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Case Report

Dermatoscopic, Ultrasound and Histopathological Findings of Bullous Pilomatricoma: A Case Report

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Abstract

Pilomatricoma is the second most common benign tumor in the pediatric population after the epidermal cyst. It presents clinically as a firm, painless, solitary, well-defined nodule. Bullous presentation is rare, so we present below a clinical case of this variant in which we highlight its dermatoscopic, ultrasound and histopathological characteristics.

Keywords: Pilomatricoma; Bullous Pilomatricoma; Ultrasound

Introduction

Pilomatricoma, also called pilomatrixoma or calcifying epithelioma of Malherbe, is a benign adnexal dermal tumor derived from the matrix of the hair follicle [1]. It is located mainly on the face, neck and proximal upper extremities, while the trunk and lower extremities are less affected [2]. It most commonly presents in the first and second decades of life [3].

Clinically it is a firm, mobile, slow-growing and asymptomatic nodule. While nodules in isolation are most common, patients can also present with multiple. Size ranges from 0.4 to 20 cm [4]. Clinical forms include: mass (giant), pigmentation, mixed, ulcerative, keloid-like, multiple eruptive, familiar and bullous types [4,5].

This case outlines a pediatric patient with bullous pilomatricoma, a rare form which represents only 2% of all cases [6]. Dermatoscopic, ultrasound and histopathological characteristics are described, which support the diagnosis.

Case Report

A 12-year-old female presented to our dermatology clinic with a 2-year history of a solid tumor which had grown rapidly over 2 weeks. She was previously evaluated at another clinic and given antibiotics for a suspected soft tissue infection, which yielded no significant improvement. She denied a history of prior injury, vaccination or other trauma to the affected area.

Physical examination revealed a soft, 4 cm x 4 cm x 2 cm bulla containing yellow fluid on the left shoulder. There was no tenderness to palpation, warmth or surrounding erythema (Fig. 1).

Dermatoscopy revealed a white, homogeneous, structureless area on a yellow background (Fig. 2). Multiple bright white lines, telangiectasias and long, irregular linear vessels were distributed around the bulla were observed (Fig. 2).

Ultrasound showed a solid nodule measuring 2.5 x 2.0 cm, exhibiting well-defined contours and heterogeneous content with punctate calcifications. It was surrounded by a hypoechoic ring. Color Doppler assessment demonstrated peripheral vascularity on the superficial aspect of the lesion (Fig. 3).

Complete excision was performed and histology showed marked edema in the papillary dermis with dilated capillaries and lymphatic vessels, as well as inflammatory infiltrate and the presence of foreign body-type multinucleated giant cells. lslands of basaloid cells with abrupt keratinization and the presence of ghost cells and foci of calcification were observed in the center of the tumor (Fig. 4).

These collective findings were compatible with the diagnosis of bullous pilomatricoma. Surgery was performed to completely excise the lesion without complications; no recurrence has been identified.

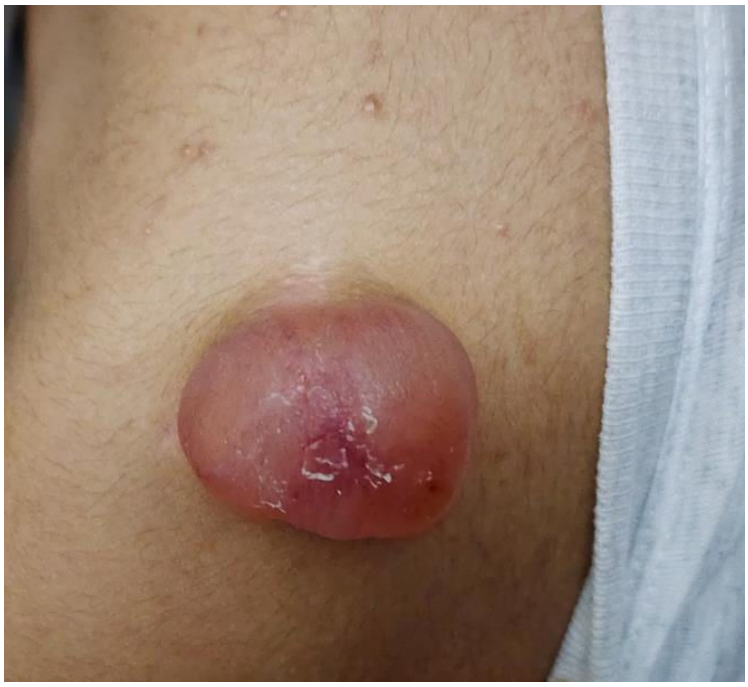
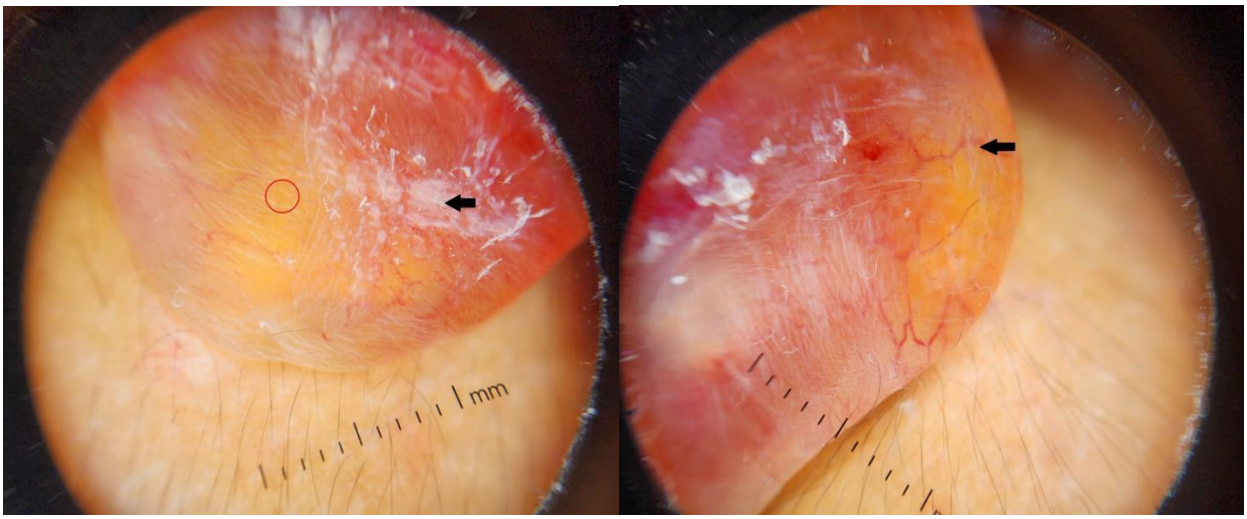


Figure 1: A 4 cm x 4cm x 2 cm bullae on the left shoulder.



A B

Figure 2: Dermatoscopic view. A: White homogeneous and structureless areas (black arrow). Yellowish background (red circle); B: Long, linear vessels around of the bullae (black arrow).

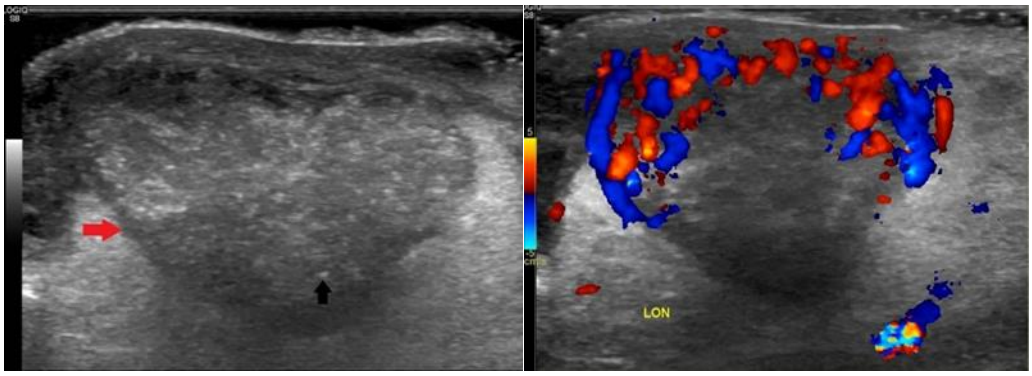


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Figure 3: Ultrasound findings. A: Heterogeneous content with punctate calcifications (black arrows) and hypoechoic ring (red arrow); B: Color Doppler shows peripheral vascularity.

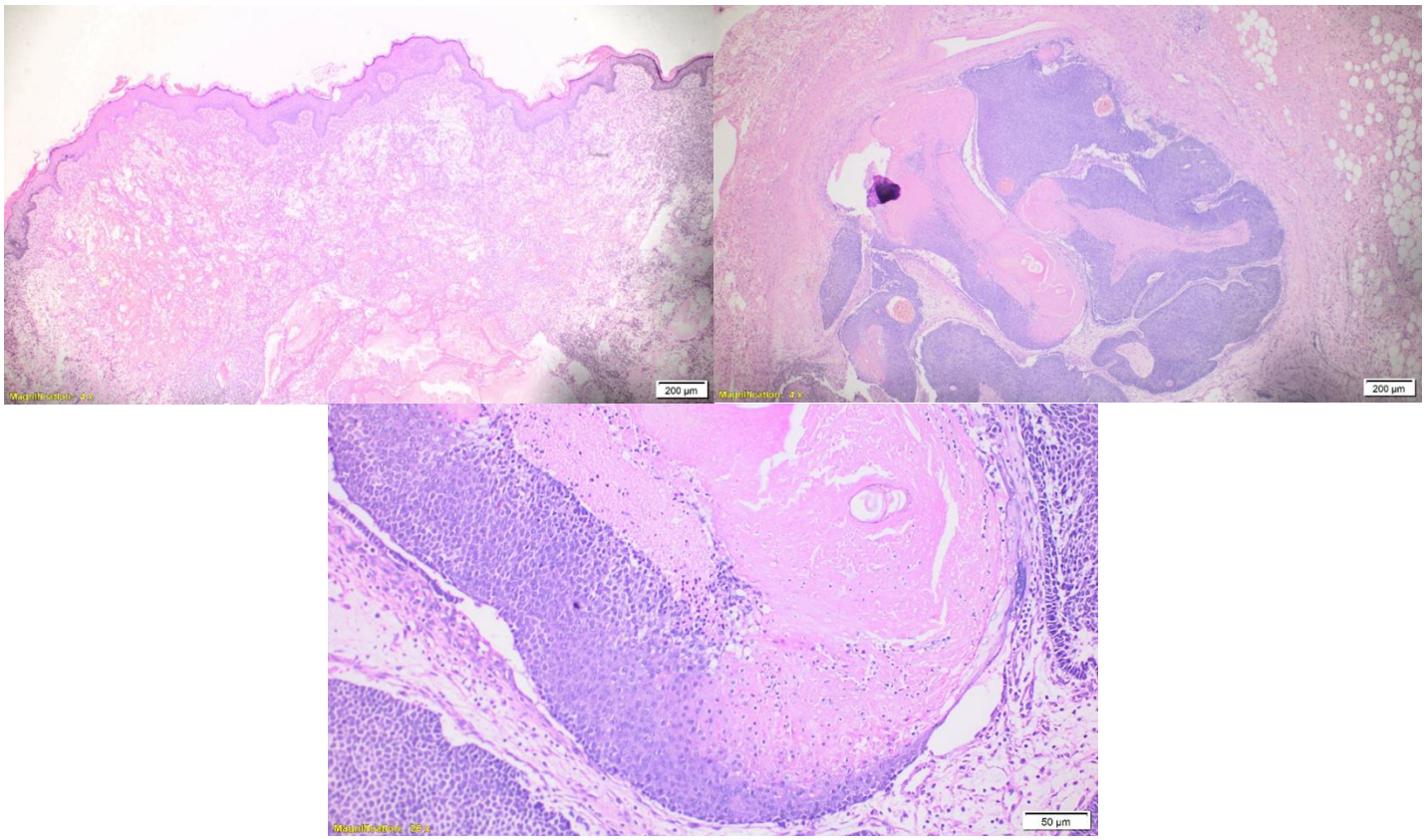

Figure 4: Histologic findings. A: Edema in the papillary dermis and dilated capillaries and lymphatic vessels (40X); B: lslands of basaloid cells with abrupt keratinization (40X); C: Abrupt keratinization and ghost cells (200X).

Discussion

Bullous pilomatricoma or lymphangiectatic variety is an asymptomatic, slow-growing tumor that can be covered by normal tissue and less frequently, by blistered skin [1]. The bullous form is preferably located on the shoulder and upper arm. The most common age range reported is 10 to 20 years. The size varies between 1 to 3 cm, with some cases reaching 6 cm. It usually presents as a solitary, flaccid, thick-walled red blister with a firm, palpable base [4].

The differential diagnosis should include other benign tumors such as epidermal inclusion cysts, dermoid cysts, dermatofibromas, hemangiomas and lipomas [7].

Pilomatricomas develop because of a genetic mutation in exon 3 of the B-catenin gene (CTNNB1), which is responsible for regulating epithelial cell adhesion [1].

Bullous pilomatricomas have obstruction of lymphatic vessels and congestion of lymph secondary to nodule growth. This leads to a dilation of the lymphatic vessels, leakage of lymphatic fluid and edema in the dermis surrounding the tumor [8]. It is also suggested that the inflammatory cells surrounding the pilomatricoma produce elastinolytic enzymes like Matrix Metalloproteinase (MMP) 9 and 12, which alter and destroy elastic fibers and collagen. This further facilitates the dilation of lymphatic vessels and allows the accumulation of fluid in the dermis, forming the blister [9,10].

Additionally, localized trauma has been described as a trigger for bullous pilomatricoma. Cases following vaccination in the shoulder have been reported, where mechanical stress and a subsequent inflammatory reaction likely led to activation of metalloproteinases and the development of secondary anetoderma [11,12].

In order of frequency, typical dermatoscopic findinds of pilomatricomas include; an erythematous background, irregular linear vessels, punctate vessels, white lines, homogeneous white areas and blue areas [13]. There are scarce reports of dermatoscopic descriptions of bullous pilomatricomas, thus far, red tortuous small vessels on an irregular white opacity, which settled on a livid-red background, has been described [14,15]. Dermatoscopic findings in our case include the yellow background described, which corresponds to the intense edema surrounding the pilomatricoma.

Reported ultrasound findings of bullous pilomatricomas are also scarce, however two ultrasound patterns have been described. The first and most common is the targetoid pattern, which consists of a hypoechoic ring corresponding to the capsule of connective tissue and a hyperechoic center due to foci of calcification and therefore produce posterior acoustic shadow [16,17]. The second pattern is a hyperechoic upper arch due to a large calcification at the upper of the tumor, producing a posterior acoustic shadow that does not allow other structures to be seen [16]. Our case shows the targetoid pattern in the center and a large hypoechoic area in the upper part of the tumor that corresponds to the significant edema that surrounds it.

Histopathologically, they are characterized by a well-circumscribed tumor surrounded by a connective tissue capsule that surrounds epithelial islands embedded in a cellular stroma [18]. Islands are composed of basaloid cells and ghost cells. Basaloid cells are usually arranged in the periphery and ghost cells are adjacent to the basaloid cells, sometimes in progressive transition and other times abruptly. These islands usually surround areas of calcification or ossification and multinucleated giant cells are also visible [18]. Reports of bullous pilomatricomas have also identified dilated lymphatic vessels, disruption of elastic fibers and collagen and lymphedema in the superficial dermis [15,19-21].

Conclusion

Treatment for bullous pilomatricoma is surgical with complete excision of the lesion, with low recurrence rates. In classic and small forms of pilomatricomas, efficacy has been described through incision and curettage.

Conflict of Interests

The authors have no conflict of interest to declare.

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