Pilomatrixoma misdiagnosed as dermatofibrosarcoma protuberans

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To the Editor: Pilomatrixoma is a benign tumor that arises from hair follicle matrix cells and has a low recurrence rate.[1] Dermatofibrosarcoma protuberans (DFSP) is a low-grade, slowly growing fibroblastic sarcoma with a high local recurrence rate and a low propensity for metastasis.[2] We report a case that was clinically suspected to be DFSP, but the surgical biopsy confirmed a diagnosis of pilomatrixoma.

A 29-year-old woman presented with a 1-year history of a localized asymptomatic mass on the posterior aspect of her neck [Figure 1A]. The lesion had begun as a slowly growing, asymptomatic, bean-sized, reddish nodule. After external friction 6 months previously, the lesion had become a rapidly growing, painful, firm, reddish mass with ulceration and bleeding. After topical application of mupirocin ointment, the pain was relieved and the mass was slightly reduced in size. No enlargement of superficial lymph nodes was found. A skin punch biopsy showed spindle cell masses in the dermis with an unclear boundary [Figure 1B]. Immunohistochemical staining showed that the spindle cells were positive for factor XIIIa, vimentin, and CD68 but negative for CD34, CD1a, and S100, and the Ki67 index was 10%. Based on these results, we clinically suspected DFSP; however, the immunohistochemical results were not consistent with DFSP. The patient then underwent surgical resection of the mass. Interestingly, the postoperative pathological examination showed that the mass was composed of basophils, shadow cells, and calcium in the lower dermis and subcutaneous fat, which was consistent with pilomatrixoma [Figure 1C and 1D]. No lesions were found in the lateral and incisional margins. No recurrence was observed during the 1-year follow-up.

In the clinical setting, clinicopathological incongruity is commonly encountered because of inadequate tissue. Roozeboom *et al*[3] reported that the agreement between the basal cell carcinoma subtype on punch biopsy and the subsequent surgical excision of primary basal cell carcinomas was only 60.9%. In the present case, the principal lesions were not taken during the punch biopsy, leading to misdiagnosis. Therefore, dermatologists must be aware of the limited diagnostic value of a punch biopsy for deeply located skin diseases, especially tumors. In addition, both pilomatrixoma and DFSP are often misdiagnosed in the clinical setting.[4,5] Pilomatrixoma usually presents as a blue-gray firm nodule of the head and neck in children or adults aged >50 years, and most of these lesions are <3 cm.[1] DFSP most commonly begins as an asymptomatic, indolent growing, indurated plaque.[2] It later becomes painful and exhibits accelerated growth, transforming into a raised nodule that ulcerates and bleeds.[2] These nodules occur most frequently on the trunk and proximal extremities, especially on the chest and shoulders in middle-aged adults.[5] Histopathologically, pilomatrixoma is characterized by basophilic cells with or without ghost cells and calcification.[4] DFSP is characterized by monomorphic spindle cells with low mitotic activity arranged in a "straw mat" pattern, and these cells invade the subcutaneous tissue through the septa and fat lobules.[5] Immunohistochemically, DFSP usually stains positively for CD34, hyaluronate, and vimentin but negatively for factor XIIIa.[2] Surgical resection is recommended for both pilomatrixoma and DFSP.[2,4] Mohs micrographic surgery is the preferred approach for localized DFSP.[2]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her names and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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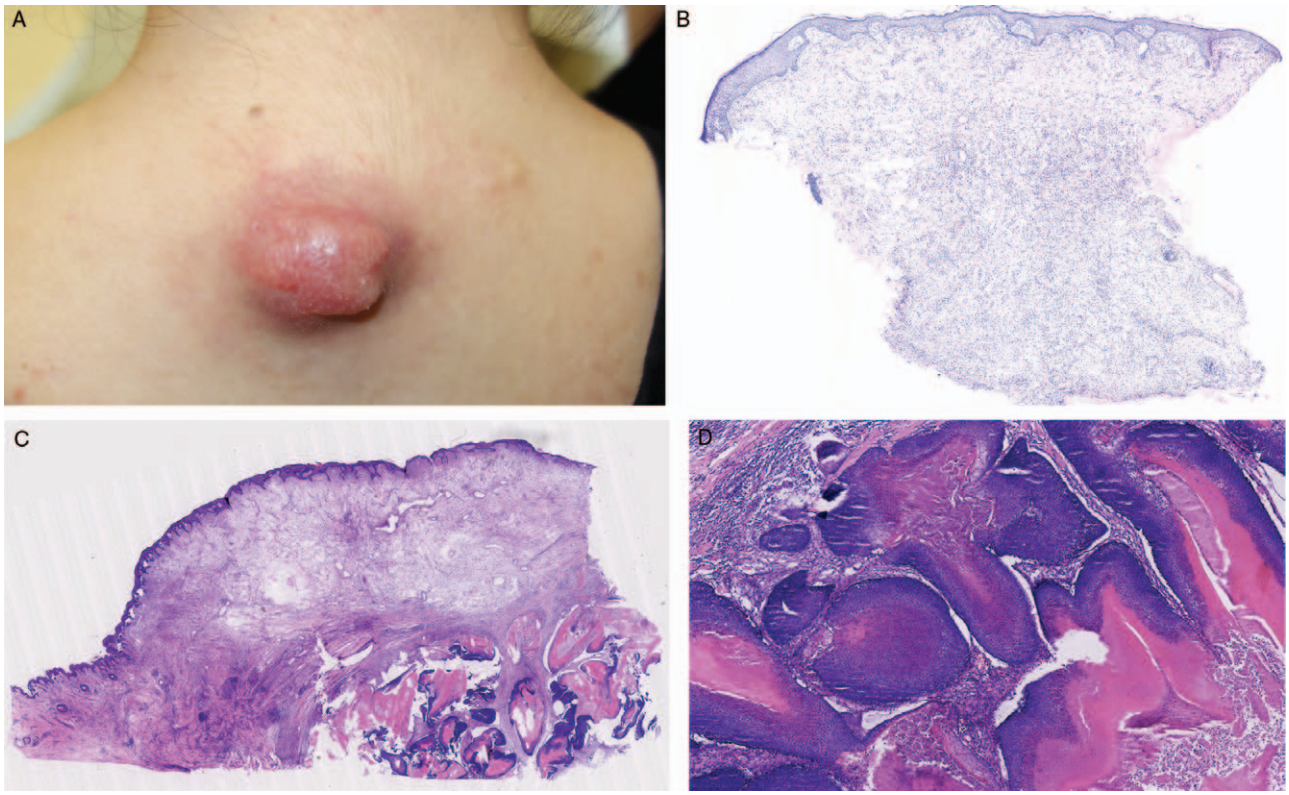

Figure 1: Clinical and pathological images of the case: (A) An asymptomatic, firm, reddish mass on the posterior aspect of the neck of a 29-year-old woman. (B) Punch biopsy of the skin showing diffuse proliferation of spindle cells in the dermis with an unclear boundary (Hematoxylin-eosin staining, Original magnification ×10). (C, D) Postoperative pathological examination showing a mass composed of basophils, shadow cells, and calcium in the lower dermis and subcutaneous fat [Hematoxylin-eosin staining, Original magnification ×10 (C), ×100 (D)].

Conflicts of interest

None.

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