Pilomatrixoma of the scalp. Case report and review of the literature.

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Case Report

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BACKGROUND. Pilomatrixoma, also known as Malherbe calcifying epithelioma, is a superficial benign skin tumor arising from the matrix cells of the hair follicle. Although these types of lesions are documented in the literature, they are often misdiagnosed as other skin conditions. We present a clinical case with initial suspicion of hemangioma and confirmatory histopathologic diagnosis of pilomatrixoma.

KEY WORDS: Pilomatrixoma, Malherbe calcifying epithelioma, head and neck surgery.

Introduction

The pilomatrixoma or calcified epithelioma of Malherbe is a benign cutaneous neoplasm of hair follicular origin with a differentiation towards the cells of the capillary cortex, occurs more frequently in children and adolescents, with a slight predominance in the female sex, Characteristically it manifests as a cutaneous lesion, firm and solitary, slow growing, for months or years before diagnosis, there are imaging characteristics that can help with the diagnosis, the definitive treatment is surgical excision with low rates of recurrence of the lesion.

Case report

18-year-old male patient, without comorbidities, without heredofamilial or toxicological history, refers clinical picture of 2 years of evolution of lesion apparently of nodular aspect of easy bleeding that initially remitted and reappears at 9 months with a new lesion and bleeding that at 3 months, A simple and contrasted cranial tomography was performed with evidence of a soft tissue tumor in the right frontal region without bone or parenchymal involvement, probably a cutaneous hemangioma (Figure 1). At the dermatologic physical examination in the right parietal region, a neoformation measuring 7 x 6 cm in diameter, with defined borders, 5 cm high, with loss of adnexa, smooth, slightly reddish, in areas of semi-soft consistency and other indurated areas (figure 2). Surgical procedure was performed under general anesthesia, excision of the tumor en bloc to the pericranium, hemostasis and rotation of the semilunar flap (Figure 3). The histopathological report presents findings compatible with totally resected Pilomatrixoma (Malherbe calcifying epithelioma), without evidence of malignancy.

Discussion

Pilomatrixoma or calcified epithelioma of Malherbe is an uncommon benign cutaneous tumor of hair follicular origin with a differentiation towards hair cortex cells first described in 1880 (1). In 1961, the term pilomatrixoma was proposed to emphasize the origin of the lesion (2). (2).A mutation in exon 3 of the β-catenin gene (CTNNB1) has been identified in most human and murine pilomatrixomas. Beta-catenin is integral to the terminal differentiation of the hair follicle during embryogenesis. (3) 60% of the lesions occur in patients younger than 20 years; however in the literature 2 peaks of incidence have been reported, the first during the first 20 years of life and the second smaller peak in individuals aged 50 to 60 years.(4) There is a slight female predominance, with a female to male ratio of 3:2 (5).

It is defined as an unusual, slow-growing, benign neoplasm, present for periods of months or years before diagnosis. It manifests as a firm cutaneous lesion located on the upper extremities, head and neck as the most common sites. (64%) followed by upper limbs (22%), trunk (8%), lower extremities (5%), and other sites (1%). (6) The right side seems to be more predominant than the left, usually ranging in size from 0.5 to 3.2 cm in diameter, (7) Typically it usually appears as a solitary mass, although in some syndromes such as Gardner, Turner, Rubinstein-Taybi or Churg-Strauss in addition to xeroderma pigmentosum or sarcoidosis the lesions may present in multiple (8).

Clinically the lesions usually present bluish-red discoloration in the area overlying the skin, due to dilation of the blood vessels and can be confused with hemangiomas as in our case. (9) Other differential diagnoses include lymphadenitis, sebaceous, dermoid and epidermoid cysts, atypical mycobacterial infections and malignant lesions such as squamous cell

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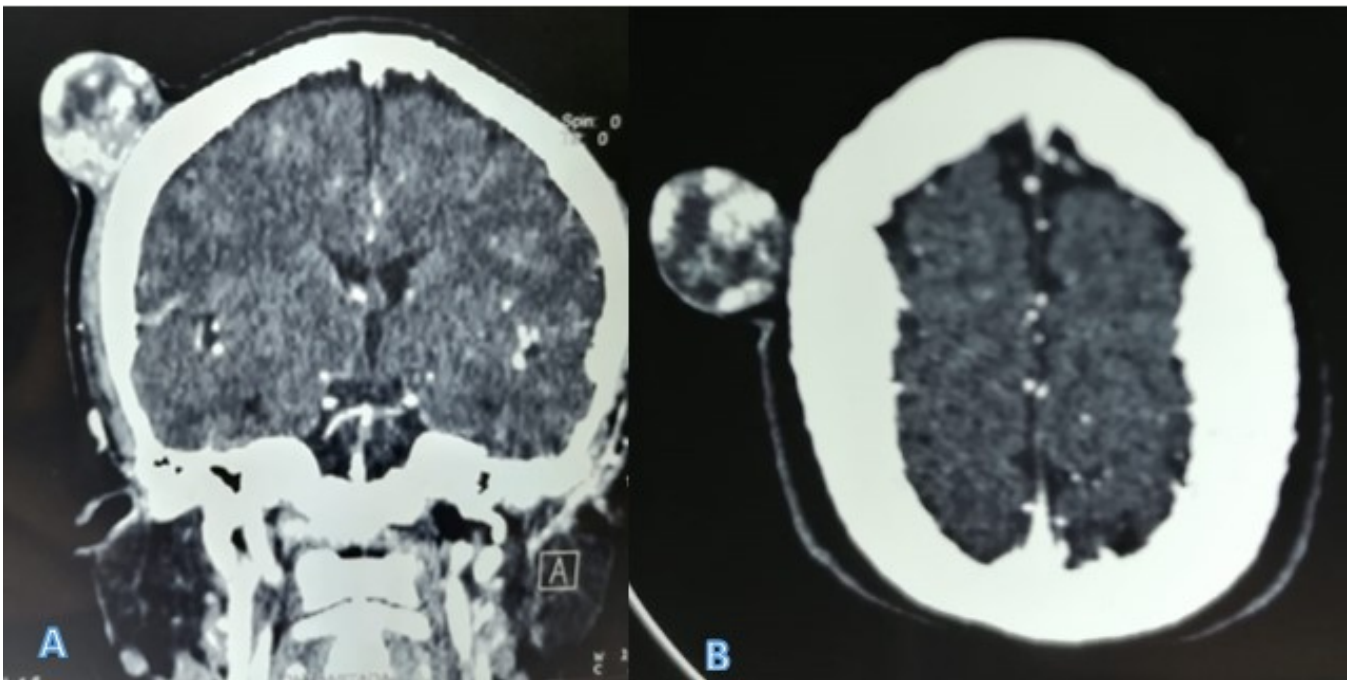

Figure 1. Contrast-enhanced tomography of the skull A. coronal reformatting B. axial section with a nodular image, with well-defined regular borders, located in the scalp towards the right parietal region, with marked enhancement to the contrast medium

arcinoma, matrix carcinoma, basal cell carcinoma and malignant melanoma.

With respect to the imaging diagnosis, ultrasound should be the first imaging study to be performed. Characteristic findings include a nodular lesion with a hypoechoic border (82.8%), as well as internal reticulations and calcifications. (10) Radiography can help to identify calcifications, but the sensitivity for pilomatrixoma is low. CT is used more frequently and describes calcifications as well as subcutaneous fat around the lesion usually intact without strands or edema (11) of similar characteristics MRI shows a clearly demarcated mass of soft tissue density containing microcalcifications. The treatment of choice for pilomatrixomas is surgical excision with histologic confirmation. Most studies report that complete surgical excision with clear margins is almost always curative. (12)

Conclusion

Pilomatrixoma is a common benign subcutaneous neoplasm in the first decades of life with origin from the adnexa, however sometimes the clinical and imaging characteristics condition misdiagnosis, the importance of proper recognition of this pathology allows treatment in a timely manner.

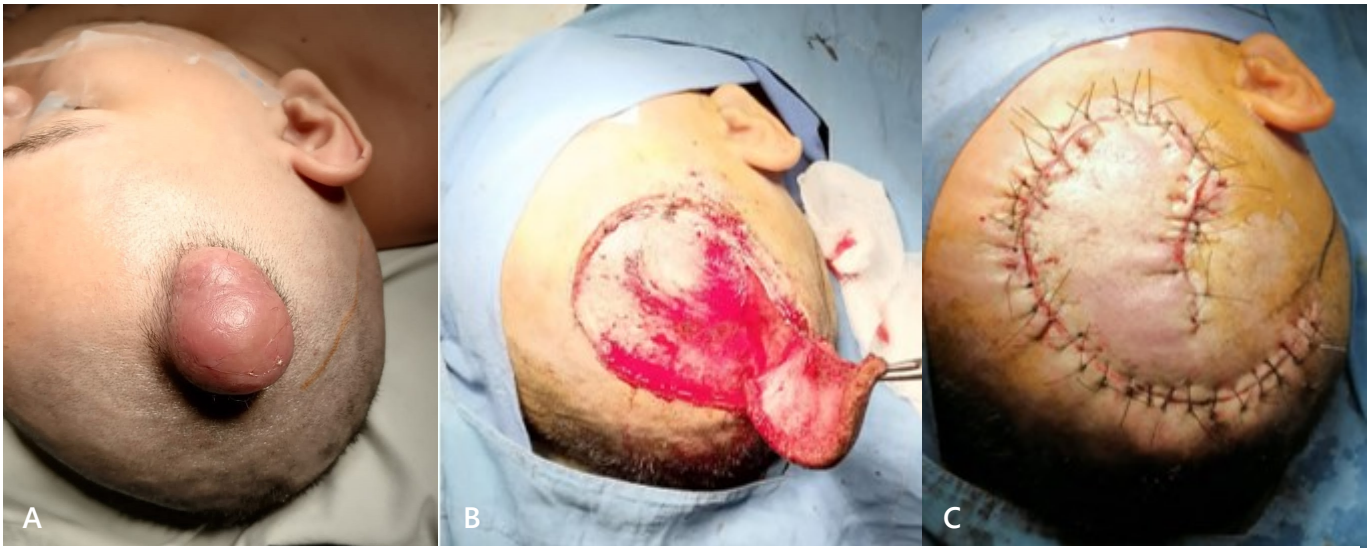

Figure 2. A. Neoformation of the scalp at the right parietal level. C and D. surgical excision of the lesion and creation of a semilunar flap.

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Conflicts of interests

There was no conflict of interest during the study, and it was not funded by any organization.

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