytic naevi appear to be the most common cause of secondary cutis verticis gyrata.

Pigmentation : May be uniform, or has medium or dark brown speckles, symmetrically disposed in a tan or light-brown field or a reticular pattern.

Presence of hair : The events leading to the nevomelanocyte accumulation may also

have effects on the surrounding tissue possibly due to changes in the local cytokine environment of the nevomelanocytes. Coarse, long, darkly pigmented hair may be present at birth, appear within the first year of life or 2 of life or be delayed for several years. Hairs are often concentrated in the centre.

Features suggestive of Atypical appearance:

Poor demarcation and / or irregular outline.

Haphazard pigmentation, very dark brown, black, or blue black or

discontinuous pigmentation.

Associated atypical histological features.

In Whites very darkly pigmented congenital melanocytic naevi may have atypical histological features .

As a child grows, the congenital melanocytic naevi should grow relatively proportionally and continue to mature.[18] History of disproportionate growth, especially after 6 months, or change in a non- uniform manner is of concern for possible melanoma.

Classification of congenital melanocytic naevi is based on:

1. Absolute Size

2. Body surface area

3. Prediction classification

4. Ease of Removal

Based on absolute size:[19]

Small : < 1.5 cm

Medium : 1.5- 10 cm

Large : 10 – 20 cm

Giant : > 20 cm

Classification on the basis of body surface area of the lesion:

Giant congenital melanocytic naevi :

Lesion as large as the patient's palm for face / neck, twice that area for other anatomic sites or involvement of 30% of the body surface area or 900 cm^2 in adults.

Prediction classification:

Giant naevi have also been described as comprising 9 cm on a child's head and 6 cm on a child's body.

Congenital melanocytic naevus may grow in proportion to the affected anatomic site. Small or medium congenital melanocytic naevus in the newborn period may become medium or large by late childhood or adulthood respectively.

Based on ease of removal:

Classified as small congenital melanocytic naevus if excised easily, and wound defect closed primarily without significant deformity , without using skin flaps or grafts.

ASSOCIATIONS OF CONGENITAL MELANOCYTIC NAEVUS:

Neurofibromatosis type 1

Lipomas

Vascular Naevi – Haemangiomas / Vascular malformation

Neurocutaneous Melanosis

Other tumors:

Schwannoma / Neuroid tumour

Rhabdomyosarcoma / Fibrosarcoma/

Lymphangioma/ Mastocytoma/ Sebaceous naevus

Skeletal abnormalities:

Spinal Dysraphism

Club Foot

Pes cavus

Hammer toes or claw toes

Congenital melanocytic naevus with Neurofibromatosis type I:

There is a significant association between giant congenital melanocytic naevus

and neurofibromatosis. This could be due to their common origin from neuroectoderm[21].Sometimes nodules may arise in congenital melanocytic naevus that have similar histopathological features of neurofibroma or schwannoma[22].

S-100 protein immunohistochemical staining helps to differentiate neurofibroma and neurofibroma like nodules of congenital melanocytic naevus. Lisch nodules in neurofibroma may even be due to nodular naevi of the irides. NF-1 compared with sporadic NF has been associated with congenital melanocytic naevus . Giant congenital melanocytic naevus with neurofibroma –like changes and spina bifida occulta has been reported.[23]

Congenital melanocytic naevus with lipoma:

Giant congenital melanocytic naevus associated with lipoma and neurofibroma has been reported.[25] A study was done on agouti mouse, investigating a possible role of the central nervous melanocortin system in the control of adiposity through effects on nutrient partitioning and cellular lipid metabolism independent of nutrient intake.

Central nervous melanocortin system, the most potent brain circuit known to control food intake, also regulates peripheral lipid metabolism.The melanocortin neurons and melanocortin receptors in the central nervous system directly and potentially affect cellular glucose utilization, lipid intake and triglyceride synthesis in the periphery. These reasons may be responsible for the occurance of lipoma in a giant naevus.

Neurocutaneous Melanosis (NCM) :

During normal embryogenesis, melanoblasts migrate from the neural crest to the leptomeninges and skin.[33] Dysregulation within this migratory pathway can result in increased proliferation of melanocytes and abnormal deposition of melanocytes and melanin in the leptomeninges, an entity known as neurocutaneous melanocytosis.

Patients with NCM may also have excess deposits of melanocytes in the skin, which can manifest clinically as the presence of multiple satellite melanocytic naevi, multiple medium-sized congenital melanocytic naevi, or large congenital melanocytic naevi. Patients with large congenital melanocytic naevi, a relatively rare condition affecting approximately 1 in 20 000 newborns, are at significantly high risk for developing NCM. It is reported that large congenital melanocytic naevi on the posterior axis (paraspinal, head, and neck regions) and in the presence of many satellite naevi may define a subgroup of patients at greatest risk for developing NCM.

Clinical Markers likely to develop NCM[27,28]

- Large congenital melanocytic naevus on the head or neck and on the posterior central axis.

- Satellitosis :

>3 small congenital melanocytic naevus but without a concomitant large congenital melanocytic naevus.

>20 satellite lesions .High Satellite numbers along with the presence of large

congenital melanocytic naevus.[29]

Neurocutaneous melanosis may be asymptomatic or symptomatic[26]

Asymptomatic NCM :

Asymptomatic neurocutaneous melanosis should be monitored clinically for progression to symptomatic neurocutaneous melanosis.

Symptomatic NCM :

1. Symptoms are headache, irritability, lethargy, recurrent vomiting, photophobia, paresthesias, meningismus, dysarthria, ataxia, loss of developmental mile stones including loss of previously achieved bladder and bowel function, seizures, disproportionate growth of head circumference, any focal neurologic signs.

2. It is common during the first 5 years of life.

3. It has poor prognosis.

4. As treatment,complete eradication of the intra cranial melanocytes is difficult and often it is impossible.

5. Palliative procedure like ventriculoperitoneal shunting can be done.

Spina bifida occulta : (occult spinal dysraphism)[23]

Developmental abnormalities of the skin, vertebral bodies and spinal nerves may occur without exposed neural tissue.

Cutaneous markers of occult spinal dysraphism:

- A midline mass such as lipoma

- A rudimentary tail protruding form the lower back

- Capillary haemangioma

- Midline deep dimples in the lumbo-sacral area

- Lumbo sacral dermal sinus

- Pigmented neavus

- An abnormally hairy patch

- Deviation of the gluteal crease

- Hamartoma

- Mongolion spot

- Atretic meningocele (a cigarette burn type of skin marking)

- Anorectal abnormalities

Giant congenital melanocytic naevus or those with other cutaneous

abnormalities described above, may warrant imaging.

Congenital melanocytic naevus with Vascular Naevi:

Very extensive congenital melanocytic naevus may be intermixed with elements of vascular malformation, haemangiomas,[24] increased numbers of mast cells, cartilage, calcification, and even bone.

Complications Of Congenital Melanocytic Naevi:

1. Ulceration

2. Pruritus: Increased number of mast cells may be present in giant congenital melanocytic naevus causing pruritus.

3. Pigmentary disturbances may occur.

4. Proliferative nodules can occur over the naevus.

5. Cutaneous malignant melanoma:

Congenital melanocytic naevus expands with growth of the child. The risk of melanoma development is proportional to the size of the congenital nevus[40,41,42]

The potential for large congenital nevi to become malignant is significant and is an important consideration in the treatment and management of this entity.

Multiple studies have attempted to elucidate the cumulative risk of developing cutaneous melanoma in patients born with congenital melanocytic naevus[43,44,45]

A recent study at the University of Pennsylvania Medical Center reported a 5.7% cumulative 5-year risk of developing a cutaneous melanoma in patients with large or giant congenital melanocytic naevus.

A study of the Dutch nationwide pathology database reported a Standardized incidence rate of 12.2% of developing melanoma in congenital melanocytic naevus. This study observed an increase risk of melanoma (in patients with congenital melanocytic naevus) of 6.4% for men and 14.1% for women when compared to general population rates.

Furthermore, patients with giant congenital melanocytic naevus had an increased risk of 51.6% compared to general population rates.

Rates of malignant potential for small and medium congenital melanocytic naevi are reported between 0.8% and 4.9%.Very large congenital naevi account for less than 0.1% of cutaneous melanomas, whereas small varieties of congenital nevi because of its increased incidence may account for 15% of cutaneous melanomas.

Reports of lifetime risk of developing a melanoma for patients with a large congenital melanocytic naevus range from 6.3-12.2%.

Malignancy should be suspected with focal growth, pain, bleeding, ulceration, significant pigmentary change, or pruritus.

Regarding giant nevi, 50% of the malignancies develop by 3 years of age, 60% by childhood, and 70% by puberty. Approximately 40% of the malignant melanomas

observed in children occur in large congenital naevi.

6. Leptomeningeal melanoma:[39]

Melanocytic proliferations affecting the central nervous system (CNS) of children may be classified as meningeal melanocytosis, primary melanoma, or metastatic melanoma. Meningeal melanocytosis often is associated with giant congenital pigmented nevi (preferentially involving the midline, the head and neck) representing the lethal condition neurocutaneous melanocytosis. Primary or metastatic melanomas, although extremely rare in children, can occur in the brain and its coverings and are associated with a poor prognosis.

7. Hypertrophic scarring and keloid:

Hypertrophic scarring and keloid can occur as a complication of surgical treatment procedures like excision and skin grafting.

8. Psychosocial morbidity:

In cases of giant naevi ,psychological stress can occur as a result of cosmetic disability.

VARIANT OF CONGENITAL MELANOCYTIC NAEVUS:

Desmoplastic Hairless Hypopigmented Naevus: [48]

Desmoplastic hairless hypopigmented naevus has characteristic clinical and histopathologi features.

Clinical features are

Hard indurate texture

Alopecia

Progressive loss of pigment

Irregular mottled border

Pruritus

HISTOPATHOLOGY:[49,50,51,26]

Although histopathologic features are cited as being useful in distinguishing naevi as congenital or acquired, there are no known features with absolute specificity and sensitivity.

Naevus Cell differ from ordinary melanocytes by being arranged in clusters or nests.It is a round cell rather than a dendritic cell.It retains pigment in their cytoplasm rather than transferring it to the neighbouring keratinocytes.

Types of Naevus Cell:

There are 3 types of naevus cells. They are Type A, B, C cells

Type A naevus cell: It is present in upper dermis, resembles epithelioid cells.It is round to cuboidal, has abundant cytoplasm containing varying amounts of melanin granules.

Type B naevus cell: It is smaller than type A cells, resembles lymphoid cells. It has less cytoplasam and less melanin.It usually occurs as well defined aggregates or cords of cells.

Type C naevus cell: It is present in lower dermis, resembles fibroblast or schwann cell.It occurs as elongated cells with spindle shaped nucleus. It rarely contains melanin.

Inverted Type A naevus cell: It is Type A cells present in deeper dermis

Non giant (small and Intermediate) congenital melanocytic naevi:

They may show features of congenital naevi or acquired naevi. They may have junctional, compound or intradermal architecture. Presence of melanocytes around and within hair follicles, in sweat ducts and glands, in sebaceous glands, in or in intimate association with vessel walls, in arrector pili muscles, in perineurium of nerves will be present in congenital naevi.

There will be extension of melanocytes between collagen bundles singly or in double rows and also extension into the deep reticular dermis and subcutis can occur in congenital naevi. Some congenital naevi are entirely junctional.In congenital naevi deep dermal involvement appear to increase with the size of the lesion.

Special forms of Non giant congenital melanocytic naevi :

I.Cerebriform congenital naevus:

It is of intradermal naevus pattern with neuroid changes simulating neurofibroma

II.Spotted grouped pigmented naevus:

It may be of intradermal naevus pattern.It may be eccrine centered or follicle centered.

Eccrine centered: Each eccrine sweat duct is tightly enveloped by nevus cells. Hair follicles are involved only slightly.

Follicle centered :Naevus cell nests mainly found around the hair follicles

III. Acral naevus:

It may be of compound naevus pattern. In the upper dermis pigmented nevus cells are present.In the lower dermis non pigmented naevus cells are present around blood vessels and eccrine glands.

IV. Desmoplastic hairless hypopigmented naevus:

Intense dermal fibrosis which is progressive in nature is present.They have scarce naevus cells and hypotrophic or absent hair follicles.

Features Simulating melanoma:

1. Asymmetry and poor circumscription of the naevi
2. An increased number of single melanocytes instead of nests of melanocytes in the naevus.

3. Single melanocytes that are not equidistant from one another.

4. Scattered single melanocytes present above the dermo-epidermal junction.

5. Confluence of nests of melanocytes.

Features against melanoma:

1. Absent cytologic atypia

2. Absent frequent mitoses

Giant congential melanocytic naevi:

There are 3 patterns : 1. Compound or intradermal naevus pattern

2. Neural naevus pattern

3. Blue naevus pattern

1. Compound or intradermal naevus pattern :

These are the most commonly seen patterns.In these patterns involvement of skin appendages and blood vessels by naevus cells occur.

2. Neural naevus pattern :

Melanocytic elements appear like neuroid tubes and naevic corpuscles.It may resemble neurofibroma histologically.

3. Blue naevus pattern :

It may have elements of blue naevus either common or cellular type or the entire lesion may consist of giant blue naevus pattern.

Leptomeningeal Melanocytosis:

In leptomeningeal melanocytosis there occurs diffuse infiltration of the leptomeninges with pigmented melanocytes.The blood vessels entering the brain and spinal cord may be surrounded by melanocytes.

There may be areas of infiltration of the brain or spinal cord with melanocytes. Leptomeningeal melanoma arising as a complication can infiltrate the leptomeninges and form multiple nodules in the brain.

Melanoma in giant congenital naevi:

Melanoma arising in giant congenital naevi has undifferentiated 'blastic' cells resembling lymphoblasts with little or no melanin or it may resemble large epithelioid cells similar to those of many melanomas.

Course and Progonsis:

Congenital melanocytic naevus do not remain static after their appearance at birth, have a dynamic evolution during body growth.

With few exceptions, congenital melanocytic naevus generally expand in direct proportion to growth of a given anatomic zone,although disproportionately rapid area expansion of some congenital naevi may occur during early infancy. Lesions in fully grown individuals should remain stable.

Elevation, Surface pigmentation may occur. Hairless at birth may develop long, dark, coarse hair or may maintain a relatively normal hair density.

Lightly pigmented may become more darkly pigmented, and darkly pigmented may become less pigmented. Spontaneous pigmentary regression with a desmoplastic reaction may occur.

Halo phenomenon:

Regression of congenital melanocytic naevus is accompanied by halo phenomenon. Halo may be atypical, discontinuous and eccentric.

Histopathology of halo naevi:

Compound congenital melanocytic naevus with an eccentric area of regression with inflammatory infiltrate may be seen.

Differential diagnosis

1. Acquired nevomelanocytic naevus:

This is a common mole. It is a collection of nevomelanocytes in the epidermis (junctional), in the dermis (intradermal), or in both areas (compound).[52]

2. Becker's naevus:

This large unilateral lesion is usually seen on the shoulder of males and consists of a sharply but irregularly demarcated area demonstrating hyperpigmentation and hypertrichosis.[52]

3. Café-au-lait macules:

These flat, light brown surface lesions are associated with neurofibromas. Congenital blue naevus: This lesion is a small, well circumscribed, dome-shaped nodule of slate blue or bluish-black color.[52]

4. Dysplastic melanocytic naevi:

A high incidence of melanoma is observed in patients with dysplastic melanocytic nevi. Since removing all the pigmented lesions in these patients is impractical, lesions demonstrating recent changes in color and appearance are removed.

5. Lentigo:

This condition occurs in areas exposed to the sun and possesses a uniform dark-brown color and an irregular outline.

6. Mongolian spots:

These lesions typically occur in the lumbosacral region as a bluish discoloration resembling a bruise.

7. Naevus sebaceous:

This lesion is usually located on the scalp or on the face as a single lesion and is present at birth. A nevus sebaceous is a circumscribed, slightly elevated hairless plaque, typically not pigmented like acongenital melanocytic naevus. In puberty, the lesion becomes verrucous and nodular and may show areas of linear distribution.

8. Naevus spilus:

A naevus spilus is a light brown patch or band that is present since birth. In childhood, it becomes dotted with small dark brown macules.

9. Pigmented epidermal naevi:

This condition is characterized by a persistent linear, pruritic lesion composed of red, scaling, verrucous papules arranged in one or several lines.

10. Melanoma [52]

Investigations:

1. Biopsy:

Complete excisional biopsy can be done which lead to precise diagnosis. Partial punch or shave biopsy may lead to misdiagnosis.

2. Special Stains:

- Silver Stain : Melanocytic naevi can be stained using silver nitrate staining.
- Ammoniated silver nitrate or Masson – Fontanna stain: Produces a densely black reaction product with melanin.
- Dopa Stain:It can be used for melanocytic naevi

3. Immunohistochemical Staining:

By using antibodies to S-100 protein, HMB-45, Melan – A (MART-1) melanocytic naevi can be stained.

4. Dermoscopy (Epiluminescence microscopy):

Synonyms:

Dermatoscopy

Surface microscopy

Incident light microscopy

It is a simple, non-invasive technique in which a liquid, usually immersion oil, is applied to the lesion, which is then examined with a hand-held lens magnification usually 10x or a commercially available device. In experienced hands, it improves both the sensitivity and specificity for the clinical diagnosis of melanoma and other pigmented and non-pigmented lesions,although those without experience and the appropriate training do not realize this benefit.

Morphological features that are otherwise not visible to the naked eye are observed using this technique. It may reveal a reticular pattern or globular / cobblestone pattern in congenital melanocytic naevi and may be useful in the identification of small foci of melanoma.

In melanoma atypical pigment netwok pattern is present with irregular, variable and widened lines that end abruptly at the periphery.Brown globules correlate with pigmented nests of melanocytes in the papillary dermis.Black dots are focal collections of melanocytes and clumps of melanin in the stratum corneum.A blue grey veil represents regression in melanoma.

5. Digital dermoscopy:

It permits computerized digital dermoscopic images to be retrieved and examined at a later date so that comparisons can be made and changes detected over time.

6. Imaging studies:

X-ray back (for congenital melanocytic naevi on posterior axis location) to rule out spina bifida occulta should be done.

- MRI: To rule out

1. Neurocutaneous Melanosis (NCM)

2. Spinal Dysraphism: Occult spinal dysraphism with tethered cord can occur with congenital melanocytic naevi.

MRI Finding:

Asymptamatic NCM:

Focal areas of high signal on T1-weighted images strongly suggestive of melanosis in one or multiple areas of the brain including the temporal lobe, cerebellum, pons medulla. Also middle carnial fossa cyst, chiari type I malformations may be present.

Symptamatic NCM findings:

Evidence of leptominingeal thickening surrounding the brain and spinal cord and mild to moderate hydrocephalus may be present. Signs of increased ICT may be found.

Management:

Two problems to be considered in managing these lesions

First: Potential for malignant change and the resulting need for prophylactic removal.

Second: Cosmetic concern.

Surgical excision with reconstruction is the mainstay of treatment. Chemical peels, dermabrasion, and laser treatments are adjunctive treatment choices. All of the adjunctive treatment methods have been associated with scarring. Furthermore, adjunctive treatment measures have not been demonstrated to decrease the malignant potential. If surgical excision is not feasible, management consists of regular follow-up examination and high-quality photographic documentation for life.

Surgical therapy

- Attempts to remove a large congenital melanocytic naevi should occur early in life, although waiting until age 6 months before operating decreases anesthetic and surgical risks.[53,54,55]

- If direct closure after complete excision is not possible, reconstruction may include serial excision, excision with skin grafts, skin flaps, tissue expansion with subsequent flap rotation or full thickness skin grafting, autologous cultured human epithelium, artificial skin replacement, and free tissue transfer about tissue expansion.[53,55,56]

    - The goals of treatment are to remove all or as much as feasible of the naevi and reconstruct the defect, preserving function and maintaining the aesthetic appearance.

    - Each case requires tailoring of the operation(s) to fit the anatomic defect.

    - The presence of an enlarging nodular mass indicates malignant change and requires immediate treatment. This mass may represent a rare neuroectodermal sarcoma.

    - The incidence of malignant melanoma appears higher in the scalp, back, and buttocks and requires removal first. This increase in incidence is likely secondary to the total body surface area.

- Excision begins in the 6-9 month range, placing procedures 3-6 months apart.

- Special attention was given to giant congenital pigmented nevi of the face by Zuker at the Hospital for Sick Children in Toronto. Complete early excision was recommended because of the cosmetic deformity and because of the life-threatening potential for malignant transformation. [57]

- Cultured epidermal autografts (CEA) have been used successfully to obtain surface coverage after excision of giant hairy nevi.[58]

- Dermal regenerate templates (Integra) prior to skin grafting have been reported in the literature as a substitute for tissue expansion and rotation flaps.[59,60]

- Evaluation of all small and medium congenital melanocytic naevi for prophylactic excision should take place before the patient is aged 12 years. After this age, malignant potential rises sharply.

- Rhodes, Kaplan, and Zaal advocate prophylactic excision of all congenital melanocytic naevi whereas Sahin believes small and medium naevi can be monitored by regularclinical examination.

Adjunctive therapy:

- The phenol chemical peel technique has been used to treat nevi that are too large for excision or that are in locations in which excision would lead to

undesirable scarring. Multiple peels were required, and the best results were in lightly pigmented, superficial lesions. Surgical excision was still deemed the primary intervention. Dermabrasion is useful as an adjunct to increase the depth of the peel and to contour surface irregularities.[61]

- Dermabrasion independently has led to a high incidence of hypertrophic scarring(14.6%) without removal of malignancy risk.
- Multiple treatments with the normal-mode Ruby laser produced immediate thermal damage to the superficial nests of nevus cells and subsequent remodeling of the superficial connective tissue. [62,63,64,65]
    - When the thickness of the subtle microscopic scar reached 1 mm, it masked the underlying residual nevus cells and achieved a good cosmetic result.
    - Follow-up visits for at least 8 years after laser treatment showed no evidence of malignant change in the treated areas.
    - Results of Ruby laser therapy have been varied and malignant risk reduction not determined.
- Successful reduction of pigmentation of giant naevi with high-energy pulsed carbon dioxide laser has been reported. Aesthetic results have ranged from acceptable to hypertrophic scarring (in 50% of patients in one

study).[66,67]

Drawbacks of surgery:

1. A naevus cannot be removed without leaving a scar.

2. It is impossible to remove every cell of a large naevus.

Some people choose to leave their naevi intact. Others choose to remove them. There are good reasons for both choices. Overall it is a highly personal decision.

AIM OF THE STUDY

1. To study the presenting age group and sex distribution of congenital melanocytic naevus.

2. To study the different clinical types of congenital melanocytic naevus.

3. To study the localization of congenital melanocytic naevus.

4. To evaluate the associated cutaneous and systemic conditions of congenital melanocytic naevus.

5. To study the complications of congenital melanocytic naevus.

MATERIALS AND METHODS

In the period between August 2007- September 2009, children and adults who attended the Department of Dermatology, Government General Hospital, Chennai and Dermatology outpatient ,Institute of Child Health, Egmore were included in the clinical study of the congenital melanocytic naevus. They attended Government Hospital for congenital melanocytic naevus as the main complaint or detected during a visit for other dermatological problems.The clinical findings were recorded in the proforma. The studied parameters were presenting age group of congenital melanocytic naevus and sex distribution of congenital melanocytic naevus, clinical types, localization of naevus, presence of hair on the naevus, associated cutaneous or systemic conditions if any, complications, skin biopsy wherever necessary, radiological evaluation in cases involving head and neck region and also treatment of congenital melanocytic naevus in certain cases.

OBSERVATIONS & RESULTS

During the period between August 2007- September 2009 out of the cases attended Dermatology OPD 77 cases were found to have Congenital melanocytic naevus.

Congenital melanocytic naevi are classified according to the size of the lesion into small congenital melanocytic naevi (< 1.5cm), medium congenital melanocytic naevi (1.5- 10 cm), large congenital melanocytic naevi (10-20cm), giant congenital melanocytic naevi (>20 cm).

Prevalence Of Different Types of Congenital Melanocytic Naevi

Table-1

Type of Naevus	No.of Naevus
Small	40
Medium	22
Large	3
Giant	12
Total	77

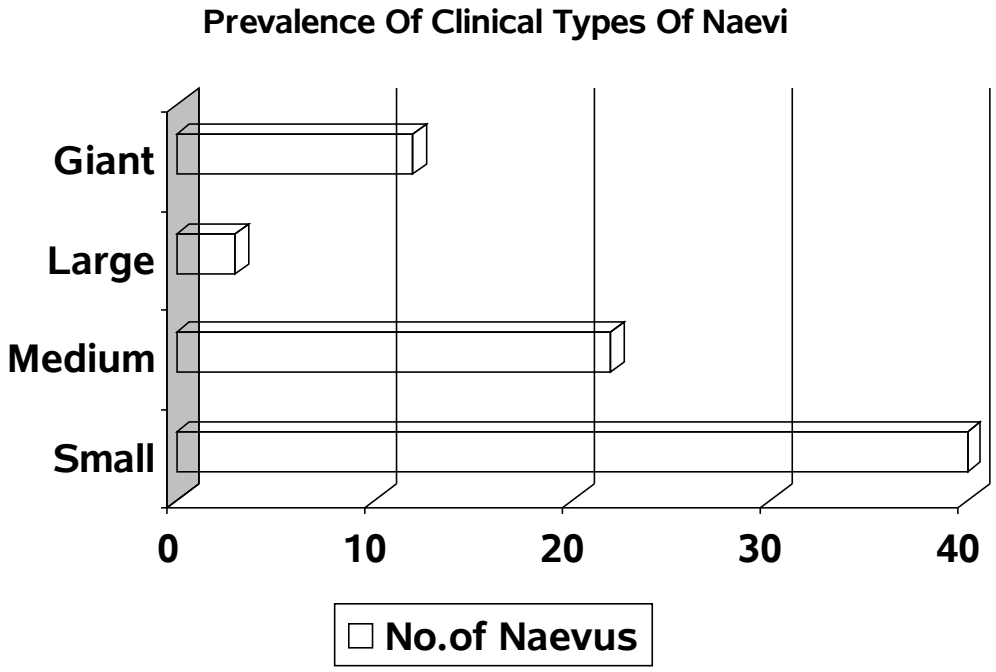

Figure – 1

Presenting Age Group Of Congenital Melanocytic Naevi

Table-2

Type of Naevus	Children (Yrs)						Adol scent	Adults (Yrs)					Total
	< 2	2 -4	4-6	6-8	8-10	10 -12	12 - 18	18 - 25	25-35	35-45	45-55	>55	
Small	2	3	2	2	3	3	7	8	7	1	1	1	40
Mediu	2	1	3	2	1	2	3	2	3	2	1	0	22
Large	0	0	0	0	1	0	0	0	2	0	0	0	3
Giant	2	1	1	10	0	1	0	2	3	1	0	0	12
Total	6	5	6	5	5	6	10	12	15	4	2	1	77

Presenting Age Group Of Congenital Melanocytic Naevi

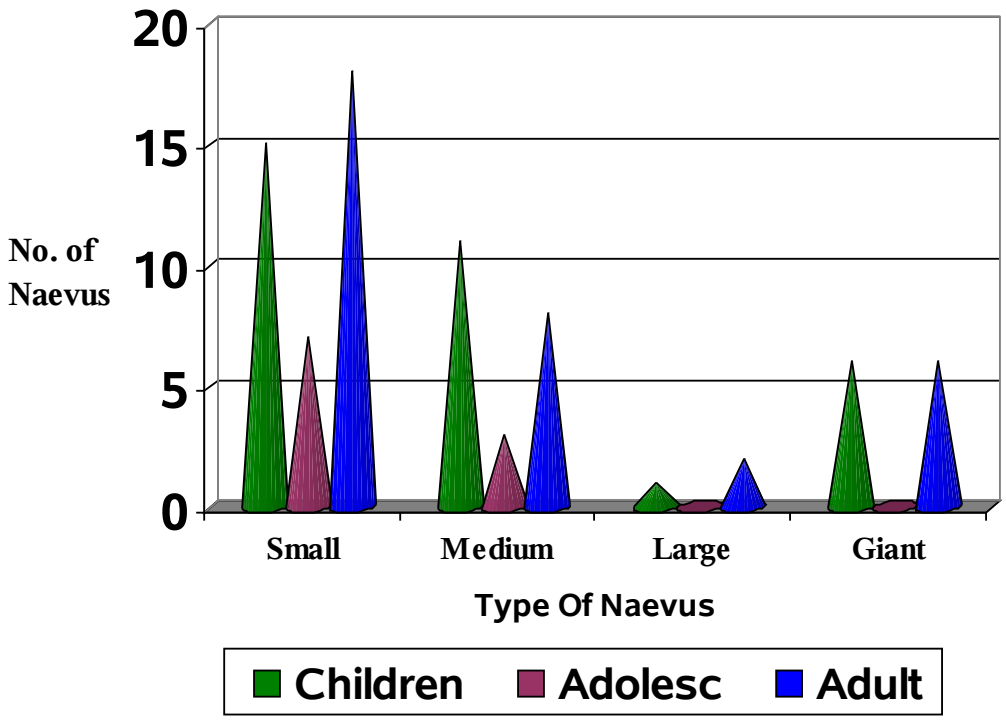

FIGURE-2

Considering the type of congenital melanocytic naevus and age at inclusion in the study, we found more or less homogenous distribution of small congenital melanocytic naevus, medium congenital melanocytic naevus , large and giant congenital melanocytic naevi in children and adults.

In adolescent age group, small, medium, large and giant congenital melanocytic naevi, were less in number than in children and adults. The reason for this finding could be, the number of adolescent population included in this study was less in number.

Sex Distribution Of Congenital Melanocytic Naevi

Table-3

Sex	Type of Naevi				Total	%
	Small	Medium	Large	Giant		
Male	17	10	2	7	36	46.75
Female	23	12	1	5	41	53.25

Out of 77 Congenital melaocytic naevi , 36 naevi were found in males and in females 41 were noted comprising 46.75% and 53.25% respectively.

Percentage Of Sex Distribution

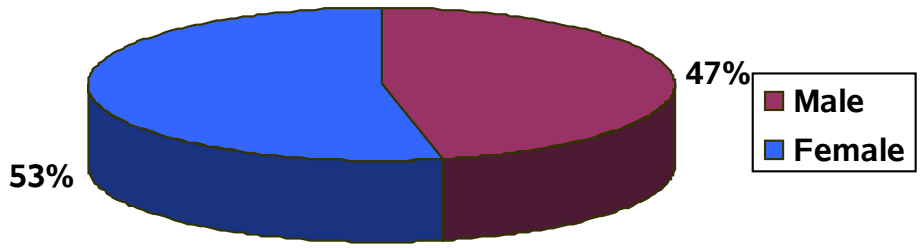

FIGURE-3

Localisation Of Small and Medium Congenital Melanocytic Naevi

Table-4

Localisation	**Type of Naevi**				**Total**	**%**
	Small	**%**	**Medium**	**%**		
1.Head & Neck	18	28.13	7	10.93	25	39.06
2.Chest & Back	14	21.88	7	10.93	21	32.81
3.Abdomen & Groin	3	4.68	4	6.25	7	10.93
4.Upper Limbs	5	7.81	3	4.69	8	12.5
5.Lower Limbs	1	1.56	1	1.56	2	3.12
6.Genitals	0	0	0	0	0	0
7.Soles	1	1.56	0	0	1	1.56

25 congenital melanocytic naevi were located on the head and neck , with small congenital naevi 18, and medium naevi 7, comprising totally 39.06 %.

21 cases of congenital melanocytic naevi were on chest and back , with small naevi 14 and medium naevi 7,comprising totally 32.81%. 8 cases of congenital melanocytic naevi including 5 small and 3 medium naevi were found on upper limbs with 12.5 %. 7 cases of congenital melanocytic naevi with small naevi 3 and medium naevi 4 were on abdomen with 10.93%. One small and one medium naevi were noted on lower limbs comprising 3.13 %. . One small type of congenital melanocytic naevus was found on the sole.

Localisation of Small and Medium Naevi:

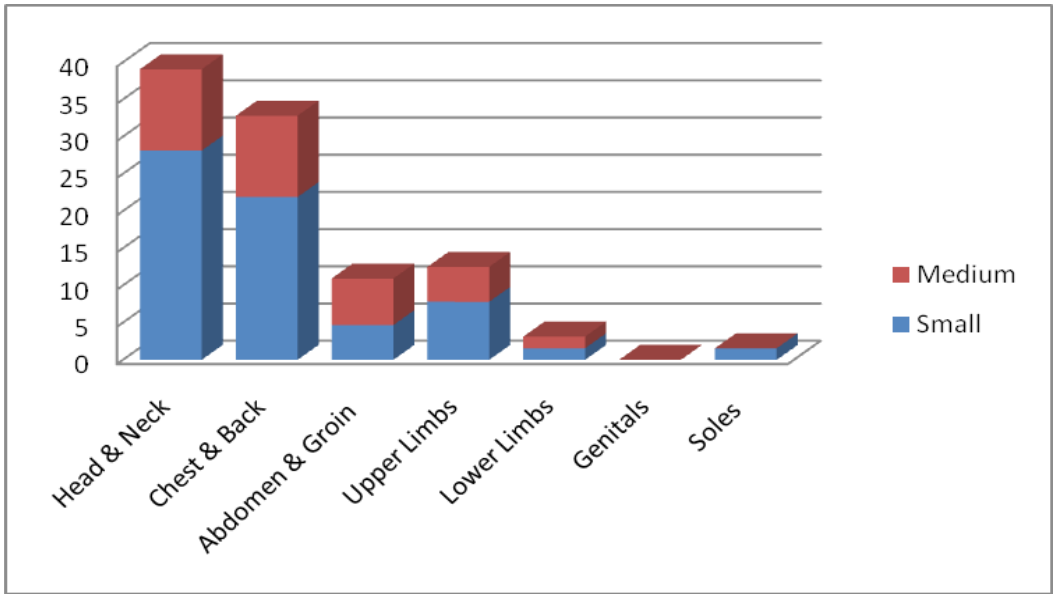

FIGURE- 4

Distribution of Large and Giant naevi:

Large naevi were noted in 3 cases . One large naevus was located on the left eyelid and in another case naevus was noted over the left forearm.One more case of large naevus was found on the right side cheek.

Giant Naevi:

2 giant naevi of bathing trunk distribution were noted.

5 naevi were on scalp ,involving in some cases face, neck, upper back

3 naevi were on lower limbs.1 giant naevus was on upper back

Multiple small and medium sized naevi were distributed over face, trunk, genitalia, and lower extremity in one case.

Associations of Congenital Melanocytic Naevi

Table-5

Associated conditions	Type of Naevi			
	Small	**Medium**	**Large**	**Giant**
1.Neurofibroma	0	0	0	3
2.Vitiligo	0	2	0	2
3.Naevus spilus	1	0	0	0
4.Portwine stain	2	0	0	0
5.Naevus of Ota	1	0	0	0
6.Halo phenomenon	1	0	0	0
7.Poliosis	0	2	0	1
8..Lipoma	0	0	0	2

Neurofibroma was found to be associated with 3 cases of giant congenital melanocytic naevi .This was the most common association noted with giant congenital melanocytic naevus in this study.

Vitiligenous depigmentation was noted within the giant naevi in 2 cases whereas in other 2 cases of medium naevi depigmentation was present surrounding the naevi. Also associated mucosal vitiligo and vitiligo in distant skin sites were present in 2 cases. Halo depigmentation was present around one case of small naevus on the cheek.

Poliosis mainly over the eyelid kissing naevus in 2 cases and in the hair over the giant congenital bathing trunk naevus was noted. In 2 cases it was associated with vitiligo.

Lipoma was found in 2 cases of giant bathing trunk congenital melanocytic naevi over the lower back in the midline .

Naevus spilus, was found to be associated with a case of small naevus. In a case naevus of Ota and portwine stain was found to be associated with a small congenital melanocytic naevus.

Distribution Of Different Types Of Hair In Naevi

Table-6

Type of Naevi	Presence of Terminal hair	%	Presence of vellus hair	%
1.Small	14	18.18	0	0
2.Medium	12	15.58	0	0
3.Large	1	1.30	0	0
4.Giant	7	9.09	3	3.89
Total	34	44.15	3	3.89

Percentage Of Terminal And Vellus Hair

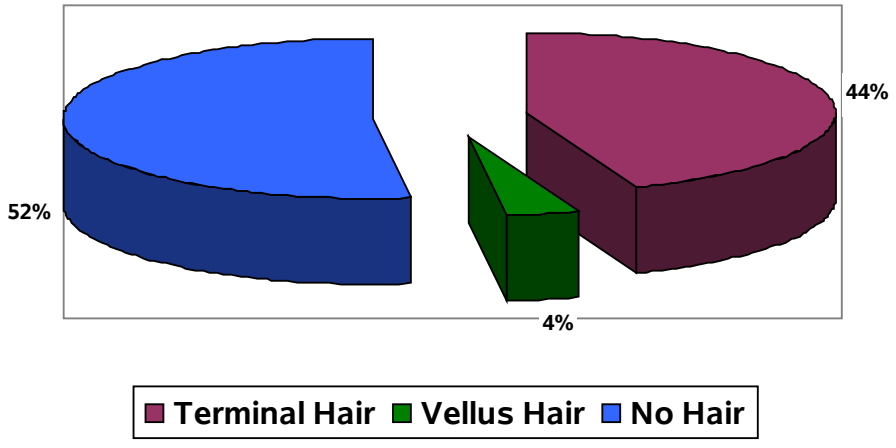

FIGURE- 5

In 77 cases of congenital melanocytic naevi, terminal hairs were present in 34 naevi, which comprises about 44.15 %. Increased vellus hairs were present in giant naevi comprising 3.89 %.

Terminal hairs are pigmented medullated hairs whereas vellus hairs are unpigmented non-medullated hairs. Increased vellus hairs on the naevus were seen mostly in young children and terminal hairs on the naevus were seen in both adults and children.

Complications Of Giant Naevi

Type of Naevi	**Complications**			
Giant	**Ulceration**	**Pruritus**	**Hypertrophic scarring & Keloid**	**Proliferative Nodules**
	1	2	1	1

Ulceration was noted in a case of giant congenital melanocytic naevus over the scalp in a child.

Pruritus was present in giant naevi over the face in one case and in another case over the scalp.

Hypertrophic scarring after plastic surgery in both the arms and also

keloidal nodules over the chest after minor trauma were noted in a child with bathing trunk naevi. Proliferative nodules were found in a case of giant naevus.

Complications Of Giant Congenital Melanocytic Naevi

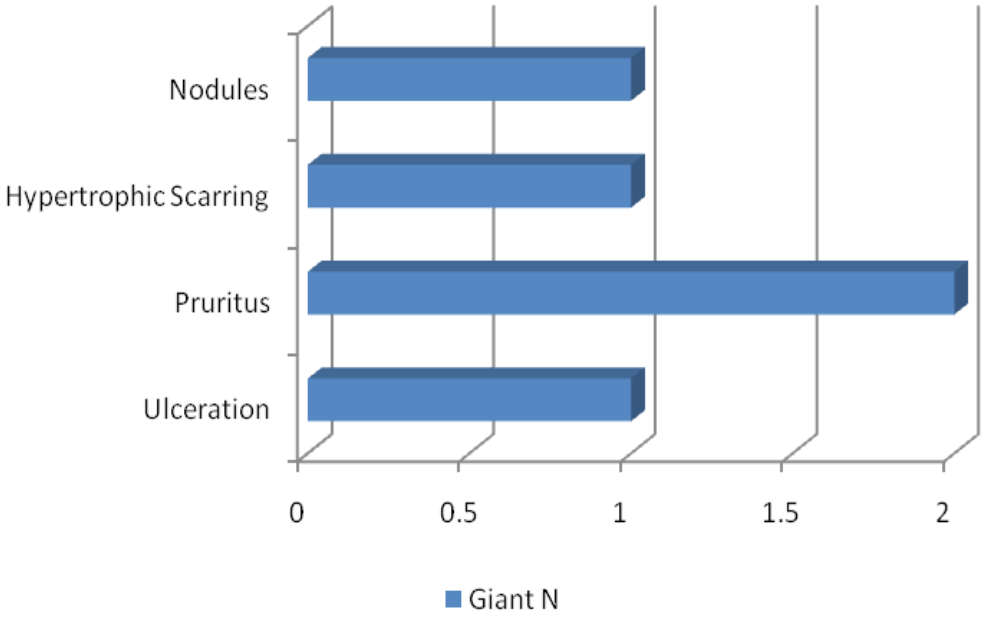

FIGURE: 6

Incidence Of Melanoma:

Out of 77 studied congenital melanocytic naevi, which were followed up for 1- 3 yrs, there was no case of cutaneous melanoma or any other cutaneous malignancy. Extracutaneous meningeal melanoma was not found by CT scan in cases involving head and neck region.

Nodular proliferations were noted in a case of giant naevus over the left cheek. Histopathological examination of these nodular proliferations were consistent with congenital melanocytic naevi.

Biopsy of giant congenital melanocytic naevi was done. Biopsy specimens taken from neurofibroma like nodules present over the naevi, showed features of neurofibroma. Biopsies taken from pigmented lesion showed features consistent with congenital melanocytic naevi.

Special staining was done in 2 cases using Masson Fontanna stain, which showed features of melanocytic naevus as black deposits of melanin in the dermis.

As treatment wide excision and split skin grafting in stages was done for 2 cases of giant congenital melanocytic naevi. Simple excision of melanocytic naevus of small size was done in one case as the patient was willing for excision. In a case of giant congenital melanocytic naevus with neurofibroma, excision of neurofibroma present over the gluteal region ,as it caused discomfort to the patient was done.

DISCUSSION

Nurimar Conceicao Fernandes et al[68] in Brazil and Vito Ingordo et al[69] in Italy, have conducted clinical study and epidemiological study of congenital melanocytic naevus respectively.

The distribution of different types of naevus in our study was homogenous in children and adults which was comparable to the above study[68] of Nurimar Conceicao Fernandes. In contrast in adolescence it was low which may be due to inclusion of low number of adolescent population in our study.

Out of 77 cases only 10 cases were adolescent, and there are equal number of 33 cases in children and adult groups. So distribution of congenital melanocytic naevi were equal in children and adult age groups and low in adolescent groups in our study.

In our study sex distribution of congenital melanocytic naevi were 46.75% in males and 53.25% in females. This was comparable with the above study[68]of Nurimar Conceicao Fernandes. In the Dawson HA et al study,[70] in a sample of 133 congenital melanocytic naevi of different sizes, the authors reported a 3:2- female : male gender ratio.[70] In Castilla EE et al study ,no significant difference was observed concerning gender. In our study sex did not influence the clinical type of naevi.

There is great controversy in the classification of congenital melanocytic naevi.The one that seems to be most appropriate classification and adopted by most authors is:[71]

Small naevus: < 1.5 CM

Medium naevus: 1.5 cm – 10 cm

Large naevus: 11cm – 20 cm

Giant naevus: > 20 cm

In our study we observed 40 cases of small naevi comprising 51.94%, 22 cases of medium naevi, with 28.57 %, large naevi 3 in number with 3.89% and 12 cases of giant naevi with 15.58%.These results were comparable with the Nurimar Conceicao Fernandes et al study[68].

Small naevi congenital melanocytic naevi are located predominantly on the head and neck, followed by chest & back and in upper limb abdomen & groin, lower limb in decreasing order of frequency in our study.

One case of small naevi was observed in the plantar region. These findings of predominant localization in head and neck area were in contrast to the Nurimar Conceicao Fernandes et al study[68] where small congenital melanocytic naevi were located predominantly on the trunk.

Medium congenital melanocytic naevi were homogenously distributed in the head & neck and chest & back but in low concentration in abdomen and groin, upper limb, lower limb in decreasing order of frequency.

This was comparable with the Nurimar Conceicao Fernandes et al study[68.] No small or medium naevus was present in the palmar or plantar region in the Nurimar et al[68] and Vito Ingordo et al studies[69].

Small congenital melanocytic naevi were single in 33 cases out of 40 cases, whereas it was multiple in 7 cases comprising 82.5 % and 17.5 % respectively.

In 77 moles terminal hairs were present in 44.15 % and vellus hairs in 3.89 %.Increased vellus hairs on naevus were seen in only children.Terminal hairs were found both in adults and children.This was comparable to the Vito Ingordo et al study[69].

The development of acquired melanocytic naevus may be related with some factors like type of skin, ethnicity,genetic predisposition and UV light exposure. More common in children of fair skin, blonde hair, blue eyes . It is also more common in areas chronically exposed to sunlight[68]. This fact has not been described for congenital melanocytic naevi.

Large naevi were located in one case over the left eyelid, and in another case on the left forearm. One more case of large naevus was found on the right side cheek.

Kissing naevi involving both upper and lower eyelids which were contiguous in nature were found in 3 cases.This shows the time of development of congenital melanocytic naevi which is after 50 days but before 6 months of gestational age when the eyelids split.

Treatment with Nd:YAG laser was tried in one case of large congenital melanocytic naevus (kissing naevus) on the left eyelid.

There was repigmentation after treatment with laser and successful result was not found.

Giant naevi were distributed more on the head and neck region. 2 cases of bathing trunk naevi, 3 cases involving lower limb and 3 cases involving genitalia were found.

Multiple small and medium sized naevi distributed over scalp, face, trunk, genitalia, extremities were noted in one child.

Out of 12 cases of giant congenital melanocytic naevi hairy component was present in 10 cases which showed terminal hairs in 7 cases and vellus hairs in 3 cases comprising 9.09 % of the total 44.15% of naevi with terminal hairs and 3.89% of vellus hairs respectively.

Neurocutaneous melanocytosis is a rare congenital disorder in which leptomeninges contains excessive layers of melanocytes and melanin.Large, giant congenital melanocytic naevi on the neck, head, and posterior midline is a risk factor for development of neurocutaneous melanocytosis[39].

Although many giant naevi were noted over head and neck ,CT scan of these patients were normal.MRI has higher sensitivity in detecting neurocutaneous melanocytosis within the first 4 to 6 weeks of life but it requires general anaesthesia.

Some authors advocate for MRI or serial neurological examination for asymptamatic patients with risk of developing leptomeningeal melanocytosis. Others question the performance of MRI as there is no treatment for cases of asymptomatic melanocytosis [39].

Out of 77 cases of congenital melanocytic naevi , which were followed up for 1-3yrs there was no case of cutaneous melanoma. None of the examined subjects reported a personal or family history of melanoma.

The studied literature refers that large and giant congenital melanocytic naevi have higher risk of developing melanomas and that the risk of medium and small congenital melanocytic naevi is still controversial.

The present sample of large and giant congenital melanocytic naevi is too small to asses the risk of malignant transformation and the follow–up time of small and medium naevi was not enough to assess malignant transformation, given that it happens only after adolescence in children. In small and medium naevi of adults also no case of melanoma was noted.

Ulceration, pruritus of giant naevi present over the scalp were noted in cases of giant naevi in children , hypertrophic scarring, keloid over the arms and chest as a result of surgical excision and grafting treatment of bathing trunk naevi, in a child was noted. Proliferative nodules of giant naevi present over the face was noted in an adult case.

Appearance at birth or later of a nodular or hyperpigmented area within a congenital melanocytic naevus simulates malignant melanoma and prompts biopsy. Although their clinical and pathologic features seem ominous, proliferative nodules typically are benign and may regress, although atypical features cause greater concern.

In a study[86] a subset of 30 samples containing both the congenital melanocytic naevus and proliferative nodules was analyzed for immunohistochemical reactivity for melanocytic (S-100 protein, HMB45, melan-A), lymphocytic (CD45), cell-cycle/proliferative (Mib-1, p16, p21, p27, cMyc), apoptotic (p53, Bax, c-kit, CD95), and anti-apoptotic (bcl-2) markers.

Both congenital melanocytic naevus and proliferative nodules had similar expression of melanocytic, lymphocytic, and most cell-cycle/proliferative and apoptotic markers, including Mib-1, p16, p21, p27, c-Myc, Bax, CD95, and bcl-2. A greater proportion of proliferative nodules than congenital melanocytic naevus were reactive for p53 and c-kit. p53 and p21 expression in congenital melanocytic naevus and all types of proliferative nodules were inversely correlated.

When ordinary and atypical proliferative nodules were compared, the atypical proliferative nodules more frequently expressed p53, Mib-1, Bax, and bcl-2, but less frequently expressed p21. The c-kit expression in nearly all proliferative nodules and its absence in nearly all congenital melanocytic naevus is potentially useful for recognition of proliferative nodules, suggests a delayed melanocytic maturation process in proliferative nodules, and may be likely indicative of their benign nature. p53 reactivity in concert with a lack of p21 up-regulation by immunohistochemistry suggests that a p53 mutation may be present in proliferative nodules, although the immunohistochemical findings alone cannot exclude possible overexpression of wild-type p53. Regressive, involutional, or maturational changes were observed in sequential samples from 4 patients. No patient developed malignant melanoma or another melanocytic nevus-associated malignancy during the follow-up period.

These findings underscore the similarities between proliferative nodules and the underlyingcongenital melanocytic naevus and suggest that maturational, proliferative, and apoptotic processes are involved in their clinical evolution. There have been reports of balloon-cell change in a large congenital melanocytic nevus[87.]

Neurofibroma was found in association with giant naevi in 5 cases.There are several reports of the association of garment naevus with neurofibroma, lipoma, and spina bifida occulta.[73] The authors have explained the constellation of these findings on the basis of a defect in the neural crest, which is considered to be a common origin of melanoblasts, schwann cells, sensory ganglia, bone, fat, muscle and blood vessels.

Immunohistochemistry helps in differentiating congenital melanocytic naevi nodules from neurofibroma. S100 staining is present only in melanocytes but not in neurofibroma or schwannoma.Myelin basic protein, glial fibrillary acid protein are positive in neurofibroma.

Lipoma was noted in 2 cases of giant congenital melanocytic naevi.The melanocortin neurons and melanocortin receptors in the central nervous system directly and potently affect cellular glucose utilization, lipid uptake and triglyceride synthesis in the periphery. This most likely occurs through effects on the autonomic outflow, thereby efficiently shifting substrate metabolism to modulate energy storage and adiposity.

The authors of this study conclude that largely independent of changes in food intake, the central nervous melanocortin system directly and rapidly controls triglyceride synthesis, lipid deposition and lipid mobilization in white adipose tissue.

The sympathetic nervous system connects white fat cells directly with homeostatic control areas in the central nervous system including melanocortin positive neurons in the hypothalamus as well as in other non-hypothalamic forebrain areas, midbrain and brainstem areas.

The central nervous melanocortin system modulates sympathetic nerve activity in white adipose tissue and that sympathetic nervous system signaling via functional β-adrenergic receptors is required for the central nervous melanocortin system-induced changes in adipocyte metabolism. [74]

These mechanisms may explain the occurrence of lipoma in giant congenital melanocytic nevus. There are pluripotent stem cells in the dermis and subcutis, which, on appropriate stimulus, differentiate into a specific tissue. This would probably explain the occurrence of lipoma in a giant congenital melanocytic naevus and the same could be the reason for development of neurofibroma in a melanocytic naevus.

Vitiligo in association with congenital melanocytic naevus is a rare phenomenon; Nevertheless several reports have been published in the literature.[75,76,77] Vitiligo onset removes congenital naevocellular naevus cells[84]

In our study vitiligo was associated with medium and giant congenital melanocytic naevi.Case of medium sized congenital melanocytic naevus, kissing naevus of the eyelid developed vitiligo like lesions surrounding the naevus.In other medium sized naevus over the right cheek there was development of halo depigmentation around the naevus, following which she developed vitiligo like lesion in other areas of skin and mucosa.

In giant naevi, in between the naevus vitiligenous areas were present in 2 cases. Out of these 1 case also developed vitiligo in other areas of skin and oral mucosa.

Pigment regression is a process in which dermal melanocytes loss their capability to produce pigment, and what is left is only the basal layer of melanocytes. Many theories have been proposed to explain these pigmentation abnormalities.

They are natural precursors of melanin would be inhibitors of their production; as a consequence, melanocytes would lose the protective mechanisms that disable these precursors. The immune theory is the most accepted one for suppression of pigment production through humoral and cellular immunity action. Another hypothesis is that neurotoxic agents would be released close or inside the melanocytes, destroying melanin-producing cells or simply stopping their production. Transepidermal elimination of nevus cells as well as fatty degeneration are also mentioned. The antibody formation against melanocytes in naevus may cause vitiligo in other areas of skin in susceptible persons.

In the kissing naevus of the eyelid, white hairs of the eye lashes were present within the vitiligo patch.

Halo depigmentation around small congenital melanocytic naevus was noted.This naevus may undergo spontaneous regression.There are studies[79,80] reporting halo naevi with demonstration of T cells around naevi, showing the involvement of T cell immunity in regression of naevi.

Poliosis ,a localized patch of white hair resulting from the absence or deficiency of melanin in a group of neighbouring follicles was noted in a case of giant bathing trunk naevus who also had mucosal vitiligo and depigmented patches in between the naevus and in a case of kissing naevus of the eyelid without vitiligo.

A case of small naevus was found to be associated with naevus of Ota and portwine stain on either side of the face. Small naevus was present on the side of the portwine stain over the cheeks. Association of naevus with vascular malformation could be an example of twin spotting, phacomatosis pigmentovascularis. Very extensive congenital melanocytic naevus may be intermixed with elements of vascular malformation,and haemangiomas.[24]

In this study naevus spilus was present in an infant with small naevus on other body site.In this case naevus spilus could have been present since birth. There are studies [81,82,83] reporting that naevus spilus could be a congenital melanocytic naevus.

CONCLUSION

- The sex distribution of congenital melanocytic naevus was more in females than males in this study.

- Congenital melanocytic naevus is classified based on the size of the lesion as small, medium, large, and giant naevus.In this study prevalence of small naevi was more ,followed by medium, giant and large naevi in descending order of frequency.

- Localisation of small naevi were more on the head and neck region, whereas medium naevi were homogenously distributed in the head and neck ,and chest and back region in this study.

- In this study, most of the small congenital melanocytic naevi were single in distribution rather than multiple.

- Kissing naevus of the eyelid (Congenital divided naevus) which falls into the group of medium and large naevus was found in 3 cases which shows the time of development of congenital melanocytic naevi.

- Giant naevi of bathing trunk distribution and localization over the head and neck region, posterior axis were noted both in children and adults.CT scan of these patients were normal and involvement of central nervous system was not found in any of these cases.

- Interesting cutaneous associations like neurofibroma, vitiligo, poliosis, lipoma were observed with giant naevi in certain cases.

- Complications of giant naevi like ulceration, pruritus, hypertrophic scarring, keloid, proliferative nodules were noted in cases of giant naevi.

- Small congenital melanocytic naevi were found to be associated with halo depigmentation, portwine stain, naevus of Ota, naevus spilus in this study.

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PROFORMA

Name: Age: Sex:

Occupation Address:

Informant:

H/o present illness:

H/o onset of naevus

H/o any increase in size

H/o any change in shape

H/o any change in colour

H/o pain

H/o itching

H/o ulceration

H/o any oozing or bleeding

H/o any trauma

H/o any altered sensation

H/o any other swelling in the body

H/o any delay in developmental mile stones

H/o seizure

H/o any increase in the head circumference

H/o extended toilet training

H/o urinary incontinence

H/o tingling, pins and needles in foot

H/o numbness of feet and toes

H/o leg pain (Sciatica like)

H/o low back ache / weakness in legs

H/o new or progressive scoliosis or kyphosis

H/o any other bony deformity(club foot)

Family History:

- Any other sibling affected with congenital melanocytic naevus
- H/o melanoma in any other family members.

Personal History:

Adults: Nature of work (Prolonged sun exposure)

Past History:

H/o any previous treatment.

General Examination:

Adults: As a routine

Anemia / clubbing / cyanosis / jaundice / pedal edema / lymphadenopathy.

Pulse :

BP :

Systemic : CVS / RS / Abdomen / CNS / Ocular Examination

Skeletal : Lower Back:

Examination
Any other skin lesion – Neurofibroma
Hairy patches
Dimples
Fatty tumors
Any foot or spinal deformity

Children:

- Developmental mile stones
- Head circumference
- CNS: Any focal neurological deficits
- Any Anorectal abnormality
- Any skeletal deformity (foot deformity or scoliosis),

DERMATOLOGICAL EXAMINATION

Morphology:

Size -

Shape -

Border -

Surface -

Pigmentation -

Presence of hair -

Number of nevus -

Site: Head / Neck / Chest / Back / Abdomen / UL / LL

Side: Whether right side
left side
mid line

Any atypical features:

Nail Examination

Hair Examination

Other Associated findings:

1. Neurofibromatosis
2. Lipomas
3. Halo phenomenon
4. Vascular naevi
5. Neurocutaneous melanosis
6. Spinal dysraphism
7. Club foot / Pes cavus / Hammer toes

Complications if any

1. Malignant melanoma
2. Extracutaneous Melanoma: Leptomeningeal melanoma
3. Rhabdomyosarcoma
4. Liposarcoma
5. Fibrosarcoma
6. Schwannoma

Investigations

- Biopsy
- x-ray (for CMN on the posterior axis location)
- CT Scan (for CMN on the posterior axis location)