QUIZ SECTION

MJS PUBLISHING

A Thick-walled Pink Bulla with a Firm Nodule Underneath: A Quiz

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A 20-year-old man was referred for treatment of a cutaneous "cyst" on his left upper arm. A few months previously, the lesion had grown substantially, and he had no history of trauma. Physical examination revealed a thick-walled asymptomatic pink bulla, measuring 47 × 28 mm in size (Fig. 1), with palpation revealing a firm mass within. A greyscale ultrasound image showed a heterogeneous hyperechoic mass with scattered punctate echogenic foci, along with posterior acoustic shadowing. The overlying dermis contained anechoic areas (Fig. 2A). Color Doppler imaging exhibited intratumoral hypervascularity (Fig. 2B).

What is your diagnosis?

1: Keloid
2: Epidermoid cyst
3: Bullous pilomatricoma
4: Dermatofibrosarcoma protuberans

See next page for answer.

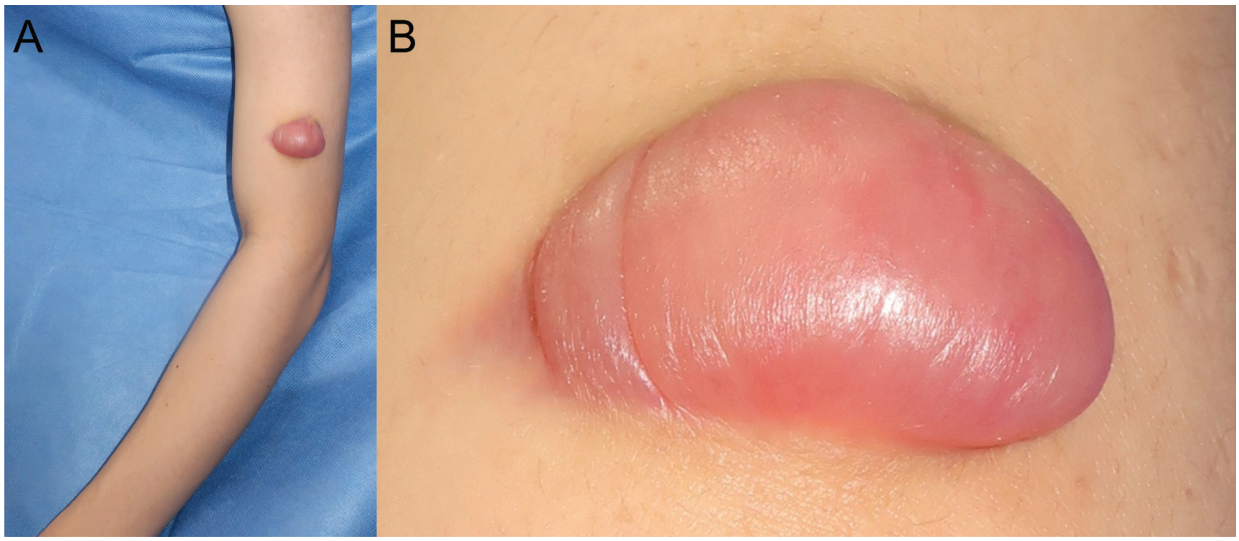

Fig. 1. Clinical presentation. (A) Photograph demonstrating a 4-cm pink bulla on the left upper arm of a 20-year-old man. (B) Close-up view.

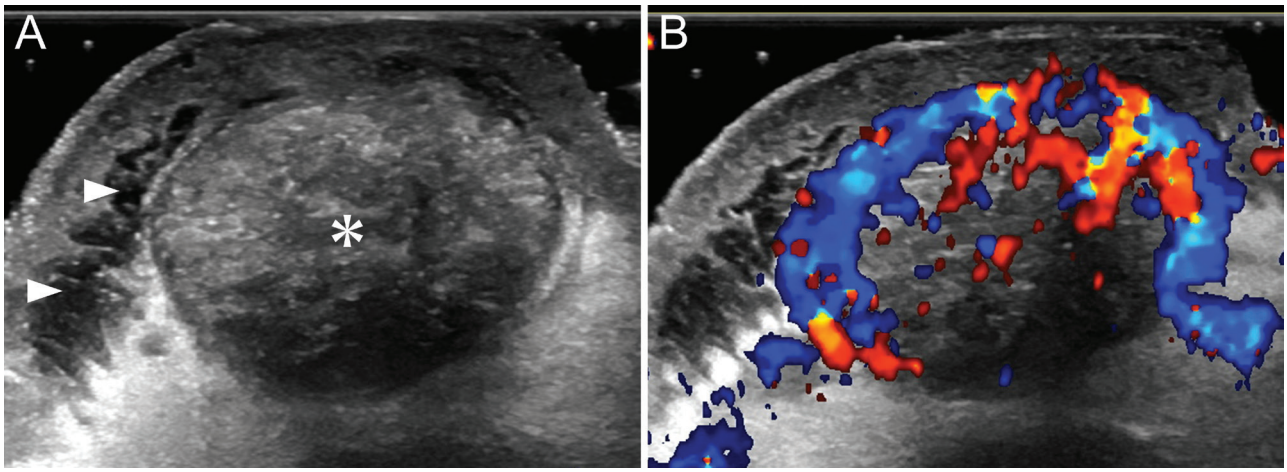

Fig. 2. Greyscale and colour Doppler ultrasound images. (A) Greyscale ultrasound image showing an oval-shaped heterogeneous hyperechoic mass involving the dermis and subcutis (asterisk) with scattered hyperechoic punctate foci and posterior acoustic shadowing. The dermis contains anechoic areas (arrowheads). (B) Colour Doppler imaging showing intratumoral hypervascularity.

ANSWERS TO QUIZ

A Thick-walled Pink Bulla with a Firm Nodule Underneath: A Commentary

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***Diagnosis*: Bullous pilomatricoma**

Excisional biopsy revealed a dermal and hypodermal nodule consisting of basophilic and eosinophilic shadow cells (Fig. 3A). The overlying dermis showed separation of collagen bundles in an oedematous stroma (Fig. 3B). Higher magnification showed scattered calcifications within the nodule (Fig. 3C). Accordingly, a diagnosis of bullous pilomatricoma was established.

Bullous pilomatricoma, also called anetodermic pilomatricoma, is an uncommon variant that presents clinically as a semi-transparent wrinkled blister with a firm nodule underneath (1). The lesions occur mostly in children and young adults (1). The surface coloration varies: most are red, and the others are blue, purple, or normal skin colour (1). While pilomatricoma mainly affect the head and neck (2), the bullous variant occurs predominantly on the upper arm and shoulders (1). Theories have been proposed to explain the mechanism of bullae formation, including mechanical irritation, obstruction of lymphatic vessels resulting in congestion and leakage of lymphatic fluids (1), and degradation of elastic fibres and collagen by matrix metalloproteinases-9 and -12 (3). In our case, the patient reported no history of trauma, and immunohistochemistry for D2-40 did not show lymphatic vessels lining dermal spaces.

Bullous pilomatricoma can be misdiagnosed as keloid and dermatofibrosarcoma protuberans (DFSP) (4). In the present case, a cyst was initially suspected. To avoid misdiagnosis, enhanced awareness of bullous pilomatricoma and palpation of the mass within a blister is critical. The clinical context of its mimickers may help exclude the differential diagnoses. Keloid usually occurs in areas of previous trauma and its development is associated with genetic and environmental factors. DFSP commonly affects the trunk and leg (5). For clinical diagnosis of an epidermoid cyst, identification of a punctum is useful, and compression of the cyst accentuates the appearance of the punctum (6).

Ultrasound may help distinguish bullous pilomatricoma from other differential diagnoses. On ultrasound, pilomatricomas most often appear as hyperechoic nodules with posterior acoustic shadowing and show vascularity on colour Doppler imaging (5). As for the bullous variant, the tumour shares sonographic features with the common type, and the stroma, consisting of remarkably oedematous connective tissue, can show anechoic lagoons (7), similar to those seen in our case. In contrast, epidermoid cysts typically present as hypoechoic nodules with posterior acoustic enhancement and lack vascularity (5). DFSP mainly presents as a homogeneous, isoechoic/hypoechoic mass with vascularity (5). Keloid presents as a hypoechoic mass with or without vascularity and rarely shows hyperechoic spots representing calcification (8).

In conclusion, we present a case of bullous pilomatricoma with sonographic evaluation. This variant is uncommon, and our report will enhance its awareness. Ultrasound and colour Doppler may help in the identification of pilomatricoma before invasive treatment.

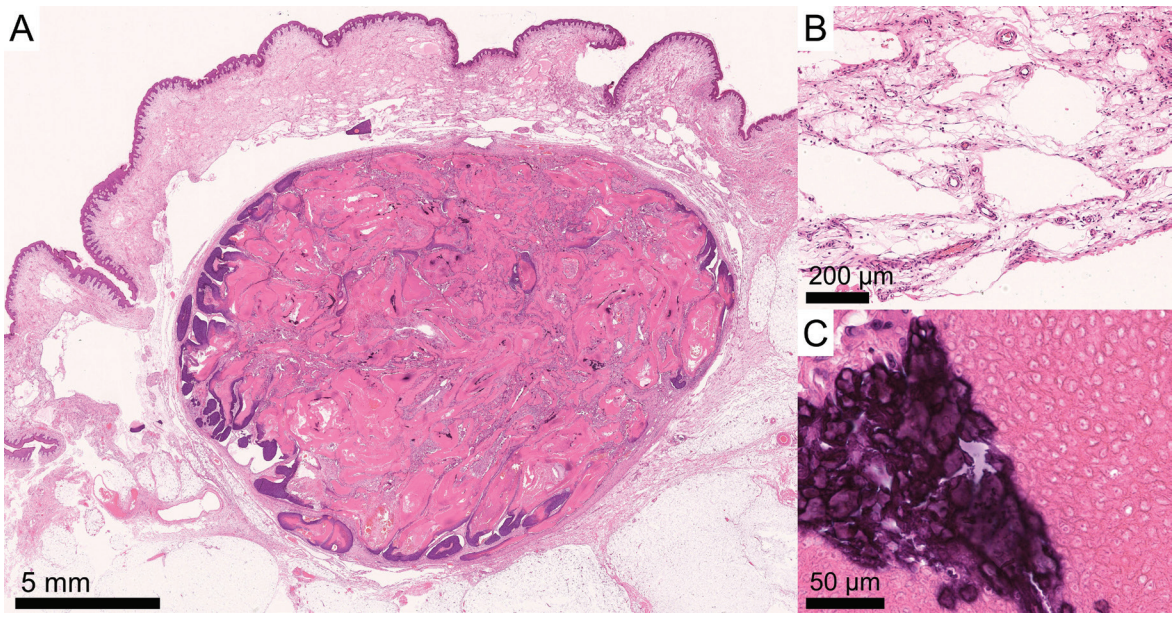

Fig. 3. Histopathology. (A) Histopathology of a resected specimen revealing a well-circumscribed nodule involving the dermis and subcutis composed of basophilic and eosinophilic shadow cells with scattered foci of calcification (haematoxylin–eosin [HE] stain, original magnification ×10). (B) Markedly oedematous dermis with separation of collagen bundles (HE, ×200). (C) Close-up view of calcification and shadow cells (HE, ×400). Scale bars=5 mm (A), 200 µm (B), and 50 µm (C).

The authors have no conflicts of interest to declare.

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