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CASE REPORT: PIGMENTED EPITHELIOID MELANOCYTOMA - A RARE CASE REPORTED FROM CAPFS' BSF COMPOSITE HOSPITAL, KADAMTALA.

Pathology

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ABSTRACT

Pigmented Epithelioid Melanocytoma (PEM) is a recently proposed entity which includes melanocytic tumors previously designated as 'animal type melanoma' and epithelioid blue nevus of Carney's complex. We report a case of PEM in an 8 year old male child who presented with a blackish nodule on the scalp. Excision biopsy of nodule was subjected to histopathological processing and IHC studies. The characteristic histomorphology and positive melanocytic marker in spindle, polygonal and epithelioid cellular components of the skin confirmed the diagnosis.

KEYWORDS

Pigmented epithelioid melanocytoma, Animal type melanoma, Epithelioid blue nevus

INTRODUCTION

Pigmented Epithelioid Melanocytoma (PEM), a heavily pigmented, rare, low grade melanocytic tumor has predilection for children, adolescents and young adults with a wide age range with histological similarities to equine melanocytic disease. Reports suggest that these lesions frequently metastasize to lymph nodes. However, their clinical behavior appeared to be better than conventional melanomas. PEM can arise in the context of a Carney's complex, or as a sporadic case. In our case, it was a sporadic lesion.

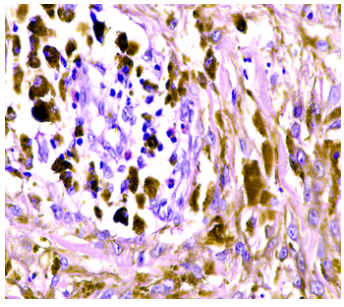
Case Report

An 8 year old male child was brought to the Central Armed Police Forces' Hospital, BSF (Border Security Force), Siliguri with blackish nodule on the scalp on back of head, for last three months. It slowly grew in size. He had difficulty in combing hair. There was no tenderness on the nodule. On examination, there was no palpable lymph node. There is no family history of such nodule. The nodule was resected and sent for histopathology examination. No lymph node was sent separately.

Gross Pathology

Received a dark nodule measuring approximately 1.5x1.0 cm. Cut section was solid and blackish in colour. Cut sections of the tissue was subjected to routine processing and stained with haematoxylin and eosin staining. IHC was done with S100 and HMB 45.

Figure 1: C/S of the swelling revealed solid blackish areas.



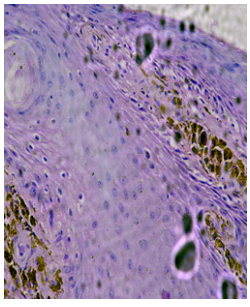


Fig 2 **Fig 3**

Figure 2 &3: 40x Views of H&E sections of skin lesion revealing heavily pigmented lesion in the upper dermis with spindle cells and epitheloid cells.

Histopathology

Sections from the submitted tissue revealed epidermis showing increased basal cell pigmentation. Upper papillary dermis revealed nests, sheets of spindle cells, polygonal and epithelioid cells with intense intracytoplasmic, brown pigmentation, having minimal nuclear pleomorphism, along with occasional mitoses.

Bleaching was positive, confirming that the brown pigment was melanin. Immunohistochemistry with S100 and HMB 45 was strongly and diffusely positive in all the cell types.

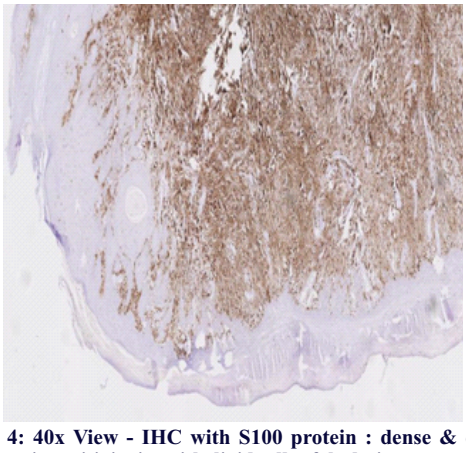


Figure 4: 40x View - IHC with S100 protein : dense & diffuse cytoplasmic positivity in epithelioid cells of the lesion.

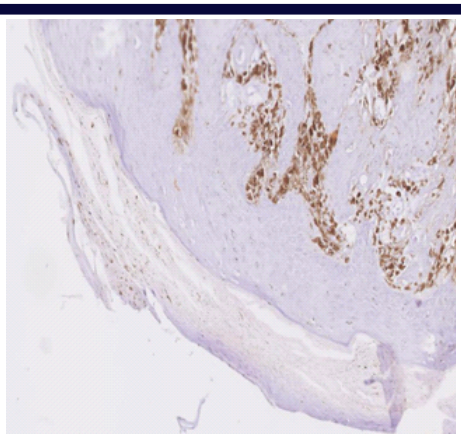


Figure 5: 40x View - IHC with HMB45 protein : diffuse cytoplasmic positivity in epithelioid cells of the lesion.

The presence of spindle, polygonal and epithelioid cells with intense intracytoplasmic melanin pigmentation, limited to the papillary dermis supported by intense S-100 protein positivity, diffuse HMB45 protein positivity suggested Pigmented epitheloid melanocytoma.

DISCUSSION

Pigmented Epithelioid Melanoma (PEM) is a recent terminology of a rare melanocytic neoplasm of low grade/intermediate/borderline neoplasm, occurring in all age groups. Equine or animal type melanoma (ATM) was first described in 1832 in old grey horses by Dick [(1)]. Although uncommon, metastasis to regional lymph nodes and as well as distant sites like spleen, liver, bone marrow and parotid gland have been reported [(2, 3)]. These tumors were compared with Epithelioid Blue Nevus (EBN) of Carney's complex. There association was first reported in 1996. However, they can occur sporadically also [(4)]. Till date three death cases have been reported [(5)].

In 2004, the term 'Pigmented Epithelioid Melanocytoma 'was coined by Zembowicz et al following clinicopathological study of 40 patients with tumors previously diagnosed as ATM and EBN [(6)].

PEM has a unique clinical and histological presentation. It is a slow growing lesion. It is reported in patients of wide range of age group but predominantly seen in children, adolescents and young adults. Most PEM occurs de novo, but rarely do they arise in association with a compound, dermal or congenital nevus. It is found that PEM has no relation with ethnic background neither exposure to sun is considered a factor for its pathogenesis [(6)]. PEMs express melanoma antigen recognised by T- cells (MART)-1, S-100 (figure 4) and (HMB)-45 (figure 5).

Although sentinel lymphnode metastases were found to be frequent (43%), there was no distant lymph node metastasis. The prognosis is better than conventional melanoma [(6)].

Clinical differential diagnosis often includes blue nevus, combined nevus or melanoma [(7,8)].

CONCLUSION

We present a case report of PEM in an 8 year old male child on the scalp without regional lymph node metastasis in a sporadic setting. The patient is kept under follow up for any regional lymph node metastasis. The PEM is a recently described, low/ intermediate/borderline melanoma which is heavily pigmented lesion showing positive melanocytic IHC markers having better prognosis than conventional melanoma with metastatic potential.

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