Pathologic Quiz Case

A 69-Year-Old Man With a Brown-Black Facial Papule

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A 69-year-old white man with severely sun-damaged skin presented with a cutaneous, brown-black, firm papule measuring 0.5 cm on his right cheek. The lesion was excised completely. Histology revealed a well-circumscribed pigmented tumor in the superficial dermis (Figure 1, A). The tumor contained a biphasic cellular population, including epithelial and melanocytic elements (Figure 1, B). The epithelial cells were variably sized and contained moderate to abundant amphophilic cytoplasm that abruptly turned into nucleus-free ''shadow'' cells (Figure 1, C). The epithelial cells had round to oval, variably sized nuclei with prominent nucleoli. Scattered mitotic figures, including atypical forms, were identified (Figure 1, D). Admixed with the epithelial cells were dendritic melanocytes with intense melanin pigmentation. Large aggregates of melanin pigment were present both in the melanocytic and epithelial cells, as well as in numerous interspersed melanophages (Figure 1, C). The tumor nodule was surrounded by a complete fibrous capsule of concentric collagen with abundant basophilic ground substance. In between the epidermis and the fibrous capsule, there was prominent telangiectasia (Figure 1, B). No granulomatous reaction or calcification was evident. The dermis surrounding the lesion contained marked solar elastosis. The epidermis overlying the dermal nodule was flattened and hyperkeratotic and had peripheral acanthosis, forming a symmetrical collarette (Figure 1, A). No local recurrence was noted at the site of the original biopsy after a follow-up of 9 months.

What is your diagnosis?

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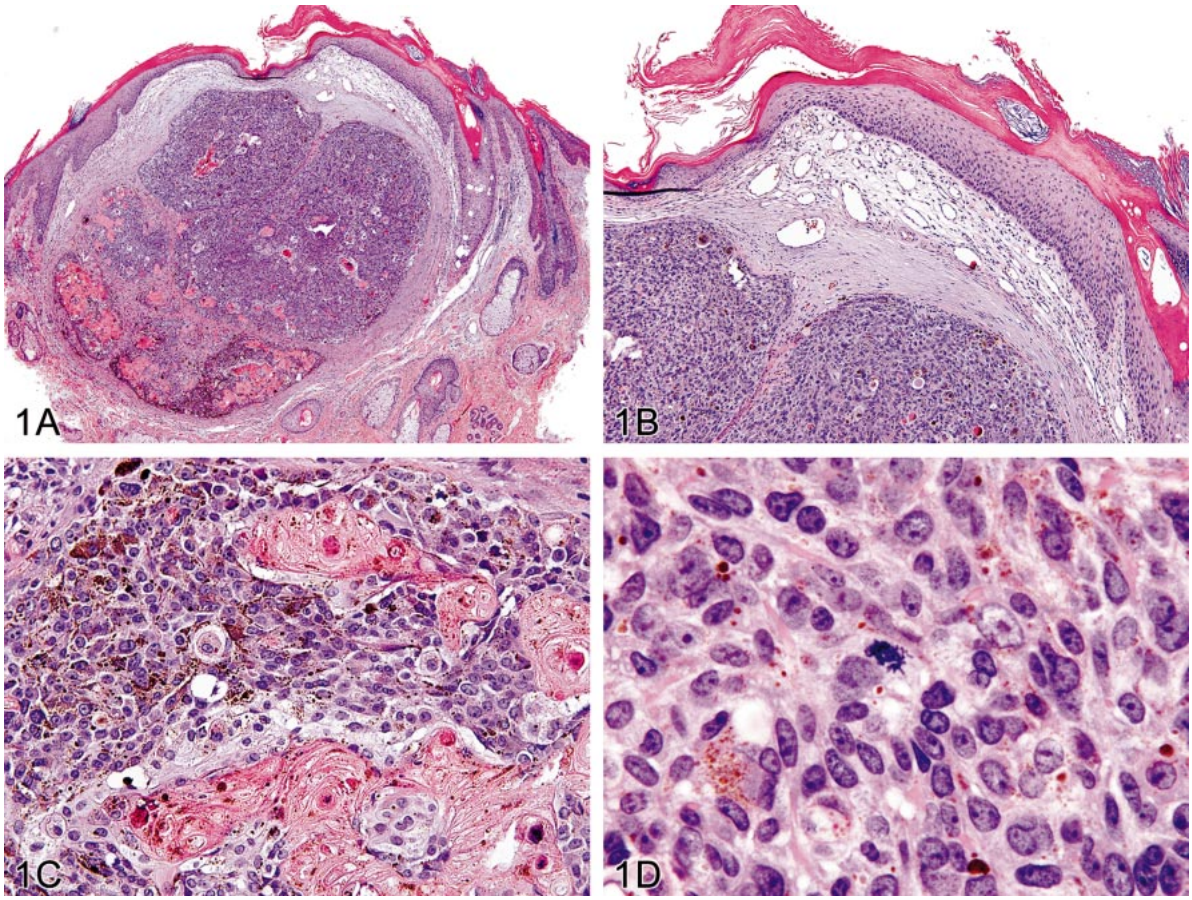

Pathologic Diagnosis: Melanocytic Matricoma

Immunohistochemical studies were performed with aminoethylcarbazol chromogen to give a red reaction product. Low-molecular-weight cytokeratin (CAM 5.2) labeled scattered epithelial cells. High-molecular-weight cytokeratin (34βE12) labeled the keratinized shadow cells. MART-1 and HMB-45 highlighted the morphology of the dendritic melanocytes, which were admixed with the epithelial cells, surrounding them with their processes (Figure 2, A and B, respectively). The microscopic and immunohistochemical features are diagnostic of melanocytic matricoma.

Melanocytic matricoma was first recognized in 1999 by Carlson et al,[1] who reported 2 cases of a pigmented matrical neoplasm distinct from pilomatricoma, composed of matrical cells and dendritic melanocytes. Subsequently, 2 additional case reports have confirmed the uniqueness of the entity.[2,3] Including the currently reported case, to our knowledge only 5 cases of melanocytic matricoma have been reported to date. Clinically, all cases have presented as dark papules arising on the sun-damaged skin of elderly patients. The clinical differential diagnoses considered usually include pigmented basal cell carcinoma, malignant melanoma, and hemangioma. Histologically, the appearance of all reported cases is quite similar and characterized by a dual cell population, including admixed epithelial and melanocytic cells. The epithelial component contains matrical, supramatrical, and shadow or ''ghost'' cells. The melanocytic cells are dendritic and darkly pigmented. The histologic differential diagnosis of melanocytic matricoma includes the pigmented variants of pilomatricoma, matrical carcinoma, basal cell carcinoma, and malignant melanoma.[1,3] Although the characteristics of melanocytic matricoma, such as circumscription, small size, and clinical history, most likely suggest a benign nature, long-term follow-up is not yet available to exclude aggressive behavior.

Recently, 2 cases of matrical carcinoma with prominent melanocytic hyperplasia were described. These tumors are, however, very different from melanocytic matricoma. They are composed of ill-defined, multinodular masses of pleomorphic, mitotically active cells with areas of necrosis. Importantly, the tumors have deep infiltrative areas and locally aggressive behavior. These lesions may represent the malignant counterpart of melanocytic matricoma.[4]

Melanocytic matricoma is morphologically different from so-called matricoma[5] or proliferating pilomatricoma, despite some debate on this issue.[6–9] Matricoma is described as having a silhouette composed of many small, discrete aggregations positioned throughout the dermis and sometimes just into the subcutaneous fat. In contrast, melanocytic matricoma is characteristically a single superficial dermal nodule. Importantly, melanocytic matricoma has a characteristic proliferation of dendritic melanocytes.

Melanocytic matricoma is distinct from pilomatricoma, both clinically and pathologically. Clinically, pilomatricoma usually presents as a nodule in younger people, while melanocytic matricoma appears to present as a papule in elderly individuals. Pathologically, pilomatricoma is a solid neoplasm with a cystic tendency, located in the deep dermis and subcutis, with calcification and granulomatous reaction usually present. In distinction, melanocytic matricoma lacks a cystic component, is more superficially located, and apparently does not tend to calcify or elicit granulomatous response. Most importantly, pigmented pilomatricomas do not tend to exhibit prominent melanocytic hyperplasia, while melanocytic matricoma invariably contains a prominent proliferation of pigmented dendritic melanocytes. As melanocytes are more prominent in the early anagen phase of the hair cycle, melanocytic matricoma may represent an earlier stage of anagen follicular differentiation, while pilomatricoma may represent a later stage.[3]

In conclusion, melanocytic matricoma is a distinctive clinical and pathologic entity that, up to this point, represents a benign pigmented papule in sun-damaged areas of older individuals, formed by a superficial, dermal, pigmented nodular proliferation of matrical and supramatrical cells admixed with dendritic melanocytes. This recently recognized adnexal neoplasm needs to be recognized by pathologists, as it often enters the differential diagnosis of malignant tumors, including melanoma.

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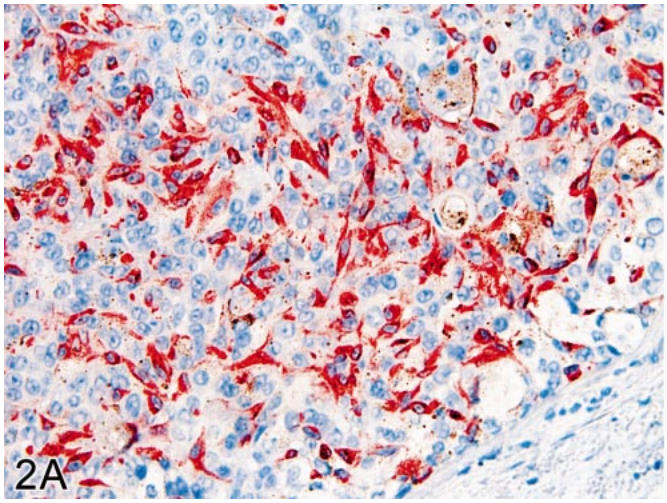

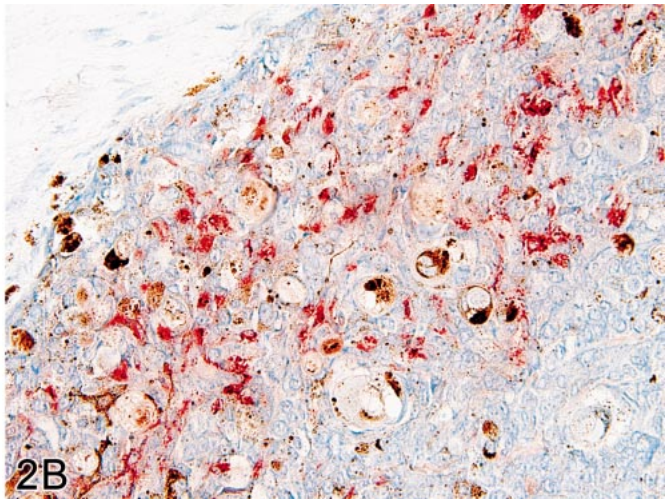

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