Case Report

Pediatric Melanoma Arising in a Background of Melanocytic Nevi

Abstract

Malignant melanoma is an aggressive neoplasm primarily involving the skin. They may arise de novo or from a premalignant melanocytic lesion. Melanomas are primarily known to occur in adults. Pediatric melanomas (PM) are rare and predominantly occur de novo following ultraviolet deoxyribonucleic acid (DNA) damage. They may also be associated with the presence of congenital melanocytic nevi. We report a rare case of a 6-year-old child with multiple melanocytic nevi subsequently diagnosed with melanoma of the back and metastatic disease.

Keywords: *Child, malignant melanoma, skin neoplasm*

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Introduction

Malignant melanoma is an aggressive malignancy, usually involving the skin. It is commonly associated with sun exposure, and its incidence increases with age.[1] It has a relatively lower incidence in pediatric populations: <1.8/1,000,000 in children younger than 10 years and 2.4/1,000,000 in children aged 10 to 14 years.[2] The incidence of melanoma in India is <0.5/1,000,000.[3] However, only a few case reports of melanoma in children have been reported from India.[4] The present case is an example of this rare condition and illustrates the severity of PM in a patient with multiple melanocytic nevi.

Case Report

A 6-year-old female child presented with pigmented, proliferative growth measuring 2.5 × 1.5 × 1 cm on the back for 3 months. On examination, numerous hairy nevi (>30) ranging in size from 0.5 cm to 7 cm were noted over the trunk and neck, which had been present since birth [Figure 1]. There was no family history of similar lesions. Axillary nodes were palpable on examination.

Contrast-enhanced computed tomography (CECT) of the thorax and abdomen showed multiple enlarged heterogeneously enhancing nodes in the left axillary region. Gross ascites, peritoneal deposits, omental caking, and extensive bony lesions were also noted. Radiological findings were suggestive of metastatic disease [Figure 2].

Histopathological examination of the lesion showed an infiltrating neoplasm composed of pleomorphic cells with prominent nucleoli. Intracytoplasmic melanin pigments were noted [Figure 3]. A biopsy from the left axillary node showed infiltration by similar cells [Figure 4]. Immunohistochemistry (IHC) for HMB45 (Human Melanoma Black), SRY-related HMG-bOX transcription factor 10 (SOX 10), and Pan-cytokeratin (pan-CK) was performed, which confirmed a diagnosis of nodular melanoma with nodal metastasis.

In view of the disease stage (stage IV—T4bN2bM1c) and the patient's general condition, the family was counseled and palliative management was planned. Unfortunately, the patient expired within 3 months of diagnosis.

Discussion

Melanomas are malignant tumors arising from melanocytes, most commonly in the extremities and anorectum.[5] Childhood melanomas are rare, and risk factors include age, family history, genetic susceptibility, and congenital nevi.[1,6] In addition, total body nevus count is an important risk factor, the risk being 15 times higher in individuals with more than 10 nevi, greater than 5 mm in diameter.[1] Melanomas are

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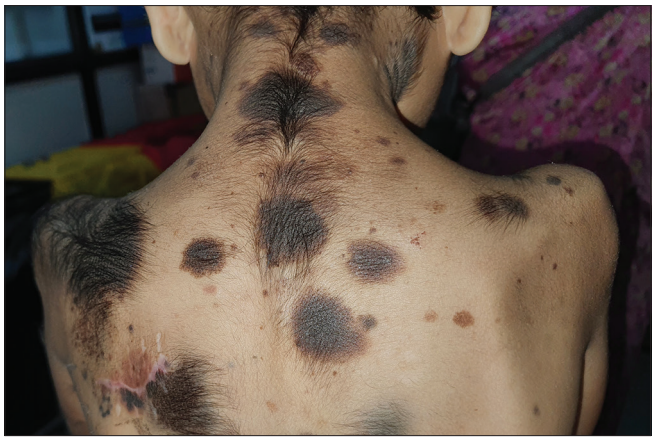

Figure 1: Multiple hairy nevi over the back

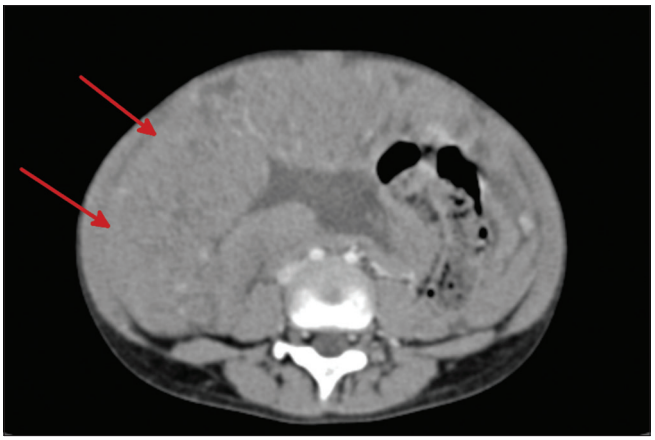

Figure 2: Peritoneal deposits (red arrows) with omental thickening (CECT abdomen)

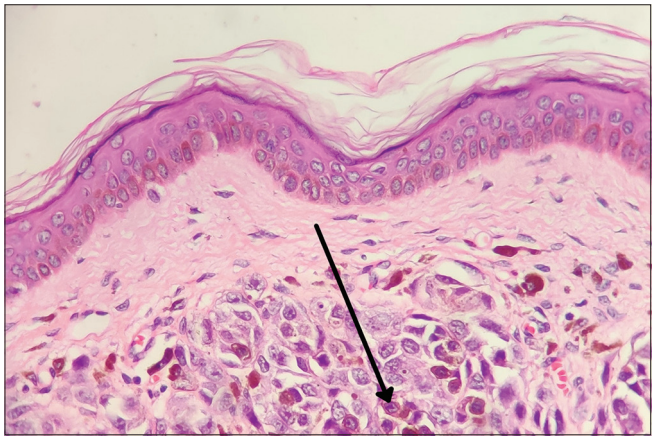

Figure 3: Biopsy of the lesion over the back. Dermis shows diffuse infiltration by pleomorphic cells with prominent nucleoli and cytoplasmic melanin (black arrow) (H and E 400×)

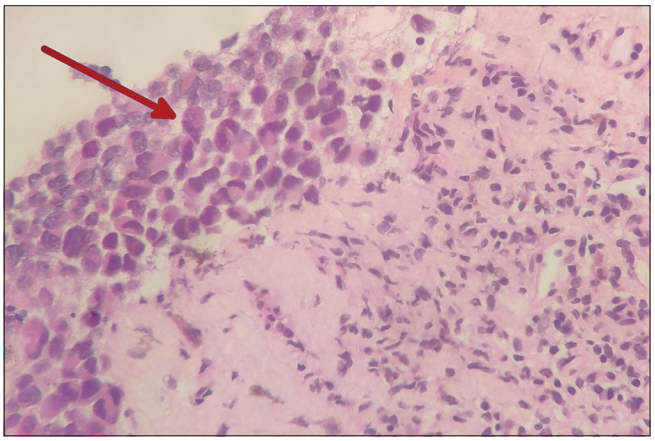

Figure 4: Tumor cells (red arrow) with adjacent lymphoid cells in the lymph node (H and E 400×)

traditionally present with lesions showing asymmetry, border irregularity, color variability, diameter greater than 6 mm, and evolution (ABCDE criteria). PM often presents atypically with amelanosis, bleeding, uniform color, variable diameter, and de novo development.[1] This divergent presentation highlights a possible issue in the early detection and diagnosis of melanomas in the pediatric population, in addition to its low incidence in the first place. Dermoscopy is a more sensitive clinical tool than the ABCDE criteria. A modified pediatric two-step algorithm aids in diagnosis; melanomas show structureless areas, shiny white lines, and atypical vessels, whereas congenital nevi commonly show patterns of dots and globules and reticular networks.[7]

Previous studies have found a female preponderance in PM.[6,8] A higher frequency of melanoma has been noted in older children, with younger children found to have worse overall survival.[8] However, age does not seem to have an independent effect on survival rate.[9]

Additionally, disease-free survival is significantly influenced by the site of origin, ulceration, tumor (T) status, node (N) status, and stage.[9]

According to the National Comprehensive Cancer Network (NCCN), surgery is the treatment of choice for melanomas.[1] The extent of surgery, however, is based on the stage of the disease, ranging from solely a wide local excision (WLE) (stage I) to WLE with complete lymph node dissection (stage III). In cases with distant metastasis (stage IV), treatment may include resection of both the primary tumor and metastasis, after considering the location and progression of the disease.[1] Options for adjuvant chemotherapy in pediatric metastatic melanomas are limited. Interferon alpha-2b (IFNα-2b) is used as an adjuvant in adults following resection. Some studies now suggest that IFNα-2b may be feasible for use in PM as well.[1,6] Ipilimumab, a monoclonal antibody that activates the immune system by binding to cytotoxic T-lymphocyte-associated antigen 4 (CTLA4), has been in use in adult metastatic melanomas for years.[1] Recently, it has been approved for use in the treatment of unresectable or metastatic melanoma in the pediatric population as well.[6] The role of prophylactic excision in congenital nevi has been suggested for suspicious lesions. However, no evidence-based surgical guidelines exist.[10] A

case-to-case approach is therefore the consensus approach at present.[10]

Follow-up of PM typically includes physical examination and lymph node surveillance.[6] CT and positron emission tomography (PET) scans, usually performed for follow-up in adult melanomas, are not pursued as aggressively in children due to the possible risk of cumulative radiation exposure and its adverse effects.[6]

Conclusion

The present case is an example of this rare condition and illustrates the severity and widespread dissemination of PM in a patient with possibly initially benign melanocytic nevi. Melanomas, though rare, can occur in childhood. However, given its rarity, it is important for healthcare providers to be aware of such an entity, to facilitate a differential diagnosis of this disease. Clinical examination, careful histopathological examination, and use of IHC allow for a conclusive diagnosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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