Case Reports and Reviews

Case Report

Pilomatrixoma of the face: a case report and literature review

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Abstract

Pilomatrixoma is a benign skin neoplasm of the hair follicle. It's an often misunderstood adnexal tumor confused with other skin lesions mostly affecting pediatric population Head and neck represent the usual locations.

We report a rare case of pilomatrixoma in an adult patient located on the face treated surgically without recurrence, different diagnostic and therapeutic aspects of this pathology will be recalled.

Introduction

Pilomatrixoma, calcifying epithelioma of Malherbe, is an adnexal skin tumor developed at the expense of the hair matrix [1]. This is the most common of the hair follicle tumors most commonly seen in children; it is often unknown and confused with other skin lesions, the most common locations are the head and neck.

Treatment is surgical, complete excision to obtain a very low recidivism rate.

We report a rare form of facial pilomatrixoma in an adult treated surgically with a good outcome.

Case report

TA 46-year-old patient with no specific history of pathology who is seeing for swelling in the right temporal-zygomatic region , evolving for eight months, clinical examination finds a nodular swelling of 3cm/2cm in diameter, hard, painless, adherent to the skin but mobile in relation to the deep plane, The skin in sight is normal looking. (Figure 1)

A soft tissue ultrasound showed a heterogeneous hypoechogenic lesion with vascularization and internal calcifications measuring 29 mm long axis, suggesting a pilomatrixoma first. The patient undergoes a total resection of the tumor under local anesthesia (Figure 2)

The tumor nodule was encapsulated, indurate, measuring two and a half centimeters long axis (Figure 3).

Histological analysis had confirmed the diagnosis of pilomatrixoma over the presence of mummified cell islets whose cytoplasmic limits are clearly visible with focal calcifications, focal keratinizations are also observed. The evolution was favorable, with an absence of recurrence after 1 year of regular control.

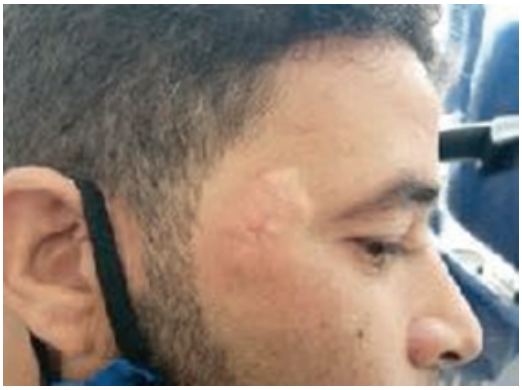

Figure 1. *Temporo-zygomatic localization of pilomatrixoma.*

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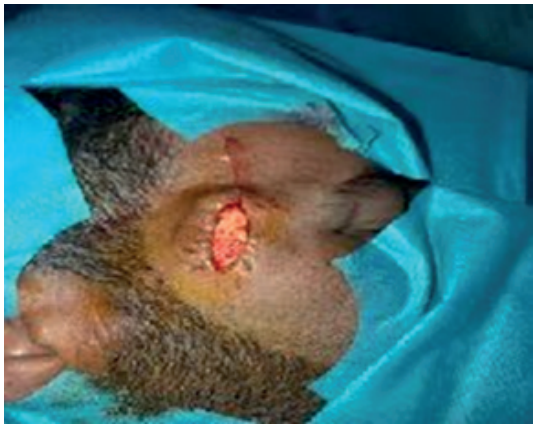

Figure 2. *Operational Aspect*

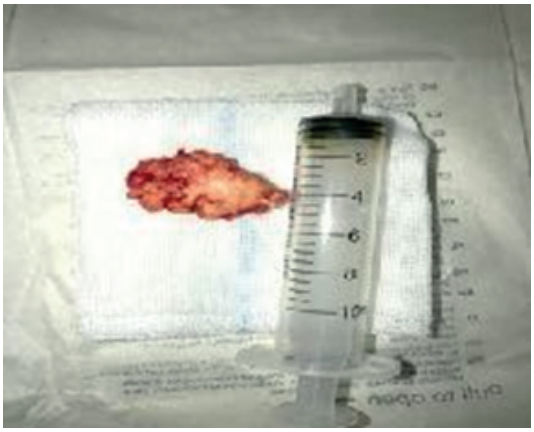

Figure 3. *Pilomatrixoma Total Excision Piece*

Discussion

Pilomatrixoma, or calcifying epithelioma of Malherbe, is a benign adnexal skin tumor developed from the elements of the hair matrix.

It is a rare tumor (0.12% of skin tumors), mainly affects children before the age of 10[2], Pulvermacker et al reported a large exclusively pediatric series of 89 operated pilomatrixomas [3].

It is more common in the female sex with an average sex ratio of 1.5 female patients for one male, and which is located in the upper part of the human body in more than one in two cases. [1].

Clinically, it is a subcutaneous nodule, unique, asymptomatic, firm (of stony consistency), round or oval shape, adherent to the cutaneous plane but not the deep planes making it thus mobilizable, the skin in sight is often bluish. There is no satellite lymphadenopathy. There may be the evocative sign «of the tent», described by Graham and Merwin, which, when the skin is placed in tension by the underlying tumor, it possible to observe a non- smooth surface made up of multiple facets and angles reflecting the amount of calcium deposited in the tumor [3], Other clinical forms can be observed and be perforated, ulcerated, anetodermal with erythematous skin next to the lesion or pigmented [4] which explains the diagnostic errors found in the literature; in our case the skin looked normal.

Pilomatrixoma is usually a single tumor. However, some patients develop, simultaneously or successively, several pilomatrixomas [5]. It is currently recognized that the occurrence of pilomatrixoma is more common in patients with Steinert myotonic dystrophy; the disease is often multiple and family forms are more frequent. Similarly, in Gardner's syndrome associating rectocolic polyposis with extradigestive signs, the occurrence of pilomatrixoma is also more frequent [6].

Paraclinical examinations are not specific but sometimes they differentiate it from many other differential diagnoses such as squamous and pilar cyst, subcutaneous fibroma, subcutaneous calcinosis but especially malignant pilomatrixoma or trichomatricial carcinoma with significant aggressive potential [7].

This is how the ultrasound shows a very limited subcutaneous mass with a «target» aspect: an echo centre and a hypoechogenic fine circumference; the existence of a posterior shadow cone will reflect the presence of calcification.

Magnetic resonance and computed tomography provide no additional element [8].

The diagnosis of pilomatrixoma must remain clinical and be confirmed histologically by showing very limited cellular islets are circumscribed by an individualized capsule. The architecture is composed on the periphery of basophilic cells, in the center of a clear area made of characteristic cells called «mummified» (without nucleus, with abundant eosinophilic cytoplasm, calcifications) separated by an inflammatory transition zone with giant cells. [7]

The treatment of pilomatrixomas is surgical, allowing the diagnosis of certainty and treating the patient. [9]

This resection must be done in a healthy area with safety margins varying from 5 to 10mm, often requiring removal of the superficial skin tissue. With well-performed surgery (R0), the recurrence rate is low at around 1.5% [10].

Conclusion

Pilomatrixoma is a benign and rare tumor in adults.

The cervical-facial seat and the female sex are the usual characteristics; his diagnosis is often unknown in preoperative because of his great polymorphism clinical, confirmation is histological and treatment is surgical.

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