Conjunctival Pigmented Epithelioid Melanocytoma: A Clinicopathological Case Report

Pigmented epithelioid melanocytoma (PEM) is a rare, melanocytic skin and mucosal tumor with low-grade malignancy. It is a recently defined histopathological entity. It encompasses epithelioid blue nevus of the Carney complex, a familial lentiginosis and multiorgan neoplasia syndrome, and most tumors previously described as animal-type melanoma (ATM).1,2

Dick3 first described ATM in gray horses in 1832. The similarity between the equine and human skin variant was noticed later by Darier.4 In 2004, Zembo-wicz et al5 observed the same features in 41 ATM and 11 epithelioid blue nevus specimens and proposed the term of PEM. Because of its unique demographic characteristics, clinical presentation, histological features, and intermediate malignant potential between a benign blue nevus and common melanoma, PEM was allocated into a separate nosological category of borderline melanocytic tumors.6 We present, to our knowledge, the first clinicopathological case report of conjunctival PEM, initially diagnosed as ATM.

Report of a Case. A 47-year-old white man had a 38-year history of a slowly enlarging, darkly pigmented nodule on the superior palpebral conjunctiva of his right eye. Five smaller, pigmented spots adjacent to the lesion had recently appeared (Figure 1). Presuming a diagnosis of conjunctival melanoma with local satellite metastasis but without excluding a benign lesion, we performed an excisional biopsy. Histopathological analysis showed a proliferation of heavily pigmented fusiform and epithelioid melanocytes arranged in solid, confluent sheets and infiltrating the tarsal plate and orbicularis muscle but not the epithelium. Large epithelioid cells with prominent eosinophilic nuclei were also found. Nuclear pleomorphism was mild to moderate. Mitoses were rare (1/40 high-power fields) and no atypical mitoses were seen. There was no necrosis (Figure 2). The tumor cells expressed S-100 and Melan-A proteins and reacted with HMB-45 antibody. Results on GNAQ and GNA11 genetic analysis of exon 5, searching for mutations identified in 83% of blue nevi,8 were negative. At that time, a diagnosis of ATM was made.

A general checkup showed unremarkable findings without regional lymphadenopathy. To increase safety margins, we performed an uneventful re-excision using an Abbé-Mustardé lower eyelid flap to reconstruct the upper eyelid. Seven years later, the patient displayed no metastasis or local recurrence.

Comment. We describe a patient with conjunctival PEM. Pigmented epithelioid melanocytoma is a distinct, low-grade variant of skin and mucosal melanoma. It involves all age groups, with a preponderance in young adults. There is no predilection for sex or body localization, suggesting that sun exposure is unlikely to be a major factor in its pathogenesis.1

Clinically, PEM appears as a thick, darkly pigmented nodule or plaque. Histopathological features consist of a proliferation of spindle, epithelioid, and large epithelioid melanocytes—abundant with melanin—arranged in sheets and/or nests localized in the dermis, from where they may infiltrate the subcutaneous fat or, rarely, the epidermis. Cells show occasional atypia. Positivity for immunohistochemical markers is the same as in melanoma.

Pigmented epithelioid melanocytoma behaves less aggressively than conventional melanoma. Although sentinel lymph node metastases as high as 46% have been documented, liver metastasis occurred in only 1 case, with the patient being well 2 years after resection. One lethal case has been reported.2,3 It is not clear whether, after excisional biopsy, re-excision with margins as wide as those for classic melanoma is indicated.2

In conclusion, we describe the first patient, to our knowledge, with conjunctival PEM. It is important to differentiate PEM from both a benign blue nevus and classic conjunctival melanoma, as PEM, usually
manifesting in younger patients, has a specific, low-grade malignant potential requiring adapted management.

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