

Case Report

Pigmented epitheloid melanocytoma: a low grade/intermediate/ borderline melanoma

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ABSTRACT

Pigmented epitheloid melanocytoma is a recently described entity, low grade malignant lesion, encompassing epitheloid blue nevus of carney complex and animal type melanoma, from which they are histologically indistinguishable. Excision biopsy of skin and lymph node were subjected to histopathological processing and IHC studies. The characteristic histomorphology and positive melanocytic marker in spindle, polygonal and epitheloid cellular components of both the skin and left supraclavicular lymph nodal lesions confirmed the diagnosis. We report a rare case of pigmented epitheloid melanocytoma of the anterior chest wall with metastasis to left supraclavicular lymph node.

Keywords: Pigmented epitheloid melanocytoma, Low grade melanocytic tumor, Animal type melanoma, Epitheloid blue nevus

INTRODUCTION

Pigmented epitheloid melanocytoma (PEM) is a low grade melanocytic tumour which has predilection for children, adolescents and young adults with a wide age range. It is a rare, heavily pigmented melanocytic tumour with an indolent behaviour and histological similarities to equine melanocytic disease. Importantly, the results suggested that these lesions frequently metastasize to lymph nodes. Yet, their clinical behaviour appeared to be better than conventional melanomas. Therefore, they coined the term PEM to emphasize, the unique nature of this tumour and need to distinguish it from benign nevi and malignant melanoma. They proposed that PEM is a neoplasm which can arise in the context of a carney complex, or outside of the complex in a sporadic setting.

CASE REPORT

A 35 years old female presented with left supraclavicular swelling since 3 months with no other symptomatology. Past history and family history were nil significant. Routine investigations done, were found within normal limits. On examination a solitary discrete swelling measuring about 3.5 x 2 cm in the left supraclavicular region in between the sternal and the clavicular heads of the left sternocleidomastoid muscle, hard in consistency with no tenderness or any local rise of temperature was noted. FNAC smears studied from neck swelling, revealed sheets of large, round to polygonal cells with abundant pale eosinophilic cytoplasm, central to eccentric vesicular nuclei with prominent nucleoli, admixed with lymphocytes, against a haemorrhagic background. Occasional binucleate forms were noted. There was no

evidence of any necrosis, pleomorphism or mitosis. Possibilities of Metastatic Paraganglioma / Epitheloid tumour were suggested.

Patient was posted for surgery. On table, before the patient was anesthetized, a thorough examination revealed, an ulcerated and pigmented lesion measuring 1.75 x 1.5 cms on the anterior chest wall. No other skin lesions/ lymph nodes were noted. On further probing into the history, the patient revealed that, she did not realise that the chest wall lesion was of any significance. Anterior chest wall lesion was subjected to a wide local excision along with excision biopsy of left supraclavicular lymph node and both the specimens were submitted for histopathological examination.



Figure 1: Gross of skin lesion measuring 2.5x1.5x1cm, with a central area of ulceration, surrounded by pigmentation.



Figure 2: Gross of lymph node mass measuring 3x2x1cm.

Gross pathology

Received 2 biopsy bits, from chest wall and the neck lymphnode. The neck lesion was nodular, grey white

mass measuring 3 x 2 x 1 cm. Cut section was solid grey white with focal friable and necrotic areas. Received a skin covered elliptical, chest wall lesion measuring 2.5 x 1.5 x 1 cm with central ulceration, surrounded by hyperpigmentation. Cut section showed pigmentation of the epidermis with underlying fibrofatty tissue. Representative bits were taken, from both chest wall and neck lesions, subjected to routine processing and stained with haematoxylin and eosin staining.



Figure 3: C/S of the lymph node mass revealed solid grey white & necrotic areas.

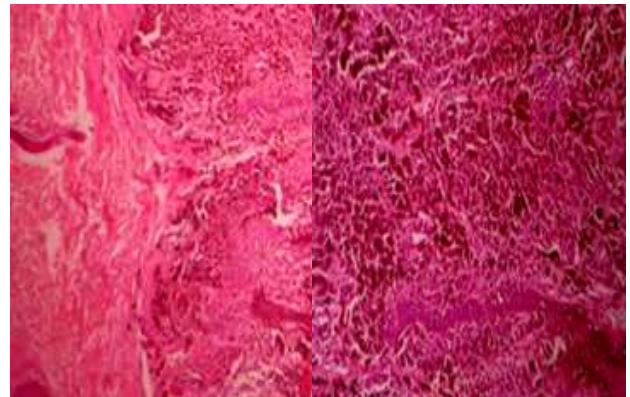


Figure 4 & 5: 10x & 40x Views of H&E sections of skin lesion revealing heavily pigmented lesion in the upper dermis with spindle cells and lobules of epitheloid cells.

Histopathology

Sections from the anterior chest wall lesion revealed structure of skin. Epidermis showed increased basal cell pigmentation. Upper papillary dermis revealed islands, nests, sheets of spindle cells, polygonal and epitheloid cells, exhibiting a confluent growth pattern, characterized by intense intracytoplasmic, brown pigmentation, having minimal nuclear pleomorphism, along with occasional mitoses.

Sections from left supraclavicular lymph node revealed remnants of native lymphnodal tissue at the periphery

with rest of the node being replaced by tumour tissue. The tumour is composed of clusters, vague lobules and sheets of epithelioid cells separated by sparse spindle cells. These epithelioid cells are having abundant vacuolated to pale eosinophilic cytoplasm with central vesicular nucleus and conspicuous nucleolus. Some of the cells are showing faint intracytoplasmic brown pigment. Areas of necrosis are seen with mild pleomorphism and occasional mitoses. The epithelioid cells seen in the dermis are having similar morphology as noted in the lymph node lesion.

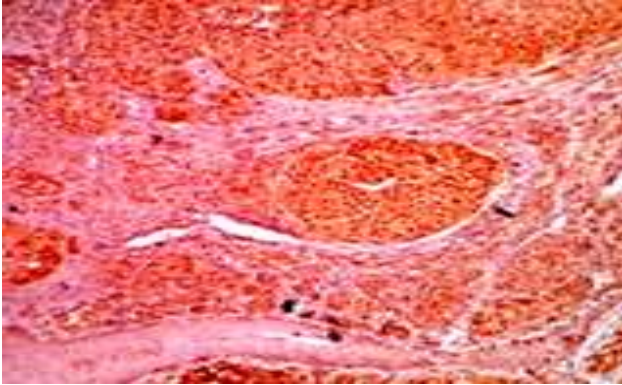


Figure 6: 40x View of IHC with S100 protein: dense & diffuse cytoplasmic positivity in both spindle & epithelioid cells of the anterior chest wall, skin lesion.

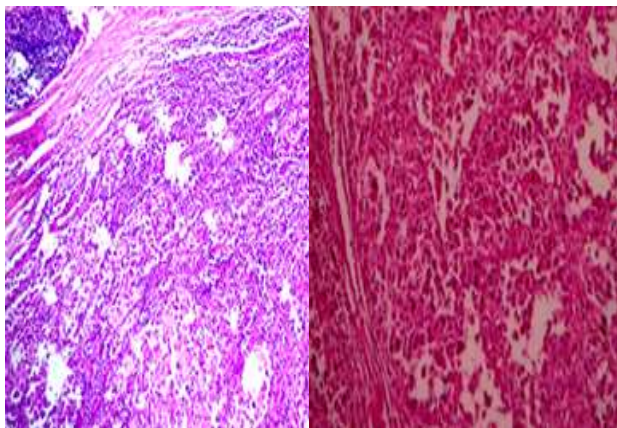


Figure 7 & 8: 10x & 40x Views of H&E sections of lymph node lesion revealed metastatic, predominantly epithelioid cell lesion, arranged in sheets, nests, lobules along with few spindle cells, having sparse & faint pigmentation. The tumour is replacing most of the native lymph node, leaving behind remnants of cortical lymphoid tissue at the periphery.

Bleaching was positive, confirming that the brown pigment was melanin and was substantiated by IHC. Immunohistochemistry with S100 was strongly and diffusely positive in all the cell types, of both chest wall and the lymph node. The possibility of paraganglioma was ruled out, as clinically pigmented skin lesions do not exist & S-100 protein positivity was noted exclusively in the sustentacular cells.

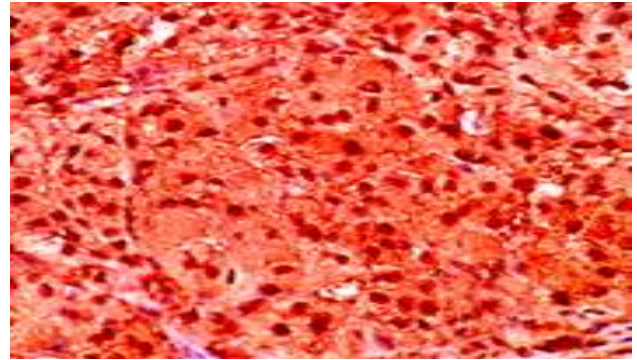


Figure 9: 40x View - IHC with S100 protein : dense & diffuse cytoplasmic positivity in epithelioid cells of the lymph node lesion.

Thorough evaluation of the history, clinical examination and special investigations, including ECG, CT- Scanning, ruled out the existence of other stigmata of carney complex. It is a well known fact that the morphologies of metastatic lesions may differ / vary from the primary neoplasm. Amelanotic variants of epithelioid blue nevus do exist.

Cells of spindle, polygonal and epithelioid morphology, intense intracytoplasmic melanin pigmentation, low grade morphology, growth limited to the papillary dermis, intense S-100 protein positivity and lack of other features of carney complex, suggested the diagnosis of low grade melanocytic tumour with the possibility of Pigmented epithelioid melanocytoma of Clark's level III (equivalent to stage III of breslow' thickness with lymphnode metastasis) limited to single node of 3 cm in diameter), in the sporadic setting. The metastatic deposit revealed predominance of one cell type, composed of epithelioid cells with sparse intracytoplasmic pigmentation.

DISCUSSION

Pigmented epithelioid melanocytoma (PEM) is a recently recognized rare melanocytic neoplasm of low grade/ intermediate/ borderline neoplasm, occurring in all age groups. Equine or animal type melanoma was first described in 1832 in old grey horses, consisting of nodules of heavily pigmented cells.¹ Although these neoplasms have an indolent course, they can rarely metastasize to the regional group of lymph nodes². Very rarely metastatic deposits have also been found in spleen, liver and bone marrow.³ These tumors were compared with original series of epithelioid blue nevus (EBN) from patients with the carney complex from which they were histologically indistinguishable.

In 2004, the term "Pigmented Epithelioid Melanocytoma" was coined by Zembowicz et al following clinicopathological study of 40 patients with tumours previously diagnosed as Atypical melanocytic tumor (ATM) and EBN.⁴ PEM is a distinct clinicopathological entity with unique clinical

presentation and histological features. It presents as slowly growing dermal with recent changes in a pigmented lesion or tumour with blue or blue/grey colouration. PEM occurs in patients over a broad age range but mainly encountered in children, adolescents and young adults. Most PEMs arise de novo, but occasionally they arise in association with a common compound, dermal or congenital nevus. It is reported that PEM has no ethnic predilection and suggested that exposure to sun is unlikely to be considered a major factor in the pathogenesis.

PEM is a unique low-grade variant of melanoma with frequent lymph node metastases but having indolent clinical course. Although sentinel lymph node metastases were found to be frequent (43%), there was no spread beyond regional lymph nodes. The prognosis is relatively good compared to conventional malignant melanoma. PEM is to be considered as a provisional histologic entity encompassing both animal-type melanoma and epithelioid blue nevus.⁵

These tumors were formed by deep dermal (mean Breslow's thickness 3.3 mm) proliferation of heavily pigmented epithelioid and/or spindle melanocytes.⁶ There are no histologic criteria to distinguish between metastasizing and non-metastasizing types of PEM. Ulceration was the only feature more common in PEM than epithelioid blue nevi of carney complex. Otherwise, they are histologically indistinguishable. Loss of expression of a carney complex gene, cyclic adenosine 3' 5' monophosphatase and dependent protein kinase regulatory sub unit 1 alpha was observed in the majority of PEM cases.⁷ From an immunohistochemical point of view PEMs express melanoma antigens recognised by T-cells MART-1, S-100 and human melanoma black (HMB)-45.

CONCLUSION

In summary, we present a case report of Pigmented epithelioid melanocytoma - A low grade/ intermediate/ borderline melanoma, presenting as anterior chest wall pigmented lesion with metastasis to left supraclavicular lymphnode, in a sporadic setting. The follow-up for a period of six months showed no evidence of loco regional recurrence/ distant metastasis. PEM is a recently described entity, heavily pigmented, low grade melanocytic tumour, having indolent clinical behavior,

histological similarities to epithelioid blue nevus of carney complex and animal type melanoma, from which they are histologically indistinguishable, positive for melanocytic IHC markers, with tendency to loco-regional lymph node metastasis, carrying better prognosis than conventional melanomas, occurring in the context of a carney complex/sporadic setting. Pathologists and surgeons should recognize and accept the new concept of Pigmented epithelioid melanocytoma (PEM).

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Ethical approval: Not required

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