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Case Report

Pigmented epithelioid melanocytoma - A case report

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Abstract

Pigmented epithelioid melanocytoma (PEM) is a borderline melanocytic neoplasms or a low-grade melanoma which is rare. It affects all age groups. The prognosis is relatively good compared to conventional malignant melanoma. We presented here a case of 3 years old Saudi girl presented with an asymptomatic brown-black lesion on her left forearm. At clinical examination, the lesion had a diameter of 0.6 cm, appeared round, exophytic, and intensely pigmented with well-defined edges and bluish reflections. All features strongly suggested nodular melanoma and the lesion was excised and histopathology showed asymmetrical growth of heavily pigmented atypical epithelioid and spindle cells. A pigmented epithelioid melanocytoma, so-called animal-type melanoma, was diagnosed. The depth of invasion was classified as Clark level IV, with maximum thickness of 3 mm.

Key words

Pigmented epithelioid melanocytoma, Animal-type melanoma, Malignant melanoma.

Introduction

Pigmented epithelioid melanocytoma (PEM) is an unusual variant of malignant melanoma with relatively indolent behavior. Dick first described it and noted its predilection for gray horses and named it as equine-type melanoma in 1832 [1]. In 1925, Darier delineated the parallelism between these lesions and similar neoplastic processes in humans, to which he gave the name melanotic sarcoma [2]. In 2004, Zembowicz, et al. [3] proposed the term pigmented epithelioid melanocytoma (PEM) for a group of lesions including cases previously diagnosed as human animal-type melanoma [4] and epithelioid blue nevus [5]. PEM was first described as epithelioid blue nevus in patients with Carney complex. Current experience indicates that PEM is best considered as a borderline melanocytic neoplasms or a low-

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grade melanoma. PEM is extremely rare. It affects all age groups. The prognosis is relatively good compared to conventional malignant melanoma.

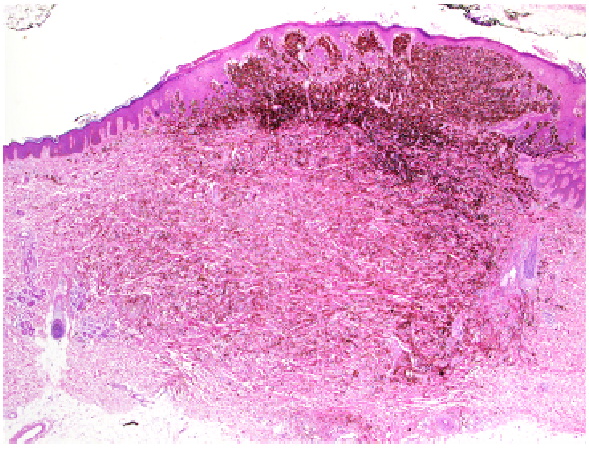
Case report

A 3 years old Saudi girl presented with an asymptomatic brown-black lesion on her left forearm. The nodule had been observed since birth and increasing in size in a few months. There was no past history or family history of malignant melanoma, blue nevus or Carney complex. At clinical examination, the lesion had a diameter of 0.6 cm, appeared round, exophytic, and intensely pigmented with well-defined edges and bluish reflections. All features strongly suggested nodular melanoma and the lesion was excised and histopathology showed asymmetrical growth of heavily pigmented atypical epithelioid and spindle cells. **[Photo - 1]** Junctional nests and few single melanocytes in epidermis were found. **[Photo - 2]** Deep dermal melanocytes showed pigmented melanocytes indicating loos of maturation. **[Photo - 3]** Mitosis, necrosis, ulceration, and lymphocytic infiltrate were absent. A pigmented epithelioid melanocytoma, so-called animal-type melanoma, was diagnosed. The depth of invasion was classified as Clark level IV, with maximum thickness of 3 mm.

Discussion

Zembowicz, et al. [3] recently proposed the term PEM for a spectrum of melanocytic tumors previously diagnosed as human animal-type melanoma [4] and epithelioid blue nevus [5]. This proposal was based on prospective analysis and follow up of 41 cases of lesions of suspected animal-type melanoma and their comparison to 11 examples of epithelioid blue nevus from the original series of Dr Carney. Zembowicz, et al. [3] concluded that both groups of lesions were either histologically indistinguishable or had considerable histological overlap. PEM is a distinctive clinic-pathological variant of melanocytic tumor of unknown malignant potential. It is characterized by its unique feature of indolent behavior compared to conventional melanoma. Histopathologically, these lesions often have a wedge-shaped configuration and are composed of heavily pigmented dermal melanocytic tumor cells with a mixture of epithelioid and spindled cells. Mitosis can be seen, but is rare. The histopathological differential diagnoses of PEM include cellular blue nevus, malignant blue nevus, Spitz nevus, deep penetrating nevus and epithelioid blue nevus. The most specific differentiating feature between PEM and cellular blue nevus is the presence of abundant epithelioid cells in PEM [6].

Photo – 1: Asymmetrical dermal growth of heavily pigmented melanocytes without maturation. (H & E, 40 X)



Zembowicz, et al. [3] reported that the lymph node metastasis was detected in 11 of the 24 cases (46%) in which lymph node sampling were performed. Because PEMs are associated with frequent sentinel lymph node metastases, we should recommend sentinel lymph node sampling in the management of these cases as a

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diagnostic procedure. The current follow up is still too short to make definitive statement about the long-term prognosis in PEM, but the experience thus far indicates that it has more favorable prognosis than conventional melanoma. However, PEM is not a benign tumor as they can cause the patient's death [4].

Photo – 2: High power Dermo-epidermal junction showed some atypical junctional melanocytes (indicated by arrows). (H & E, 400 X)

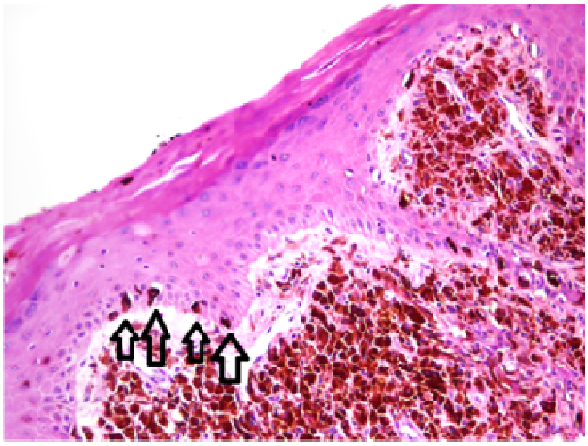
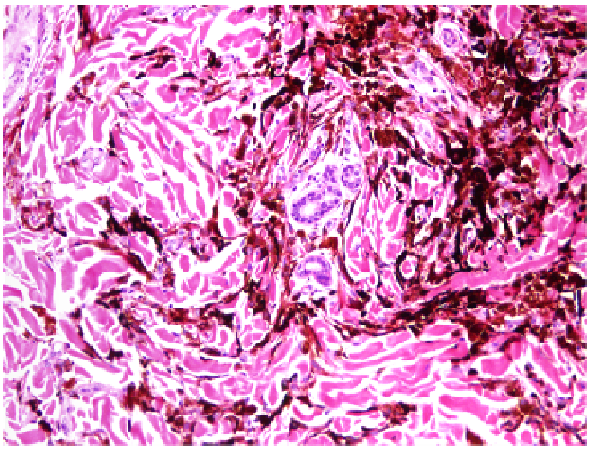


Photo – 3: Deep dermal melanocytes showed pigmented melanocytes indicating loos of maturation. (H & E, 400 X)



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