

CASE AND RESEARCH LETTERS

Ulcerated Giant Pilomatrixoma in a Child[☆]

Pilomatrixoma gigante y ulcerado en un niño

To the Editor:

We report the case of a 7 year-old boy who presented with a lesion on the left arm that had developed 2 years earlier and had suddenly increased in size in the preceding month. A Doppler ultrasound performed 1 month earlier revealed an echogenic mass of $1.6 \times 1.7 \times 1$ cm with posterior acoustic shadowing.

Examination revealed a stony-hard, exophytic, pedunculated tumor of 8 cm in diameter on the patient's left arm. The overlying skin was absent, exposing an eroded surface and a bleeding tumor bed with a crater-like appearance (Fig. 1). Nuclear magnetic resonance imaging (MRI) and magnetic resonance angiography showed an exophytic formation consisting of a heterogeneous nodule with a hypointense halo surrounded by hyperintense tissue, and a large number of tortuous vessels in the stalk (Fig. 2). The tumor was excised and the defect reconstructed with a Dufourmental flap. Histopathology revealed the presence of lobules of



Figure 1 Pedunculated exophytic tumor with eroded and bleeding surface.



Figure 2 Nuclear magnetic resonance imaging: exophytic formation with a heterogeneous nodule surrounded by a hypointense halo and abundant vascularity in the stalk.

basaloid cells showing maturation toward the surface, the formation of cells with abundant eosinophilic cytoplasm, sheets of eosinophilic keratinous debris, and pilar keratinization. The lobules were surrounded by a fibrous stroma with mononuclear, histiocytic inflammatory infiltrate and foreign body-type multinucleated giant cells. Numerous foci of dystrophic calcification were observed. These findings were consistent with a diagnosis of giant pilomatrixoma with foreign-body giant-cell reaction (Fig. 3). The patient progressed favorably and there was no recurrence after 1 year of follow-up.

Pilomatrixoma is a benign adnexal tumor with differentiation towards hair matrix cells, and is most common in children and those over 60.¹ Although the etiology of this tumor is unknown, in 75% of cases a mutation has been identified in *CTNNB1*, the gene encoding beta-catenin, a signaling protein involved in hair follicle development.²

Pilomatrixomas are slow-growing nodules measuring 1 to 1.5 cm across; they frequently calcify, giving rise to bluish discoloration of the overlying skin, which may extrude calcific material.³ Nodules can be multiple or solitary. Solitary tumors have been described in genetic

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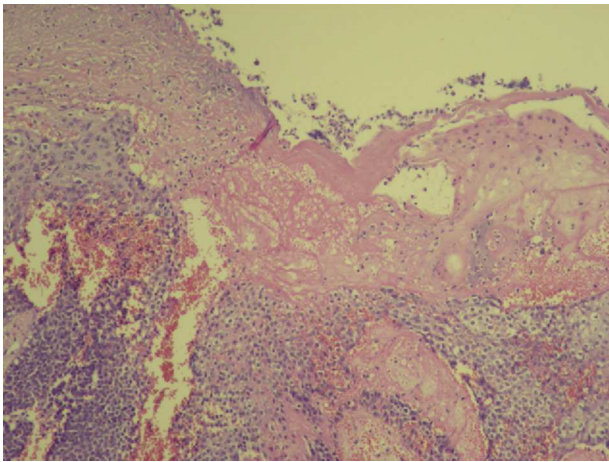


Figure 3 Histology: lobules of basaloid cells with maturation toward the surface and ghost cell formation (hematoxylin-eosin, original magnification $\times 100$).

syndromes such as Turner syndrome, Sotos syndrome, myotonic dystrophy, Rubinstein-Taybi syndrome, Gardner syndrome, trisomy 9, basal cell nevus syndrome, and xeroderma pigmentosum.⁴ Multiple pilomatricomas have been reported in healthy patients.⁵ Other clinicopathologic variants have also been described, including blistering, anetodermic, perforating/ulcerated, lymphangiectasic, and malignant forms.

Pilomatricomas of over 5 cm in diameter are considered giant. Only 4 cases of giant pilomatricoma have been described, all in adults^{6,7}: 2 ulcerated and 2 with areas of erosion. Two cases of giant pilomatricoma associated with hypercalcemia have also been described, both of which resolved after resection.⁷

The differential diagnosis includes dermatofibrosarcoma protuberans, calcinosis cutis, osteoma cutis, cutaneous lymphomas, sarcomas, squamous carcinoma, and cutaneous metastases.

The giant, ulcerated clinical form must be differentiated from the malignant form, and should be suspected in cases of sudden growth of a pre-existing lesion with ulceration and bleeding. Indicators of malignancy include necrosis, atypical mitotic figures, perineural or perivascular invasion, and the degree and extent of infiltration.⁸

Ultrasound reveals an echogenic lesion with posterior acoustic shadowing. In the present case the extension of the lesion was evaluated by MRI, which revealed an exophytic formation with hypervascularization, as described in a previous case of giant pilomatricoma.⁹

The pilomatricoma case described here is of interest due to the sudden increase in the size of the lesion; while this

characteristic suggested a malignant tumor, malignancy was subsequently ruled out by histopathology.

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References

1. Julian CG, Bowers PW. A clinical review of 209 pilomatricomas. *J Am Acad Dermatol.* 1998;39:191–5.
2. Gilaberte Y, Ferrer-Lozano M, Oliván MJ, Coscojuela C, Abascal M, Lapunzina P. Multiple giant pilomatricoma in familial Sotos Syndrome. *Pediatr Dermatol.* 2008;25:121–4.
3. Khammash M, Todd D, Abalkhail A. Concurrent pilomatric carcinoma and giant pilomatricoma. *Austral J Dermatol.* 2001;42:120–3.
4. Uchimiya H, Kanekura T, Gushi A, Fukumaru S, Baba Y, Kanzaki T. Multiple giant pilomatricoma, letter to the editor. *J Dermatol.* 2006;33:644–5.
5. Fernández Arias FJ, Marco RM, Heras Tabernero JA, Muñoz Marcos D, Barba Diaz L, Martínez Murillo P, et al. Pilomatricoma múltiple. *Casos breves. Actas Dermosifiliogr.* 2000;91:529–30.
6. Kovacic M, Rudic M, Nekić I, Lisica-Sikić N, Kranjčec Z, Simurina T. Giant pilomatricoma (benign calcifying epithelioma of Malherbe) of the neck and face. *Dermatology Surgery.* 2007;33:340–3.
7. Yamauchi M, Yotsuyanagi T, Saito T, Ikeda K, Urushidate S, Higuma Y. Three cases of giant pilomatricoma—considerations for diagnosis and treatment of giant skin tumours with abundant inner calcification present on the upper body. *J Plastic Reconstr Aesthet Surg.* 2010;63:e519–24.
8. Huerva V, Sanchez C, Asenjo J. Large, rapidly growing pilomatricoma of the upper eyelid. *Ophthal Plast Reconstr Surg.* 2006;22:401–3.
9. Lozzi GP, Soyer HP, Freuhauf J, Massone C, Helmut K, Ketty P. Giant pilomatricoma. *Am J Dermatopathol.* 2007;29:286–9.

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