AN UNUSUAL PRESENTATION OF A PILOMATRIXOMA IN THE EYELID

PRESENTACIÓN INUSUAL EN EL PÁRPADO DE PILOMATRICOMA

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ABSTRACT

Case report: A 32-year-old man developed a lesion, over a period of 1 year, at the margin of the right upper eyelid. The presumed diagnosis was a pyogenic granuloma. An excision biopsy was performed and the histopathologic diagnosis was a pilomatrixoma.

Discussion: Eyelid pilomatrixoma is commonly misdiagnosed preoperatively and is extremely rare in middle-aged patients (Arch Soc Esp Oftalmol 2006; 81: 483-486).

Key words: Eyelid neoplasm, pilomatrixoma, calcifying epithelioma, free-edge eyelid, pyogenic granuloma.

INTRODUCTION

Pilomatrixoma, also known as Malherbe’s calcifying epithelioma, is a benign tumor which occurs frequently in pediatric age. It originates in the hair follicle matrix (1). It can appear in any part of the skin comprising hair follicles although it occurs more frequently in the head, face, neck and upper limbs (2). Malherbe’s epithelioma generally appears as a unique lesion, and its association with Steinert’s myotonic distrophi has been described. This case concerns an adult with pilomatrixoma on the free palpebral edge (a location previously not described) which clinically simulated a pyogene granuloma.

RESUMEN

Caso clínico: Hombre de 32 años de edad que presenta una lesión en borde libre palpebral de párpado superior derecho de un año de evolución, con un diagnóstico clínico de granuloma piógeno. Se realiza su escisión quirúrgica y el diagnóstico histopatológico es de pilomatricoma.

Discusión: El pilomatricoma de localización palpebral es generalmente mal diagnosticado preoperatoriamente y es extremadamente raro en pacientes de edad media.

Palabras clave: Tumor palpebral, pilomatricoma, epitelioma calcificante, borde libre palpebral, granuloma piógeno.
CASE REPORT

A 32-year old male referred to our service due to a pediculated lesion with clinical diagnostic of pyogenic granuloma with a height of 1 cm and 0.6 cm diameter at the base, reddish with vascular ingurgitation, localized in the free edge of the medial third of the upper right eyelid (fig. 1), without pain or bleeding and one year evolution. The rest of the ophthalmological exploraiton yielded normal results.

The epithelioma was surgically excised by means of a total thickness pentagon comprising the lesion.

The histopathological diagnostic was pilomatrixoma with description of a solid, encapsulated mass constituted by a mass of pilomatractical cells arranged in the periphery, large keratine masses and isolated areas of phantom cells, with granulomatous reaction to an adjacent foreign body (fig. 2). Additional zoom (fig. 3) reveals small basaloid cells without cytological abnormalities. In the central area cells are bigger, with eosinophile cytoplasm and loss of nucleus (phantom cells). Between these two predominant cell types we found transitional cells. After 4 years follow-up no relapse or malignization has been observed.

DISCUSSION

Pilomatrixomae are ectodermic tumors originated in the outermost cells of the sheath of the hair follicle root (1) which have been described in eyelids and brows for over 40 years (3). They can appear at any age, with greater prevalence in the first and sixth decade of life, with very little frequency in youngsters and adults, and more predominant in men than women (2,4).

The appearance of pilomatrixoma in patients with myotonic dystrophy is more frequent than in the general population.

However, in general pilomatrixoma is not hereditary. The pathogenic mechanism of its development
is associated to mutations in the betacatenine gene (CTNNB1) and it has been confirmed that this mutation does not only occur in pilomatrixoma but in hair follicle carcinomas too, directly involving the betacatenine dysfunction as the main cause of tumor growth in the hair follicle (5).

The location of 94% of cases is the scalp, face, neck and upper limbs as well as the preorbitary region (upper eyelid and brow), which is involved in 21% of cases (4). It has been suggested that the distribution of pilomatrixoma matches the density of hair follicles in a given area.

The presence of pilomatrixoma in the free eyelid edge has not been described previously. The differential diagnostic must be made generally with an epidermic or dermoid cyst as well as with a basal cell chalacin, hemangioma, lipome and carcinoma, even though the correct diagnostic occurs between 21 and 50% of cases (2,4).

Histopathologically, pilomatrixoma is characterised by a mass made up by basaloid cells, ghost cells and calcification and sometimes ossification. The ghost cells represent necrosis areas of previously vital basaloid cells. The calcification and ossification areas appear progressively in the necrosis areas. There is also an increase of small sized vessels and the overlying dermis and epidermis are atrophic.

In general, the clinical development is benign, although malign transformations have been described (4). Treatment is surgical excision and, if done adequately, recurrence is exceptional.

REFERENCES
1. Ni C, Kimball GP, Craft JL, Wang WJ, Chong CS, Albert DM. Calcifying epithelioma: a clinicopathological analysis of 67 cases with ultrastructural study of 2 cases. Int Ophthalmol Clin 1982; 22: 63-86.
2. Pirouzmanesh A, Reinisch JF, Gonzalez-Gomez I, Smith EM, Meara JG. Pilomatrixoma: a review of 346 cases. Plast Reconstr Surg 2003; 112: 1784-1789.
3. Boniuk M, Zimmerman LE. Pilomatrixoma (benign calcifying epithelioma) of the eyelids and eyebrow. Arch Ophthalmol 1963; 70: 399-406.
4. Julian CG, Bowers PW. A clinical review of 209 pilomatrixomas. J Am Acad Dermatol 1998; 39: 191-195.
5. Hassanein AM, Glanz SM. Beta-catenin expression in bening and malignant pilomatrix neoplasms. Br J Dermatol 2004; 150: 511-516.