

Case Report

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Subcutaneous Malignant Melanoma developing in Congenital Giant Naevus with Complete Spontaneous Regression of Superficial Dermo-Epidermal Part: A Case Report

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Abstract

Giant congenital melanocytic nevi ≥ 20 cm is rare in adolescents and adults. Here we present a case of subcutaneous melanoma developing in congenital giant naevus with complete regression of superficial dermo-epidermal part. A 17 yr old girl presented with a painless and gradually increasing swelling in her right temporal region on pre-existing giant congenital melanocytic naevus (GCMN) on right hemiface. FNAC proved the case as malignant melanoma of temporal region with metastatic deposit in cervical lymph node. On histopathological examination it was found that the melanoma arising deep in the subcutaneous tissue with extensive involvement of deeper structures, extensive metastasis of cervical lymph nodes, salivary glands, deeper soft tissues with spontaneous regression of superficial epidermal and dermal part. Though regression is seen in melanomas, spontaneous regression has been mostly seen in melanomas with only intraepidermal component.

Keywords: Giant congenital melanocytic nevus; Subcutaneous melanoma; Spontaneous regression

Introduction

Giant congenital melanocytic nevus is a variant of congenital naevus characterised by its extensive size, by definition the surface area being 144 cm² or larger. They are rare, with approximate incidence of 1 in 20,000 live births. They increase the lifetime risk for malignant melanoma and neurological deficits, including leptomeningeal melanosis and epilepsy [1]. Congenital giant naevi extend into deeper tissue layers with involvement of neurovascular and deep striated muscle bundles. Conventional melanoma usually starts in the epidermis (Clarks' level I) and invades the papillary dermis (level II) to the junction of the papillary and reticular dermis (level III) and reticular dermis (level IV) before entering the subcutaneous fat (level V). In contrast melanomas arising in GCMN often extend into deeper tissues. Here we present a case of aggressive subcutaneous malignant melanoma in a giant congenital naevus with spontaneous regression of superficial part without any treatment. Though regression is seen in melanomas, spontaneous regression has been mostly seen in melanomas with only intraepidermal component [2].

Case Report

A 17 year old girl presented with a swelling in her right temple for 6 months. She had a congenital giant naevus on the right side of her face (Figure 1). The swelling was painless and had gradually increased in size. On examination, a firm lump 4 cm x 5 cm in size was present in her right temple in the area of the naevus. The lump was well defined. The skin above the lump was smooth and free from it. The lump was partly fixed to the underlying structures. A few neck nodes in level I, II and V were present. They were about 1 cm in size, discrete and firm in consistency.

FNAC from the lump revealed malignant melanoma. FNAC from the neck nodes also revealed metastatic deposits of melanoma. The patient underwent surgical excision with modified radical neck dissection. During operation the tumour was found to be free from the overlying skin. However the mass was firmly fixed to the underlying structures and complete excision was difficult (Figure 2a, 2b).

Histopathological examination revealed relatively circumscribed

expansile lobulated cellular tumour with invading margin infiltrating soft tissue and muscle (Figure 3). Tumour was composed of spindle to polygonal pleomorphic cells arranged in fascicles, nests and sheets. The individual cells had pleomorphic nuclei, clumped chromatin and prominent nucleoli and melanin pigments in cytoplasm (Figure 4a, 4b). Foci of necrosis, perineural invasion, angioinvasion, brisk mitosis, melanin incontinence was noted. Peripheral and deep margins were focally involved. The tumour was found to be unrelated to overlying



Figure 1: Patient with giant congenital melanocytic naevus in right hemiface. The melanoma appears as a swelling in her right temporal region.

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Figure 2a: After excision of the primary tumour.



Figure 2b: The lymph nodes in the neck shows black pigmentation.

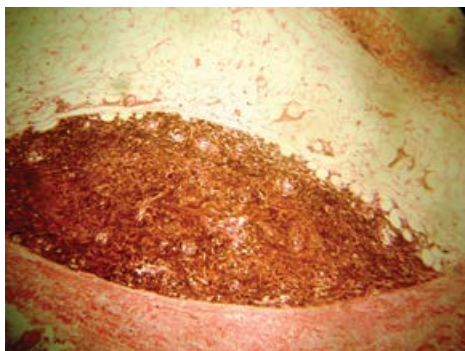


Figure 3: Local soft tissue infiltration.

skin on both gross and microscopic examinations and appeared to be originating beneath subcutaneum (Figure 5a, 5b). Sections from the skin showed unremarkable epidermis. Dermis showed elongated scattered dendritic melanocytes, fibrosis and clusters of melanophages throughout the entire thickness extending upto dermo subcutaneous junction. No naevus cells or melanoma cells were identified in epidermis and dermis. Sections from the neck dissection showed extensive metastatic deposits in multiple lymph nodes with extranodal spread, involvement of salivary gland parenchyma and soft tissue (Figure 6). The tumour cells were immunoreactive for HMB-45 and S-100. Extent of invasion- $pT_{4a}N_3M_x$

Discussion

By definition, a giant congenital naevus is ≥ 6 cm in size in children and ≥ 20 cm in size in adolescents and adults. Its incidence is 1 in

20,000 live births [1,3,4]. These lesions are thought to be pre malignant and warrant early complete excision if possible. The risk of malignant transformation of all congenital naevi ranges from 0.05% to 10.7% with risk correlating with size and location [5-8]. Melanoma have been found more commonly in patients with garment naevus than giant naevus on limbs and/or head which accounts only 0.3% [5,9]. Patients with CMN carry an approximately 465-fold increased relative risk of developing melanoma during childhood and adolescence. Men with garment naevi are at higher risk of developing malignant melanoma than women, with a male:female ratio of 2:1 [9].

Conventional melanomas that are not associated with GCMN

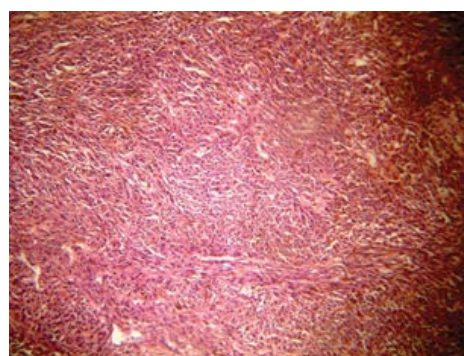


Figure 4a: Tumour consists of pleomorphic spindle to polygonal cells with cytoplasmic pigmentation. (10X).

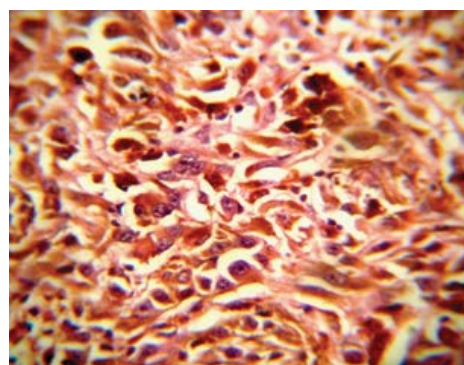


Figure 4b: Individual pleomorphic spindle to polygonal spindle cells with prominent nucleoli. (40X).

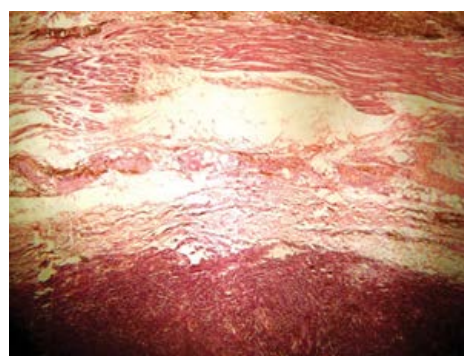


Figure 5a: Tumour arising from subcutaneous tissue. (4X).

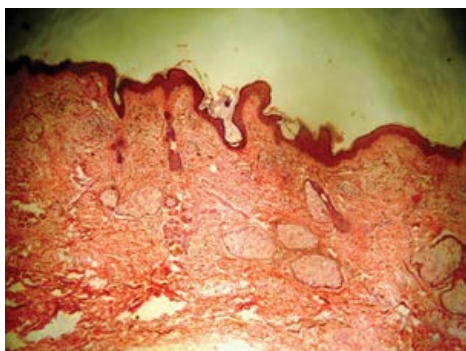


Figure 5b: The overlying skin is free.

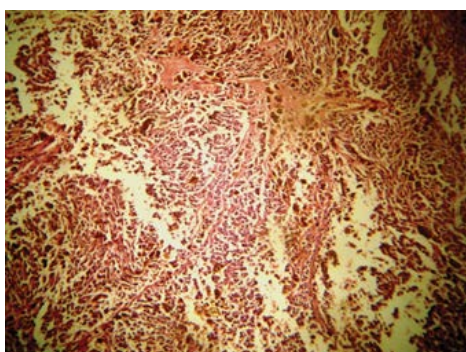


Figure 6: Lymph nodal structures are totally effaced by pleomorphic spindle tumour cells and dense pigmentation. (10X).

usually start with a lesion in the epidermal or superficial dermal layer and spread either radially or vertically [10]. In GCMN specimens, naevus cells are found to extend into deep layers of the dermis, sometimes around the lymphatic channels and vasculature, and even into the fascia or muscle [6]. It is very possible that benign naevocytes may spread via haematogenous or lymphatic routes [11].

Though regression has been described in melanomas, not many cases have been reported about spontaneous regression without any treatment. Clinically spontaneous regression is heralded by sudden onset of irregular halo around the tumour. Microscopically regression is characterised by a dense infiltrate of lymphocytes in early stage with vascular scar tissue and variable number of melanin laden macrophages in dermis in late stage.

Our case is unique in the following aspects: First, in our case the teen age girl with giant congenital hemifacial melanocytic naevus presented with subcutaneous aggressive malignant melanoma with extensive involvement of neurovascular, deep muscles, cervical lymph nodal, salivary gland and extranodal soft tissue. And second, the melanoma showed features of spontaneous regression of dermo epidermal part

of primary tumour without any treatment. The combination of these feature have, to our knowledge, not been reported in the literature.

Conclusion

GCMN are rare and melanomas developing in GCMN with regression of superficial dermo epidermal part in female adolescent is an unique case.

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