Giant pilomatrixoma: a distinctive clinical variant: a new case and review of the literature.

Javier Sabater-Abad[1], María Matellanes-Palacios[1], Laia Bou-Boluda[1], Juan José Campos-Dana[2], Pilar Alemany-Monraval[3], Fernando Millán-Parrilla[1]

Affiliations: [1]Department of Dermatology, Hospital Arnau de Vilanova, Valencia, Spain, [2]Department of Otolaryngology, Hospital Arnau de Vilanova, Valencia, Spain, [3]Department of Pathology, Hospital Arnau de Vilanova, Valencia, Spain

Corresponding Author: Javier Sabater Abad, Department of Dermatology, Hospital Arnau de Vilanova, C/San Clemente, 12, Valencia, 46015, Spain, Tel: 34-639493570, Email: javiersabater999@gmail.com

Abstract

Pilomatrixoma is a benign adnexal tumor very common in pediatric age and in young adults that derives from follicular matrix cells. Although clinically it usually presents as a subcutaneous nodule of bluish color less than 3cm in size, multiple clinicopathological variants have been described in the literature. Among these we can find the giant pilomatrixoma, a rare clinical variant that reaches a size greater than or equal to 4cm and can simulate the clinical presentation of a malignant neoplasm. We report a 59-year-old man with an exophytic and ulcerated nodule in the left parotid region that was removed with the suspected diagnosis of a cutaneous squamous cell carcinoma. Histopathological analysis showed a proliferation of basaloid cells with areas of transition to ghost cells, under granulation tissue, hemorrhage, and an ulcerated epidermis. Thus, the diagnosis of giant pilomatrixoma was made. We reviewed the literature and found a total of 53 articles that report a total of 71 cases of giant pilomatrixoma. It is important to recognize this clinical subtype of pilomatrixoma because, apart from the possibility of being clinically confused with malignant lesions, the clinicopathological differential diagnosis must be made with the proliferating pilomatrixoma and pilomatrixcarcinoma.

Keywords: giant pilomatrixoma, clinical variant, ghost cells, malignant lesions, pilomatrixcarcinoma

Introduction

Pilomatrixoma (PM) is a benign adnexal tumor that derives from follicular matrix cells. Clinically it usually presents as a solitary subcutaneous nodule of slow growth and blue-black color with a usual size less than three cm. It is a very common lesion in the pediatric age and in young adults. It is most often located in the head and neck. Multiple clinicopathological variants of PM have been described in the literature, including the bullous, perforating, anetodermic, lymphangiectatic, and giant types [1,2]. We report a new case of giant PM and review the literature to better characterize this clinical subtype of PM.

Case Synopsis

We present the case of a 59-year-old man with no history of interest who presented to our clinic for an exophytic and ulcerated tumor of 4.5×3.5cm that had been present in the left parotid region for 9 months (**Figure 1**). With the clinical suspicion of a cutaneous squamous cell carcinoma, the lesion was removed and the surgical defect was reconstructed with a rotation flap. The histopathological study showed a proliferation of basaloid cells under granulation tissue, hemorrhage, and an ulcerated epidermis. In some areas, the transition of basaloid cells to cells with a broader and more eosinophilic cytoplasm and a smaller nucleus could be seen (**Figure 2**). Eventually these evolved to eosinophilic

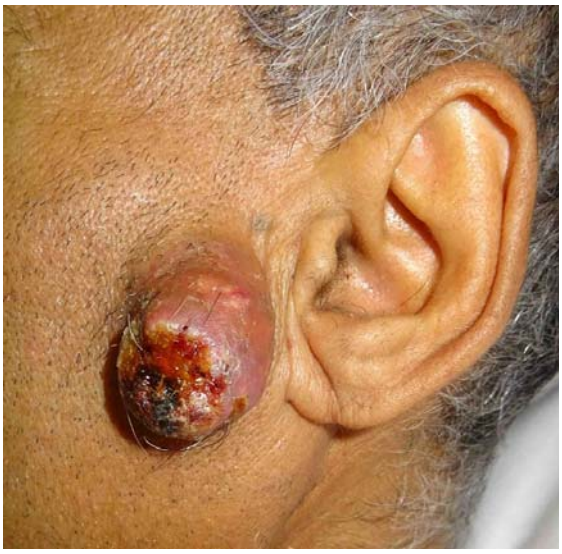

Figure 1. *Exophytic and ulcerated lesion of 4.5×3.5cm in the left parotid region.*

cells without nucleus, also known as ghost cells. Therefore, the diagnosis of giant PM was made.

Case Discussion

We have reviewed the literature in search of those PM with a diameter greater than or equal to 4cm and the first case was published in 1974 [3]. To the current date, a total of 53 articles report 70 cases of the giant variant of PM. According to the literature, it seems that giant PM is more frequent in adult men. The usual clinical presentation is a solitary lesion that has been present between one month and 40 years. The most frequent location is the upper limb followed by the parotid region and the back. The size may vary and the largest lesion reported to date was 34cm in diameter [4]. Nearly 40% of the giant PMs published are ulcerated, which together with their frequent location in the parotid region, makes the initial clinical suspicion more likely to be a malignant neoplasm [1] or a parotid neoplasm [5], such as cutaneous squamous cell carcinoma, dermatofibrosarcoma protuberans, cutaneous metastases, pleomorphic adenoma, and malignant parotid tumors. Despite the large size and ulceration of some giant PM, they are usually asymptomatic. When there are symptoms, the most common is localized pain.

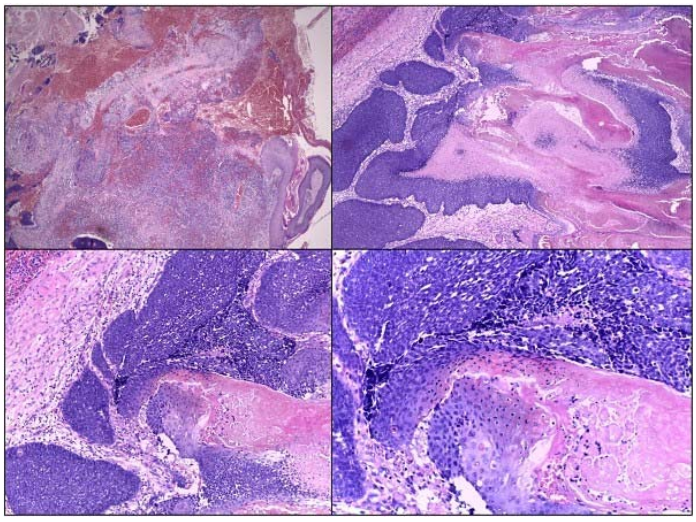

Figure 2. *Ulceration of the epidermis and abundant granulation tissue located above a proliferation of basaloid cells with areas of transition to broader and eosinophilic cytoplasm cells with loss of nuclei (ghost cells).*

Owing to the difficulty in making the clinical diagnosis, in more than half of the cases reported, an imaging test or cytological study using fine needle aspiration puncture (FNA) was performed. The most requested imaging tests were computerized tomography and magnetic resonance imaging. The most common radiological finding is a well-defined soft tissue mass, heterogeneous with calcifications, with abundant and tortuous vessels, and without invasion of neighboring structures. The most relevant cytological findings for the diagnosis of PM by FNA are basaloid cells and ghost cells, although multinucleated giant cells, nucleated squamous cells, and calcium deposits may also be helpful [6].

The histological differential diagnosis includes pilomatrical carcinoma or pilomatrixcarcinoma, a lesion that, architecturally, is usually asymmetric with ulceration, poorly defined, and shows an infiltrative growth pattern. Cytologically it is composed of pleomorphic basaloid cells with high mitotic index [2]. Regarding the perforating subtype of PM, which is a clinical variant of PM that presents as a nodule or ulcer that drains material [7], we could say that about 40% of giant PM in this review were perforating since this percentage of PM were ulcerated. We have also found a proliferating PM [2], a histopathological variant somewhat controversial because of the possibility that it is a precursor of pilomatricial carcinoma that is composed of a greater number of basaloid cells with atypia and

mitosis and a smaller amount of ghost cells. Apart from having found cases of giant PM associated with Steinert myotonic dystrophy or Sotos syndrome [8], we have also identified cases associated with malignant hypercalcemia [9] and reactive lymphadenitis [10]. Finally, treatment was surgical in all cases identified in the literature. Some required surgical reconstruction techniques. In more than half the cases located in the parotid region, a superficial parotidectomy was necessary owing to the proximity of the tumor with the parotid gland itself. Recurrences are not frequent and if they occur are related to the incomplete excision of the tumor.

Conclusion

Giant PM is a distinctive clinical variant of PM that, owing to the large size, the usual location in the parotid area, and the frequent ulceration can be confused clinically with malignant tumors or parotid tumors. It is important to know the existence of this entity and to distinguish it from a pilomatrical carcinoma.

Potential conflicts of interest

The authors declare no conflicts of interests.

References

1. Lozzi GP, Soeyr HP, Fruehauf J, et al. Giant pilomatricoma. *Am J Dermatopathol.* 2007;29:286-9. [PMID: 17519628].
2. Grohs RL, Mesbah Ardakani N, Harvey NT, et al. Giant proliferating pilomatricoma; report of a rare entity. *Australas J Dermatol.* 2017;58: e91–e93. [PMID: 27301983].
3. Krausen AS, Ansel DG, Mays BR. Pilomatrixoma masquerading as a parotid mass. *Laryngoscope.* 1974;84:528-35. [PMID: 4821511].
4. Beattie G, Tai C, Karakas P, et al. Colossal pilomatrixoma. *Ann R Coll Surg Engl.* 2018;100:e38–e40. [PMID: 29182000].
5. Cozzi DA, d'Ambrosio G, Cirigliano E, et al. Giant pilomatricoma mimicking a malignant parotid mass. *J Pediatr Surg.* 2011;46: 1855–8. [PMID: 21930004].
6. Wang J, Cobb CJ, Martin SE, et al. Pilomatrixoma: clinicopathologic study of 51 cases with emphasis on cytologic features. *Diagn Cytopathol.* 2002;27:167–72. [PMID: 12203865].
7. Ciralik H, Coban YK, Arican O. A case of perforating pilomatricoma. *Journal of Dermatology.* 2006;33:394-8. [PMID: 16700828].
8. Gilaberte Y, Ferrer-Lozano M, Olivan MJ, et al. Multiple giant pilomatricoma in familial Sotos Syndrome. *Pediatr Dermatol.* 2008;25:121-4. [PMID: 18304174].
9. Yamauchi M, Yotsuyanagi T, Saito T, et al. Three cases of giant pilomatrixoma - 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. [PMID: 20080452].
10. Namiki T, Miura K, Nojima K, et al. Ulcerated giant pilomatricoma with appearance of cutaneous malignancy on positron emission tomography/computed tomography. *J Dermatol.* 2017;44:220-222. [PMID: 27207071].