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Pigmented Epithelioid Melanocytoma: A Rare Melanocytic Neoplasm with Diagnostic Complexity

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Abstract:

Pigmented epithelioid melanocytoma (PEM) is a rare melanocytic tumor with borderline malignant potential. It exhibits histological features of both epithelioid blue nevus and animal-type melanoma. Due to its ambiguous biological behaviour, diagnosis and management present significant clinical challenges. Here, we present a case of a 32-year-old female with a solitary, pigmented lesion on her right forearm, which was diagnosed as pigmented epithelioid melanocytoma following histopathological and immunohistochemical examination. The lesion was excised surgically with clear margins, and the patient has shown no signs of recurrence or metastasis during a one-year follow-up. This report underscores the importance of accurate diagnosis and appropriate management of PEM to ensure favourable clinical outcomes.

Keywords: pigmented epithelioid melanocytoma, melanocytic tumor, pigmented lesion, melanoma, epithelioid blue nevus.

Introduction

Pigmented epithelioid melanocytoma (PEM) is a rare melanocytic neoplasm characterized by heavily pigmented epithelioid and spindle-shaped melanocytes. Initially described by Zembowicz et al. (2004), PEM encompasses lesions previously categorized as animal-type melanoma and epithelioid blue nevus (Zembowicz & Carney, 2004). While it often exhibits benign behaviour, cases with regional lymph node involvement and distant metastases have been reported, raising concerns about its malignant potential (Barnhill et al., 2018).

PEM predominantly affects young adults but can occur at any age. The lesions are typically solitary, well-circumscribed, and deeply pigmented, often located on the trunk or extremities (Reed et al., 2019). Histopathologically, PEM displays a combination of epithelioid and spindle cells with abundant melanin pigmentation. Immunohistochemical studies frequently demonstrate positivity for markers such as S100, HMB-45, and Melan-A (Busam et al., 2020). Given the rarity and ambiguous nature of PEM, accurate diagnosis is essential to differentiate it from malignant melanomas and other melanocytic lesions. This case report aims to contribute to the growing body of literature on PEM by detailing a case presentation, diagnostic approach, and clinical management.

Case Details

A 32-year-old female presented with a complaint of a darkly pigmented lesion on her right forearm, which had been present for approximately six months. The lesion had gradually increased in size but was asymptomatic, without associated pain or bleeding.

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Clinical Examination:

On physical examination, a solitary, well-defined, dome-shaped, dark brown to black lesion measuring 1.8 cm in diameter was observed on the volar aspect of the right forearm. The lesion was non-tender, non-ulcerated, and exhibited no signs of inflammation. Regional lymph nodes were not palpable.

Diagnostic Workup:

Given the clinical suspicion of a melanocytic lesion, a dermoscopic examination was performed, revealing a homogenous blue-black pigmentation with no evidence of regression structures. An excisional biopsy with a 2 mm margin was performed for histopathological evaluation.

Histopathological Findings:

Microscopic examination revealed a well-circumscribed dermal tumor composed of epithelioid and spindle-shaped melanocytes with abundant cytoplasmic melanin pigmentation. There was no evidence of significant nuclear atypia or mitotic activity. The lesion extended to the mid-dermis without infiltration into the subcutaneous tissue. (Figure a, b)

Immunohistochemical Analysis:

The tumor cells showed strong positivity for S100, HMB-45, and Melan-A, confirming the melanocytic origin of the lesion. Ki-67 proliferation index was low (<5%). These findings were consistent with a diagnosis of pigmented epithelioid melanocytoma. (Figure c, d)

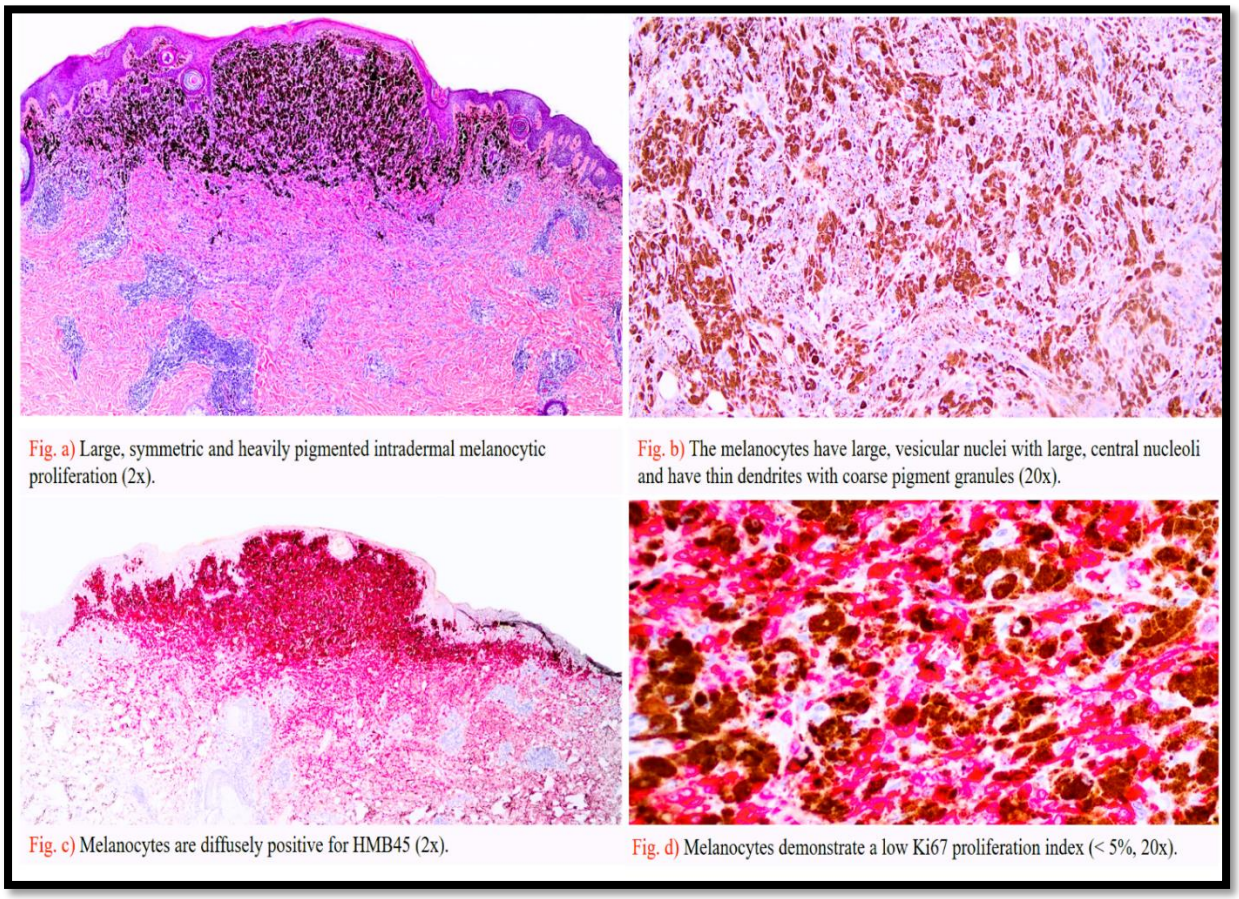

Fig. a) Large, symmetric and heavily pigmented intradermal melanocytic proliferation (2x).

Fig. b) The melanocytes have large, vesicular nuclei with large, central nucleoli and have thin dendrites with coarse pigment granules (20x).

Fig. c) Melanocytes are diffusely positive for HMB45 (2x).

Fig. d) Melanocytes demonstrate a low Ki67 proliferation index (< 5%, 20x).

Management:

Complete surgical excision with a 5 mm margin was performed. The surgical site healed without complications. Given the potential for regional lymph node involvement, a sentinel

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lymph node biopsy was considered but deferred due to the absence of high-risk histological features.

Follow-Up:

The patient was monitored clinically every three months. At the one-year follow-up, there was no evidence of local recurrence or distant metastasis.

Discussion

PEM is a rare melanocytic tumor with features overlapping benign and malignant melanocytic neoplasms. Its histopathological characteristics often pose diagnostic challenges, necessitating a comprehensive approach that includes clinical, histological, and immunohistochemical evaluations (Zembowicz & Carney, 2004).

The differential diagnosis for PEM includes blue nevus, animal-type melanoma, and conventional malignant melanoma. Blue nevi are typically smaller and lack the epithelioid cell component seen in PEM (Barnhill et al., 2018). Animal-type melanoma, though histologically similar, often exhibits more aggressive behaviour with a higher likelihood of metastasis (Reed et al., 2019).

Immunohistochemistry plays a crucial role in the diagnostic process. The strong expression of melanocytic markers such as S100, HMB-45, and Melan-A supports the diagnosis of PEM. The low Ki-67 proliferation index observed in this case is consistent with the typically indolent nature of the tumor (Busam et al., 2020).

Management strategies for PEM remain a subject of debate. Complete surgical excision with clear margins is generally recommended. The role of sentinel lymph node biopsy is controversial; it may be considered in cases with high-risk features, such as significant mitotic activity or deep invasion (Barnhill et al., 2018). In this case, the decision to forego sentinel lymph node biopsy was based on the absence of adverse histological features.

Long-term follow-up is essential given the potential for late recurrences and metastasis. Although our patient remained disease-free at one year, continued surveillance is warranted.

Conclusion

Pigmented epithelioid melanocytoma is a rare and diagnostically challenging melanocytic tumor with an unpredictable clinical course. Accurate diagnosis through histopathological and immunohistochemical evaluation is essential to guide appropriate management. Complete surgical excision with clear margins remains the cornerstone of treatment. This case highlights the importance of vigilant follow-up to monitor for recurrence or metastasis.

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