

Case Report

Giant congenital melanocytic nevus scalp: report of a rare case

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ABSTRACT

Giant congenital melanocytic nevus (garment nevus) is a kind of congenital malformation of neural crest cells with size greater than 20 cm. Malignant melanoma may develop in 2-31% of these lesions. The objective of this paper is to present a unique case of giant nevus which is rare.

Keywords: Garment nevus, Scalp, Rare

CASE REPORT

An 18 year old female was admitted in the plastic surgery department in August 2010 with multiple nodules of variable sizes on the scalp present since birth. The patient was born with a soyabean-like swelling on her left side of scalp. The skin and hair covering the neoplasm were normal. With the patient's growth, the mass enlarged gradually, and the covering skin became thick and wrinkled without other complicated symptoms. In the past 5 years before the admission, the lesion enlarged quickly and covered most of the scalp. The patient had normal consciousness and intelligence without any history of traumatic brain injury, chronic eczema, pyoderma, psoriasis, endocrine disorder, or cancer. She was born of a nonconsanguineous marriage. There was no similar case in her family.

On admission, physical examination showed a good physical development and nutritional status. The patient's head size was in a normal range. The mass on the scalp sized 22 cm × 18 cm x 2.5 cm approximately with deep thick wrinkles spread across the whole of the scalp (Figure 1). The wrinkles were about 2 cm in width and 1.5 cm in height with normal skin, with multiple nodules and focal ulceration of the skin. The whole mass looked

like the gyrus or garment over the head. The skin on the face, forehead and other regions was not thick or wrinkled. The patient had a few pigmented nevi on the extremities, some of which were hairy. Enlargement of local lymph nodes were present.



Figure 1: Clinical photograph of 18 year old female patient with giant congenital melanocytic nevus.

Laboratory examination showed normal blood, urine and stool routine with normal liver and kidney functions. X-ray of the chest and head, MRI and CT of the brain, and ultrasonography of the abdomen did not reveal any abnormality. The differential diagnosis offered was nevus/ hamartoma/diffuse neurofibroma. Complete resection of the lesion was performed.

GROSS: Received 25x20x5cm skin covered entire scalp. Surface grey tan and hair present. Elevated nodules of variable sizes present largest measuring 4x4cm. Cut section shows grey brown and dark brown areas (Figure 2 & 3). Adjacent 2 lymph nodes identified.



Figure 2: Gross photograph of specimen with deep thick wrinkles about 2cms in width and 1.5 cm in height with normal skin with multiple nodules and focal ulceration.



Figure 3: Cut section of the specimen, thickened skin with greyish white lesion in the dermis.

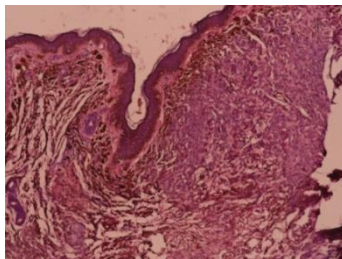


Figure 4: Photomicrograph showing surface squamous epithelium with subepithelial sheets and nests of round to polygonal nevus cells uniform nuclei with moderate amount of melanin pigment (Hematoxylin and Eosin, x100).

Histological examination showed surface hyperplastic squamous epithelium. Below the epithelium sheets of nevus cells were located in the reticular layer of the dermis extending into subcutaneous adipose tissue. The nevus cells were separated by thin collagen fibers. The mature nevus cells were arranged in nests with moderate amount of melanin pigment with bland nuclei (Figure 4 & 5). There are foci of foreign body giant cells around the hair follicles (Figure 6). The deep nevus cells were spindle shaped (Figure 7). Thus the mass was diagnosed as a giant congenital melanocytic nevus. Sections from the lymph node showed features of reactive follicular

hyperplasia. There is no evidence of malignancy. Thereafter, dermoplasty was carried out with application of graft. The graft was rejected due to skin infection. The infection was controlled and reapplication of the graft was done. The patient was uneventful during two years follow-up and at present she is doing well.

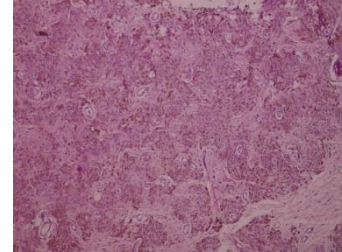


Figure 5: Photomicrograph showing nests of round to polygonal nevus cells in upper dermis (Hematoxylin and Eosin, x100).

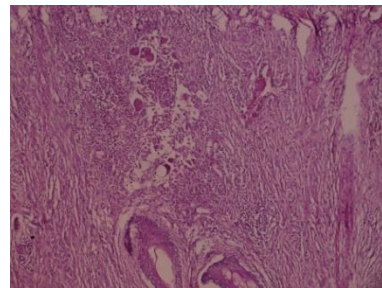


Figure 6: Photomicrograph showing nests of spindle nevus cells in dermis with foci of inflammatory cells consisting of lymphocytes, plasma cells and foreign body giant cells (Hematoxylin and Eosin, x100).

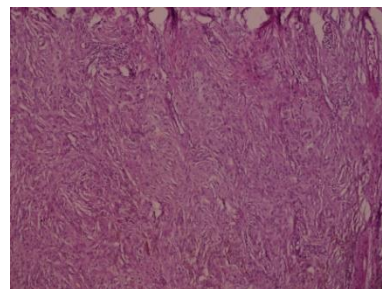


Figure 7: Photomicrograph showing nests of spindle nevus cells in deep dermis (Hematoxylin and Eosin, x100).

DISCUSSION

Congenital melanocytic nevi is neurocristopathy (a disorder of the development of the embryonic neural crest), with unknown etiology. Majority of Congenital melanocytic nevi, develop during first trimester of pregnancy. Vast majority are sporadic but familial cases are also reported.¹

Congenital melanocytic nevi vary greatly in size, macroscopic appearance and histology. There is a practical need to subdivide congenital nevi according to size, since size differences have a direct bearing on cosmetic, therapeutic options and probably on the chance of malignant transformation. James William D et al² divided congenital melanocytic nevi into three types, small sized congenital nevus < than 2 cm, medium sized nevus 2cm to less than 20cm and giant congenital melanocytic nevus (giant pigmented nevus, giant cerebriform nevus, garment nevus, bathing trunk nevus etc) > 20 cm and size more than 40cm has increased risk of malignant transformation to melanoma. Zeal LH et al³ reviewed literature and various classifications and recommended defining giant congenital nevi as congenital nevi covering 1% body surface area in the face and neck and 2% elsewhere on the body.

Giant pigment nevus (giant cerebriform nevus, garment nevus) is found in 1% of infants at birth and lesions of scalp and forehead are even rarer at birth.^{4,5} Giant pigment nevus is present in all ethnic groups, both genders with slight female predominance. Majority of the patients have normal adult lives without complications.

Clinically the lesions are often flat at birth, brown to black in color. They often grow proportionally to the body size as the child matures. As they mature they often develop thickness, become elevated although these features can be present from birth. Large congenital melanocytic nevus is a darkly colored circumscribed area of skin, sometimes covered with dense hair or hypertrichosis or proliferative nodules and / or accompanied by multiple small satellite nevi that develop at birth or during early childhood (tardive satellites).⁶

Giant congenital melanocytic nevi predispose to malignant melanoma with a reported incidence of 2% - 31%.⁸ A review of literature shows that approximately 70% of the melanoma that have occurred in patients with giant congenital nevi have occurred before puberty. Some of these melanoma have occurred in extra cutaneous sites, especially the CNS.⁷

Turkman et al evaluated⁸ 60 cases of giant congenital nevus and followed them from 1997 to 2007. In his study 34 were males, 26 were females with average age of 17.4 (range 3-32 years). The nevi was evaluated using three different classification methods. They recorded the total area of nevus in cm², greatest nevus dimensions and percentage of nevus surface area to total patient body surface area. Malignant transformation was noted in 15.4% of participants with congenital nevi, which was

confirmed on histopathology after excision. They recommend that the calculation of the total nevus area in cms is most useful method for assessment of the risk of developing melanoma in congenital nevus. Because of the premalignant potential, it is an acceptable clinical practice to remove congenital nevi electively in all patients and relieve the neurocytic load. Surgical excision is the standard treatment.⁹

CONCLUSION

Congenital giant melanocytic nevi (garment nevus) are rare scalp lesions which require surgery in early childhood because of their growth potential and possible malignant behaviour. The objective of this paper to present is a Giant congenital melanocytic nevi present since birth for 18 years without malignant behaviour.

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