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Case presentation

Bullous pilomatricoma: a rarely reported variant of pilomatricoma

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Abstract

Pilomatricomas are cutaneous adnexal tumors with matrical differentiation. We report and describe a rare variant called bullous pilomatricoma.

Case synopsis

A female in her 20s presented for evaluation and treatment of an enlarging blister on her right arm of nine months duration. She reported that the area first appeared similar to a bruise and subsequently enlarged to form a blister-like lesion. The patient could not recall any inciting trauma and denied pain or tenderness to palpation. No previous treatment was attempted. Her past medical history, past surgical history, family history, and social history were unremarkable. On physical examination, there was a 3.5 x 3cm pink bulla containing palpable firm nodules on the right upper arm (Figure 1). It extended 1.5cm from the epidermal surface. There was no background erythema. Upon palpation, a sharp drop-off was noted between lesional and normal skin. Excisional biopsy was performed. The incised specimen revealed gelatinous material containing several calcified nodules (Figure 2). Histopathology revealed dilated, thin-walled vascular structures within the papillary dermis (Figure 3). Deeper in the dermis there was a multilobulated tumor, surrounded by a fibrous pseudocapsule of compressed connective tissue elements. An admixture of basaloid and ghost cells were seen within the tumor and focal calcifications were present (Figure 4). The final diagnosis was determined to be a bullous pilomatricoma.

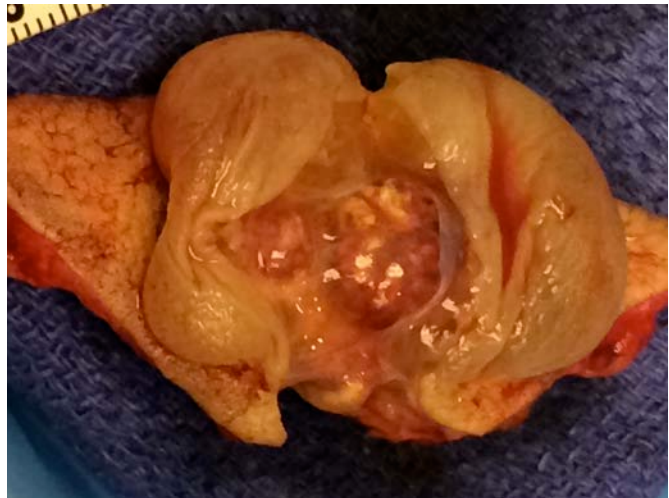


Figure 1. Pink bulla containing palpable firm nodules measuring 3.5 x 3 cm on the right upper arm. **Figure 2.** Incised lesion with central gelatinous material containing several calcified nodules.

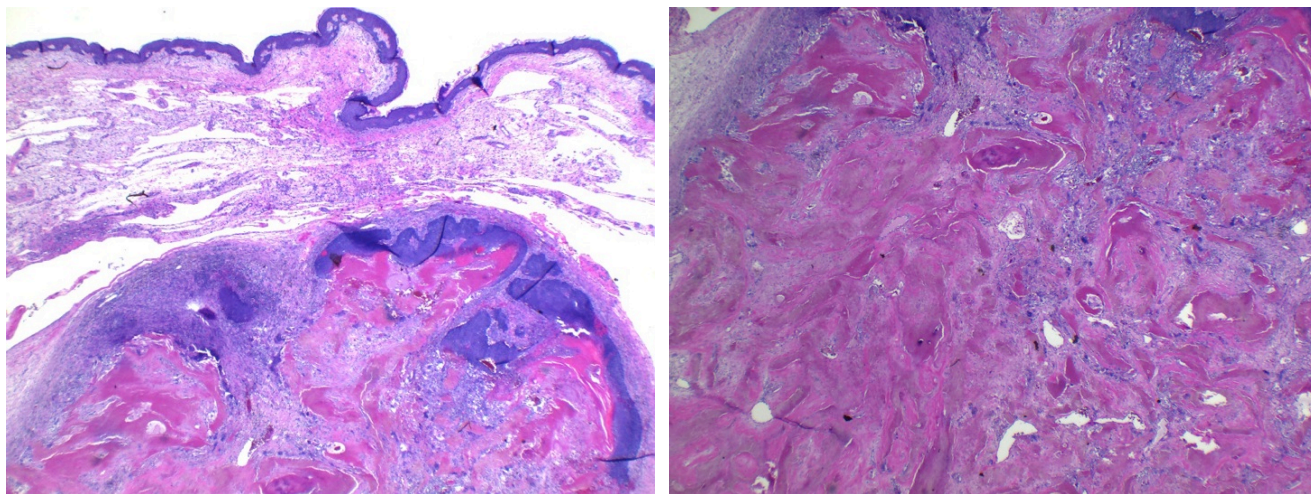


Figure 3. H&E 20x. Dilated, thin-walled vascular structures within the papillary dermis, representing dilated lymphatics. **Figure 4.** H&E 40x. Admixture of basaloid and ghost cells with focal calcifications, consistent with pilomatricoma.

Discussion

Pilomatricomas are benign cutaneous adnexal tumors with hair matrix differentiation [1-3]. Representing 20% of hair-follicle tumors, pilomatricomas tend to present as solitary, non-tender nodules on the head, neck, or upper extremities in females under the age of 20 [1]. Pilomatricomas are precipitated by mutations in CTNNB1, a gene encoding β -catenin, which is linked with upregulated hair follicle differentiation and decreased apoptosis [1,4]. As the clinical presentation of pilomatricomas may vary, histopathologic correlation demonstrating “ghost” or “shadow” cells, basophilic cells, and calcification is necessary [1,4].

Bullous pilomatricoma is a rare variant of pilomatricoma with a bullous appearance [1,3]. They most commonly occur in females between 10-20 years of age on the shoulder and upper arm [2]. A literature search limited to the English language revealed 21 reported cases of bullous pilomatricoma. The reported cases were not associated with Turner’s syndrome, myotonic dystrophy, Gardner’s syndrome, or β -catenin mutations as in traditional pilomatricomas. There are no reported clinical associations or implications for bullous pilomatricoma [1]. The most consistent factor contributing to the development of bullous pilomatricoma in reported cases was prior trauma. The differential diagnosis for bullous pilomatricoma should include lymphangioma, malignancy, bullous morphea, or secondary anetoderma [5].

The histopathology of bullous pilomatricomas has the hallmarks of pilomatricoma, including basaloid nests and eosinophilic shadow cells, but additionally exhibits ectasia of lymphatic vessels filled with lymphatic fluid [1,3]. The pathophysiology of these findings is likely related to obstruction of lymphatic vessels by a pilomatricoma resulting in congestion and dilation of the vessels with extravasation of lymphatic fluid into the dermis forming a bullous appearance [3]. Treatment is similar to that of pilomatricomas, with surgical excision being curative. There have been no reported cases of malignant transformation of bullous pilomatricoma and an excellent prognosis follows surgical excision [1].

We report this rare case of bullous pilomatricoma to remind the practicing dermatologist of this entity that is easily diagnosed and definitively treated with excision.

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