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Congenital and infantile malignant melanoma of the scalp: A systematic review

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Review

Congenital and infantile malignant melanoma of the scalp: A systematic review

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HIGHLIGHTS

- Congenital and infantile malignant melanomas are rare.
- Surgery is the mainstay of treatment.
- Prognosis of the condition remains poor.

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ABSTRACT

Congenital and infantile malignant melanomas are rare and typically carry poor prognosis. The purpose of this article was to review the data on congenital and infantile malignant melanomas of the scalp in order to understand its presentation, diagnosis, management, and outcomes of congenital melanoma of scalp. We searched PubMed, CINAHL and Cochrane databases. Ten cases of congenital and 3 cases of infantile malignant melanoma of scalp were identified. The diagnosis was confirmed by biopsy and histological analysis for confirmation. The prognosis depends on the origin of disease (congenital melanocytic nevus, transplacental metastasis, or de-novo), tumor thickness, the presence of ulceration and/or necrosis, and anatomic site (scalp lesions having poor prognosis). The most commonly used treatment of the reported cases of congenital and infantile melanoma was surgical excision of the primary lesion. Further modes of treatment may be extrapolated from the treatment of childhood and adult melanomas.

Contents

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1. Introduction

Malignant melanoma is a neoplasm arising from the melanocytes. Melanocytes are predominantly found in the basal layer of the skin epidermis (at the junction of epidermis and dermis). They are also found in the uvea of the eye, meninges, and ectodermal mucosa. The average age at diagnosis of melanoma is 57 years and is more common in men [1].

Melanoma in the children accounts for 1%–4% of all cases of melanoma, and 1%–3% of all pediatric malignancies [2]. Pediatric melanoma can be divided into four categories based on the age of occurrence: congenital (in utero to birth), infantile (from birth to 1 year), childhood (1 year to puberty) and adolescent melanoma (puberty to 21 years) [3]. Congenital and infantile malignant melanomas are the least common variety of pediatric melanomas. Only 36 cases of congenital and infantile melanomas have been reported in literature since 1925 [4–12].

Congenital or infantile Malignant melanomas are further grouped into three categories based on the origin of the disease: 1) arising within congenital melanocytic nevus (benign proliferation of melanocytes), 2) arising de-novo (from non-lesional skin) and 3) from transplacental metastasis (from maternal lesions) [13]. The purpose of this this article was to review the data on congenital and infantile malignant melanomas of the scalp in order to understand its presentation, diagnosis, management and outcomes.

2. Methods

We searched PubMed, CINAHL and Cochrane databases for articles on congenital or infantile malignant melanoma of scalp. Search strategy was to use MeSH terms of [Malignant melanoma] and [Scalp]. The search was further refined by using [infants] or [infant] or [Congenital]. The search was run on 13th April 2016 and repeated on 10th May 2016 by two authors independently. We included all the studies on malignant melanoma of scalp irrespective of study design and date of publication. Selected studies were critically reviewed for a descriptive review. We followed PRISMA guidelines [14] for reporting of the review which was registered with research registry UIN: www.researchregistry.com.

3. Results and discussion

3.1. Epidemiology of congenital or infantile malignant melanoma of the scalp

Our literature search revealed 10 cases of congenital [7–9,11,15–19], and 3 cases of infantile [13,20,21] malignant melanoma that were located on the scalp. Six of the 10 congenital scalp melanoma cases and one case of infantile melanoma arose from congenital melanocytic nevi, whereas remaining cases were de-novo in origin. Eight patients were male while five were female. Previous literature supports this male preponderance [13] and is consistent among different age groups [22].

3.2. Diagnosis

Diagnosis of malignant melanoma presents is a challenge. Biopsy is essential to establish a definitive diagnosis. The histological characteristics of melanoma include pagetoid intraepidermal spread, absence of maturation, increased cellularity and/or dermal mitotic figures [23]. No single histologic characteristic is diagnostic of melanoma. Rather it is the aggregation of histological features, which directs the diagnosis for or against melanoma [23]. Amongst the immunohistochemical markers of malignant melanoma, S-100 remains the most sensitive marker. However, the specificity of S-100 is limited [24]. More specific markers like HMB-45, Melan-A or tyrosinase are used in conjunction with S-100 [24]. Another typical finding of cutaneous melanomas is that the direct invasion of bone is exceedingly rare [25].

3.3. Prognosis and outcomes of congenital or infantile malignant melanoma of the scalp

Outcomes of malignant melanoma of scalp are generally unfavourable. Of the thirteen reported cases, 6 expired within 6 months of age [4,7,8,11], 1 was lost to follow-up and remaining were alive at least till the last follow-up [4,9].

It was believed that melanoma in children carry a better prognosis compared to adults, probably because of misdiagnosis of relatively benign conditions like spitz nevi as malignant melanomas [6,26]. However, it is now seen that prognosis depends on a number of factors. The origin of the disease is an important factor in determining prognosis. Transplacentally acquired melanomas fare the worst prognosis; in 3 out of 4 such reported cases, multi organ involvement (particularly of the liver) was noted, and death occurred within the first year of life [4,13]. Melanomas arising from a giant congenital melanocytic nevus tend to develop deep in the dermis or subcutaneous tissue; therefore, malignant melanoma are diagnosed at an advanced stage [4]. Melanomas arising de-novo appear to have a better prognosis and life expectancy than the former two; according to Asai et al. Two of the 12 patients with de-novo congenital and infantile melanoma expired in infancy [4]. Recent reports by Enam et al. and Sue et al. bring that count up to 4 [7,11].

According to Balch et al., tumor thickness and ulceration are the most important prognostic factors in patients with localized melanoma (stage 1 and 2) [27]. Ulceration (defined as absence of epidermis above the tumor) is a result of the primary melanoma invading the overlying epidermis instead of displacing it. Hence, an ulcerated melanoma has a greater metastatic capacity than its non-ulcerated counterpart of equal thickness [27]. The presence of tumor necrosis in a cutaneous melanoma is also an indicator of poor patient survival. The frequency of necrosis and its significance as a prognostic factor increases with tumor thickness [28]. Tumor necrosis was seen in5, and ulceration was seen in 6 cases of congenital and infantile malignant melanoma [7–9,11,17]; all patients with tumor necrosis had a poor prognosis.

The anatomic site of the primary melanoma also plays a role in determining the prognosis. Melanomas of scalp and neck had a particularly poor prognosis [27]. The most common site for congenital and infantile melanoma is also the head and neck region [9]. According to one study, scalp and neck melanomas account for about 6% of all melanomas but are responsible for 10% of all melanoma related deaths [29]. As compared to other anatomic locations, scalp melanomas are also more likely to metastasize to regional lymph nodes and be ulcerated [29]. The reason behind this worse prognosis of scalp and neck melanomas is not clear. A possibility could be that sentinel lymph node[1] (SLN) biopsy for the head and neck present with a number of challenges, which may influence the accuracy of the procedure [30]. These false negative SLN biopsies can lead to recurrences of melanoma. Nevertheless, such conclusions are difficult to be made for congenital melanoma due to the rarity of reported cases.

3.4. Treatment of congenital or infantile malignant melanoma of the scalp

Most of the congenital and infantile melanoma underwent

[1] Sentinel lymph node is defined as a node that receives direct independent drainage from the primary melanoma site.

surgical excision. Other modalities of treatment like interferon-α have been used more commonly in adults and children.

Role of SLN biopsies in congenital and infantile scalp melanomas is not well established. However, a recent study found it useful in the treatment of childhood melanoma [31]. The pathologic result of SLN biopsy in the most important prognostic factor for recurrence in cutaneous melanomas [32]. SLN biopsy is also useful in selecting patients who are most likely to benefit from complete lymph node dissection and other possible adjuvant therapy. The role of Interferon-α for congenital melanoma is limited because it can be associated with severe morbidity in neonates [6], even though it is currently the most widely used adjuvant treatment for melanoma in other age groups [32].

4. Conclusion

Congenital and infantile malignant melanoma of the scalp are rare, and difficult to diagnose without biopsy and histological analysis. These lesions carry poor prognosis. Bad prognostic factors include location, origin of disease, thickness, ulceration and/or necrosis of the lesion. Surgical excision remains the primary treatment modality.

Ethical approval

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Author contribution

1. Sohaib Tariq: Formulated the methodology, literature search and Write up.
2. Hussain Shallwani: Conceived the idea, literature search and Write up.
3. Muhammad Waqas: Review and Manuscript Modification.
4. Muhammad Ehsan Bari: Conceived the idea and finalised the draft.

Conflicts of interest

None.

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