Dear Editor:

Pilomatricoma, a slow-growing, benign adnexal tumor of the skin, originates from primitive cells of the hair matrix. It usually appears as a painless, firm, subcutaneous nodule, which adheres to the overlying skin. The overlying skin normally has a bluish discoloration. Although lesions with bullous changes on the overlying skin are very rare, they have been reported. Herein, we report a case of bullous pilomatricoma to enhance the recognition and proper management of this variant, which represents a particular diagnostic challenge.

An 18-year-old woman chiefly complained of a slightly tender, bullous skin lesion on her right upper back (Fig. 1A). Three months earlier, the lesion had appeared as a painless, firm nodule that enlarged with time beneath the skin. The bullous appearance was detected 1 month before her visit. The bulla was thick-walled, semitransparent, and sized 2.5 × 3 cm. A palpable mass was faintly noted inside the bulla. Systemic and laboratory examination results were normal. Preoperative bulla bisection revealed a firm central tissue core, surrounded by a sac and serous fluid (Fig. 1B). The lesion was totally excised and closed directly. The 1 × 2 cm mass inside the bulla was similar to a classical pilomatricoma: well demarcated,
lobulated, and hard. On microscopic examination, we observed typical “ghost cells”; anucleate keratinized basaloid cells (Fig. 2A, B). Endothelial cells of the dilated lymphatic channels were positive, on immunohistochemical staining, for CD34 and D2-40. Pseudocysts without endothelial cell lining were noted in the superficial dermis (Fig. 2C). The patient remained well without recurrence during follow-up for six months.

Reports on bullous pilomatrixoma are limited. To date, a total of 18 cases have been published. Bullous pilomatrixoma has a predilection for the shoulder and upper arm regions, predominantly affecting women aged 10 to 20 years.

Lymphatic fluid fills the bulla-like space. It has been postulated that the pressure surrounding the hard core of the pilomatrixoma induces obstruction of lymphatic vessels and congestion of lymphatic fluid, resulting in dilation of lymphatic vessels, with leakage of lymphatic fluid, and edema in the dermis surrounding the tumor, producing a bullous appearance. Nearly all the cases of bullous pilomatrixoma are formed by typical tumor nests—composed of shadow and basophilic cells—surrounded by a fibrotic capsule. Most cases present dilated lymphatic vessels and lymphedema in the superficial dermis.

Ghost, or "shadow," cells are the most characteristic microscopic findings in pilomatrixoma cases. They have distinct cell borders, with eosinophilic cytoplasm and a central clearing, representing the outline of an absent nucleus. Commonly, there is an associated foreign-body multinucleated giant-cell reaction and fibrosis with calcification; hence, this lesion was originally described as "calcifying epithelioma of Malherbe.”

As for the classical pilomatrixoma, surgical excision is the treatment of choice because this lesion does not regress spontaneously. The prognosis is excellent, without reported recurrences or malignant changes after excision.

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REFERENCES