Case Report

Giant Congenital Cerebriform Melanocytic Nevus of the Scalp in Adult

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Received: January 1, 2014
Accepted: February 25, 2014
Published Online: February 27, 2014
DOI: 10.5455/jihp.20140225013612

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Keywords: Congenital melanocytic nevus; melanocytes; giant; scalp lesion

INTRODUCTION

Congenital melanocytic nevi are defined as benign nevomelanocytic proliferation present at birth. They are rare lesions and a study of more than 500,000 neonates reported that 1/20000 neonate have nevi larger than 10 cm in diameter [1]. Majority of the congenital melanocytic nevi are not familial. They are usually sporadic caused by genetic mosaicism originating from a post zygotic mutation. The concept of paradigmatic inheritance best explains the sporadic occurrence of congenital melanocytic nevus and why exceptional cases of familial aggregation of this trait do not show any consistent Mendelian pattern. Review of literature shows 14 cases reports with family history of congenital nevi [2]. No clear sex predilection is reported for the development of melanocytic nevi. It is difficult to come to a conclusion based on available data of incidence and prevalence as women are more likely to seek medical attention [3]. They occur in approximately 1% of new-born and are usually classified according to their size. On the basis of their size, they are classified as “small congenital nevi” when the size is less than 1.5 cm in largest dimension and as “giant congenital melanocytic nevi (GCMN)” when the size is greater than 20 cm in largest dimension [4]. They have classical histologic criteria and are distinct from acquired nevi and constitute a well-known risk factor in the development of malignant melanoma both in prepubertal and post pubertal individuals with a reported incidence between 4.6% and 38% [5, 6]. Here we describe an adult female who lived with a huge cerebriform melanocytic nevi of the scalp for more than three decades.

CASE REPORT

A 32-year-old adult female presented with a huge cerebriform nevus on the scalp which had become painful with foul smelling discharge since the last 1-2 months. The lesion remained the same since birth. On examination nodular cerebriform lesion covering almost the entire scalp measuring about 25 cm in its longest axis with focal ulceration and yellowish discharge was seen (Figure 1). With a clinical diagnosis of nevus sebaceous, wide excision of the lesion was done with split skin graft. This case is unique due to the fact that this young female lived with this disfiguring lesion for 3 decades and never consulted any physician for cosmetic purpose and fortunately the lesion never turned malignant.
the dermis consisting of nevus cells arranged in groups and sheets infiltrating the lower dermis and subcutaneous tissue in close relation to the skin appendages and neurovascular bundles (Figures 3, 4, and 5). Grenz zone was seen and the most superficial cells in the group contained a lot of melanin. There was no pleomorphism, hyperchromatism or mitoses suggestive of malignancy. Melanin bleaching was done and intensity of staining reduced after bleaching.
DISCUSSION

GMCN commonly present as blue black patches over the back and thigh and also on the head and neck [7]. They are cosmetically distressing to the patients as well as parents. They are pigmented cutaneous lesion formed by a combination of epidermal and dermal derived nevus cells and are termed giant when the size exceeds 20 cm. In our case diagnosis was based on the following findings: 1) Presence of nevomelanocytes with in the lower two-thirds of the dermis and subcutaneous tissue 2) nevomelanocytes splaying and extending between the collagen vascular bundles of the reticular dermis in Indian files and cords 3) extension of nevomelanocytes around and extending between the hair follicles, sebaceous glands, eccrine apparatus, vessel walls and nerves 4) perivascular and perifollicular distribution of nevomelanocytes stimulating an inflammatory reaction such as figurate erythema 5) Arrector pili that may be enlarged, distorted and infiltrated by nevomelanocytes [1]. Melanin bleaching done further substantiated the diagnosis.

Approximately 1% new-born have biopsy proven nevomelanocytic nevus [8]. Lifetime risk of melanoma is of the range 5-40% [1]. Size, location, and histologic type seem to be the important factors which help to assess the risk of developing melanoma in congenital nevus. The larger the lesion, the higher the risk. The most common sites for malignant degeneration in giant nevi are the posterior scalp, neck, back, and buttock; 80% of malignant lesions occur in these areas. There is a lower risk of malignancy in giant nevi of the face and extremities, but the reason for this differential risk is not known. The risk is greater in the first decade of life and hence early intervention is recommended [9]. Poorly differentiated small round cell cancer, malignant cellular blue nevus and spindle cell malignant tumor with lamellar differentiation are other tumors described to arise from GMCN [1]. Though melanoma tends to arise in a proliferating nodule, literature shows that it has remained benign even after 30 years [10]. The verrucous and lobulated surface of GCMN as well as dermal origin of associated melanomas makes early detection difficult. Classifying a lesion as benign or malignant is often a source of diagnostic dilemma for pathologists. In our case ulceration of the nodule alerted the physician to rule out malignancy. A study of 80 pediatric patients with GMCN by Maldonado et al revealed malignant transformation in 4 of them of which 3 were fatal [11]. Nicotina et al described three cases of GMCN of the scalp which underwent excision and repair [12]. This case appears quite mysterious due to the fact that this young female lived with this disfiguring lesion for 3 decades and never consulted any physician for cosmetic purpose and fortunately the lesion never turned malignant.

Associated anomalies reported with GMCN are scoliosis, spina bifida, atrophy, asymmetry, clubfoot, elephantiasis, and cranial bone hypertrophy and neurocutaneous melanosis. The main aim of the treatment is to eliminate majority of the nevus cells by ablative approaches to improve the appearance and to reduce the risk of malignancy [7]. Management options are staged incision with grafting, dermabrasion, curettage, Q-stimulated ruby laser. Small lesions may require only simple close observation [1].

CONFLICTS OF INTEREST

Authors declare that there are no conflicts of interest.

REFERENCES


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