Anetodermic pilomatricoma: clinical, histopathologic, and sonographic findings

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Abstract

Pilomatricoma is a benign cutaneous tumor originating from hair matrix cells. Anetodermic changes in the skin overlying pilomatricomas are sometimes reported, although their precise mechanisms remain unknown. We present an unusual case of anetodermic pilomatricoma on the upper extremity of a 17-year-old boy and report its clinical, histopathologic, and sonographic findings.

Keywords: anetodermic pilomatricoma, anetoderma, cutaneous sonography, hair follicle tumor

Introduction

Pilomatricoma is a common benign adnexal tumor originating from the matrix of the hair follicles. We report an unusual case of pilomatricoma with associated anetoderma on the upper extremity of a 17-year-old boy. We describe clinical, histopathologic, and sonographic findings of this uncommon variant of pilomatricoma.

Case Synopsis

We report on an otherwise healthy 17-year-old boy who presented with a 4-year history of a painless slowly growing cutaneous lesion on his left arm. There was no history of prior trauma or injection. Physical examination revealed a 3-cm in diameter, soft, wrinkled, pink, bag-like protuberant lesion (**Figure 1**a). A firm underlying nodule was palpated within the sac. When vertical pressure was applied, the sac became flaccid, simulating a collapsed blister (**Figure 1**b). There was no regional lymphadenopathy and remaining physical examination was normal. Skin ultrasound showed a 3 x 1.6-cm, well-defined, oval, dermal and subcutaneous nodule with heterogeneous echotexture. There were internal echogenic foci in a scattered dot pattern and a peripheral hypoechoic rim (**Figure 2**a). Color Doppler sonography showed increased perilesional vascularization (**Figure 2**b).

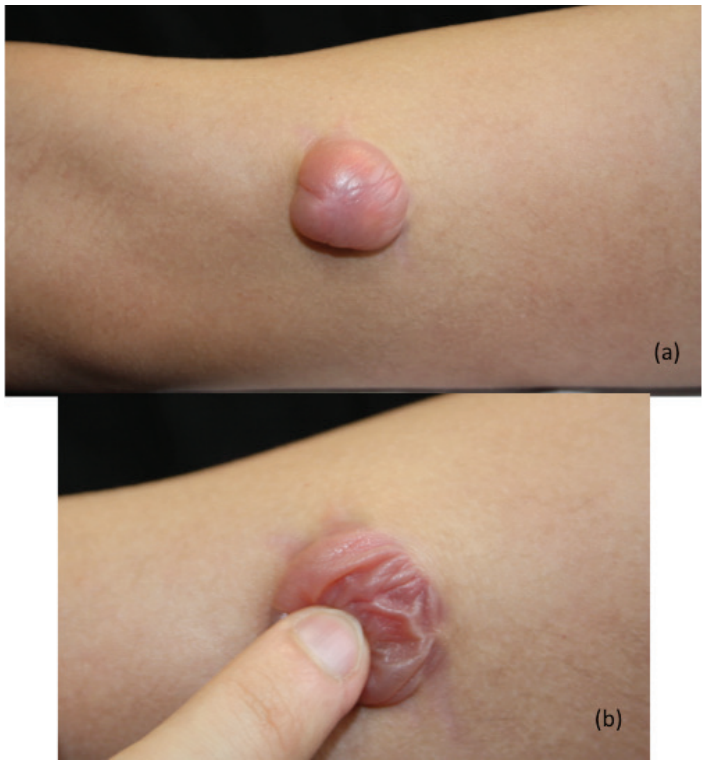

Figure 1. *(a) A 3-cm in diameter, soft-wrinkled, pink, bag-like protuberant lesion on left arm. (b) Skin overlying tumor becomes flaccid when vertical pressure is applied.*

The tumor was surgically excised. Macroscopic examination revealed a well-defined tumor with overlying bag-like skin and multiple areas of

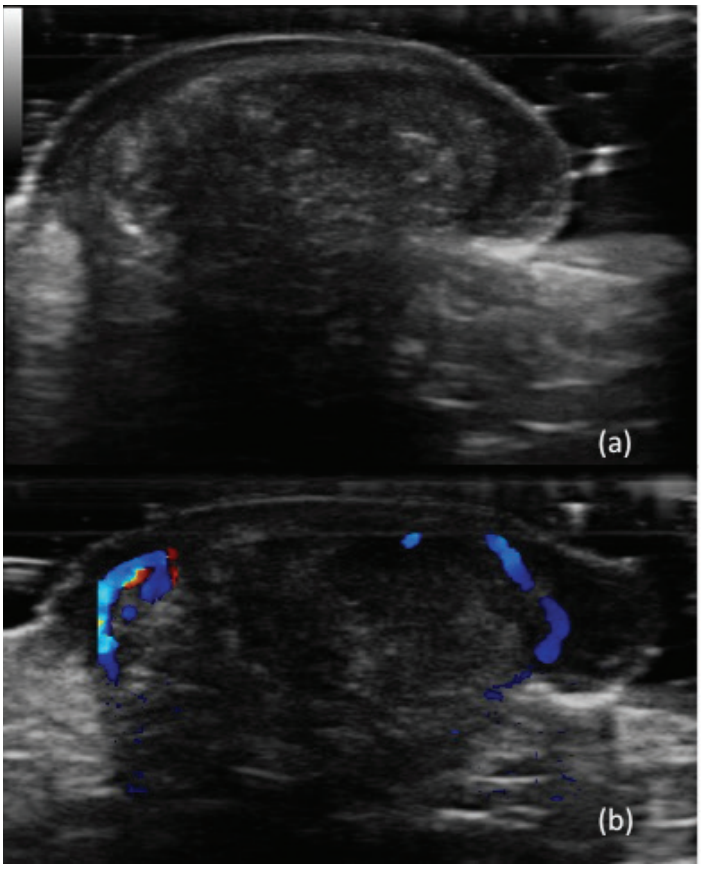

Figure 2. *(a) A well-defined, oval, dermo-hypodermic lesion with internal hyperechogenic spots and a peripheral hypoechoic rim. (b) Color Doppler sonography shows an increased perilesional vascularization.*

calcification (**Figure 3**a). Histologic examination revealed the tumor to be composed of islands of small, round, basophilic cells adjacent to pale, eosinophilic, enucleated shadow cells and transitional cells (**Figure 3**b, **3**c). Focal areas of calcification and foreign body granulomatous reaction were also seen. The orcein stain showed an absence of elastic fibers in the supra-tumoral dermis with some dilated vascular spaces (**Figure 3**d). The epidermis showed basaloid hyperplasia of keratinocytes.

Based on the clinical, sonographic, and histopathological findings, a diagnosis of anetodermic pilomatricoma was made.

Case Discussion

Pilomatricoma or calcifying epithelioma of Malherbe is a benign neoplasm derived from matrix cells of the hair follicles. Pilomatricoma typically presents as a firm, subcutaneous, solitary nodule with overlying normal skin. These tumors are usually located on the cephalic segment and scalp. It has two peak ages of presentation, in the first and sixth decades of life [1].

Anetodermic pilomatricoma, also called lymphangiectatic or pseudobullous, is an uncommon variant of pilomatricoma that constitutes 2% of these tumors. Clinically, it usually presents as pink to translucent, soft-wrinkled, atrophic or keloid-like skin over a firm, rapidly growing, subcutaneous mass. Anetodermic pilomatricoma can be depressed at the center when vertical pressure is applied (dimple sign). The most frequently involved sites are the upper arms and shoulders. The onset age is young with a female predilection [1, 2].

The etiology of anetodermic pilomatricoma is still unknown although different theories have been proposed. Li et al. postulated that mechanical trauma might play a critical role in its development by disrupting elastic fibers and lymphatic drainage, and inducing blood vessel proliferation [3]. This fact could explain why anetodermic pilomatricoma occurs mainly in areas prone to mechanical trauma. Other authors have speculated that mast cells could have a causative role by favoring elastolysis and increasing epidermal proliferation of the lesion through the activation of certain cytokines [2]. Other theories reported suggest that elastic-tissue destruction is caused by catabolic enzymes released by the tumor or the inflammatory infiltrate [4]. Mutations in β-catenin have been suggested as the causative factor for pilomatricomas [5].

Anetodermic pilomatricoma is histologically characterized by findings at three levels: a) a basaloid hyperplasia of keratinocytes in the epidermis, which is not always found; b) dilated lymphatic vessels in the dermis above the pilomatricoma with a lack of collagen fibers and absence of elastic fibers, which gives it a myxoid aspect; c) deep dermal pilomatricoma with islands of basaloid cells, similar to the matrix cells of hair follicles. At this level, eosinophilic shadow cells are exhibited with a transition zone of retained nuclei between them. Deeply basophilic calcium, focal areas of ossification and necrosis, and foreign-body granulomatous reaction with multinucleated giant cells may also be observed [1, 6]. Areas of transepidermal elimination of pilomatrical cells have been reported in some cases [7].

There are a limited amount of reports with a description of ultrasound findings of anetodermic pilomatricoma in indexed literature [8-10]. Anetodermic pilomatricoma show similar sonographic findings than conventional pilomatricoma, enabling a specific diagnosis. The tumor appears often as a well circumscribed, non-homogeneous echogenic nodule. Neither the peripheral hypoechogenic rim nor the variable peripheral vascularization are specific for pilomatrixoma. The only key sonographic finding so far is the early calcification of the tumor, which indeed is often (but not always or just in the later course of the disease) visible as hyperechogenic spots. Its imaging depends very much on the ultrasound device used and most certainly on its quality [8-10].

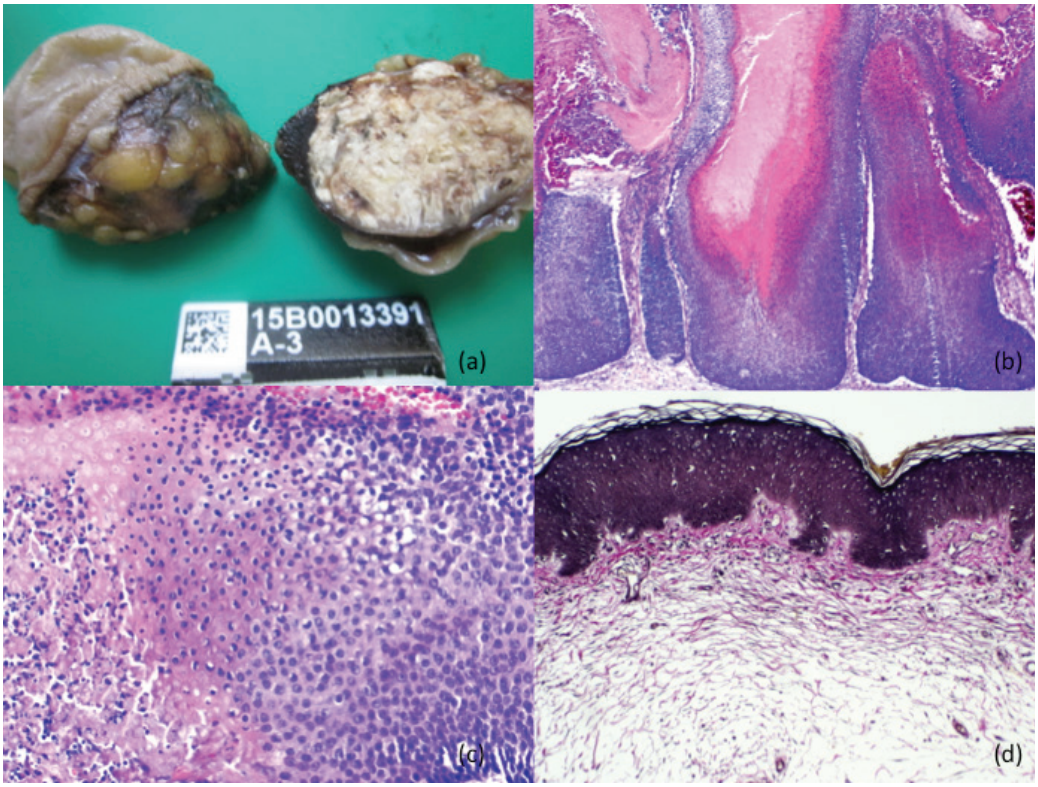

Figure 3. *(a) Bag-like skin over the pilomatricoma and multiple areas of calcification. (b, c) Histopathology shows islands of basaloid cells, eosinophilic shadow cells and transitional cells (H&E, original magnification x20; x40). (d) Absence of elastic fibers in supratumoral dermis with some dilated vascular spaces and basaloid hyperplasia of keratinocytes (orcein stain, original magnification x20).*

In some cases, the differential diagnosis should include basal cell carcinoma and malignant melanoma. In both cases, sonographic findings may help us to distinguish them. Basal cell carcinoma usually presents as a well-defined lesion with small hyperechoic nodules in the lesion and malignant melanoma is an ill-defined lesion with abundant vascularization within the tumor [11].

In conclusion, anetodermic pilomatricoma is a rare variant of pilomatricoma, which presents identifiable clinical, histopathologic and sonographic findings. Clinicians should be aware of this clinical type of pilomatricoma.

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