Dermatology Practical & Conceptual

Pigmented Epithelioid Melanocytoma: Avoiding Overtreatment in a Challenging Diagnosis

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Case Presentation

A 16-year-old male presented with a 3×3 mm pigmented lesion on the left upper arm, noted to have appeared approximately one year earlier and to have progressively darkened. Dermoscopic evaluation revealed homogeneous black-blue pigmentation without a pigment network. The lesion was completely excised. Histopathologic examination revealed a predominantly intradermal compound melanocytic proliferation. The junctional component was located within a hyperplastic epidermis and consisted of a few compressed nests of small epithelioid melanocytes. The dermal component was wedge-shaped and composed of numerous melanophages admixed with nests and single epithelioid melanocytes with amphophilic, variably pigmented cytoplasm and round nuclei with evenly distributed chromatin and visible nucleoli. Immunohistochemistry demonstrated loss of PRKAR1A expression. RNA extracted from FFPE tumor tissue was screened for fusion transcripts associated with a range of cancer types; no fusion events were identified involving ALK, BRAF, NTRK1, NTRK2, NTRK3, ROS1, or RET. The case was reviewed in a regional melanoma multidisciplinary team meeting, and a wider local excision with 5 mm margins was performed, with clear histological margins. Clinical follow-up was planned at six-month intervals for two years.

Teaching Point

Pigmented epithelioid melanocytoma (PEM) is a rare, borderline melanocytic neoplasm that typically shows indolent clinical behavior despite frequent lymph node involvement. Histologically, PEM is characterized by a heavily pigmented dermal melanocytic proliferation, often wedge-shaped, and composed of epithelioid and/or spindle-shaped melanocytes. Although sentinel lymph node positivity is observed in up to 43.8% of reported cases, progression beyond regional nodes is rare (2.9%), and mortality remains exceptionally low [1]. Loss of PRKAR1A expression, as seen in this case, supports the diagnosis and helps distinguish PEM from conventional melanoma and other pigmented neoplasms [2]. Awareness of PEM's distinct histopathologic and molecular profile is essential for accurate diagnosis and to prevent unnecessary aggressive treatment, especially in young patients.

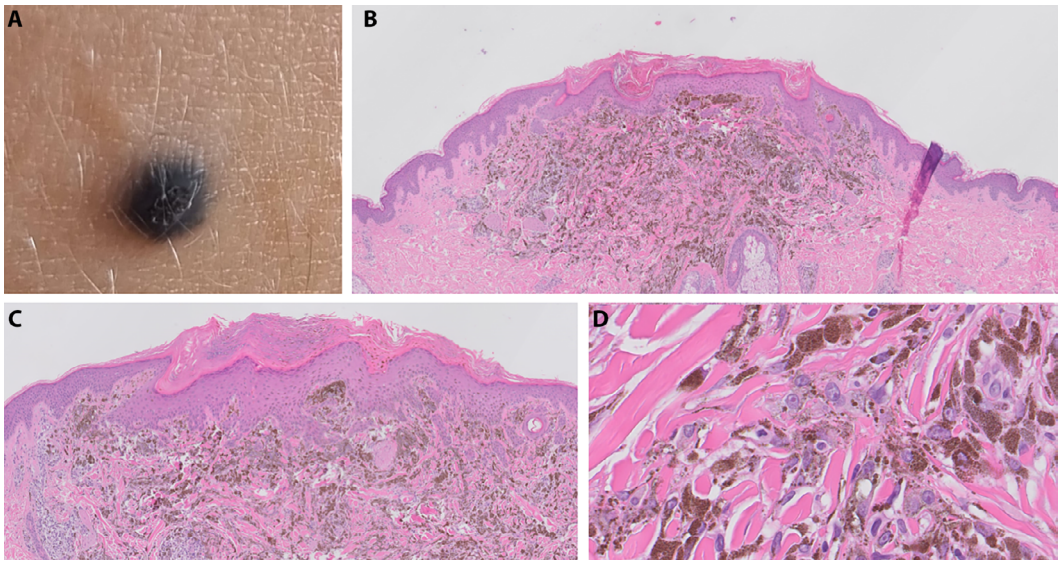

Figure 1. A: Clinical image showing a 3×3 mm well-circumscribed pigmented macule on the left upper arm of a 16-year-old male. B: Low-power histologic view demonstrating a predominantly intradermal, wedge-shaped melanocytic lesion with heavy pigmentation. C: Junctional component is minimal and located within a hyperplastic epidermis. D: Numerous pigmented melanophages admixed with single epithelioid melanocytes are seen within the dermis.

References

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