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Palpebral swelling: dermoid cyst or not?

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Summary:
Pilomatrixoma, also known as Malherbe's calcified epithelioma, is a rare benign adnexal skin tumor. We report the case of a 25-year-old woman with a pilomatrixoma on the left upper eyelid mimicking the clinical features of a dermoid cyst.
The patient was 25 years old and had no previous medical history. She came with a cystic swelling of the left upper eyelid that appeared three years ago and was progressively increasing in size. Her visual acuity was at 10/10. Ophthalmological examination revealed a well-limited tumor located at the tail of the eyebrow. A dermoid cyst was suspected because the patient wasn't sure that it appeared 3 years ago. The tumor was superficial and mobile in relation to the deep planes, so we opted for surgery without further investigation. Surgical excision took place under local anaesthesia via a small incision. Pathology revealed a pilomatrixoma of the left upper eyelid.
Pilomatrixomas are benign tumors often found in the first 2 decades of life with the medial canthus as the preferred location. Often misdiagnosed clinically, the correct diagnosis is only established after excision and histological examination. The pathological diagnosis is based on the discovery of large masses of ghost cells, associated with basophilic cells, inflammation, foreign body giant cells, calcification and ossification.
Pilomatrixomas are benign tumors that should be considered for diagnosis prior to surgical removal in children, especially when the tumor is indurated but mobile, chalky in appearance and located in the medial canthus.

Résumé :
Le pilomatricome, également appelé épithélioma calcifié de Malherbe est une tumeur cutanée annexielle bénigne rare. Nous rapportons le cas d'une jeune femme de 25 ans présentant un pilomatrixome au niveau de la paupière supérieure gauche mimant les caractéristiques cliniques d'un kyste dermoïde.
Il s'agit d'une patiente de 25 ans, sans antécédents pathologiques particuliers. Elle s'est présentée à la consultation pour une tuméfaction kystique de la paupière supérieure gauche apparue il y a trois ans et augmentant progressivement de volume. L'acuité visuelle était à 10/10 . L'examen ophtalmologique a révélé une tumeur bien limitée, située au niveau de la queue du sourcil. Un kyste dermoïde, malgré l'apparition à l'âge adulte selon la patiente, a été évoqué. La tumeur était superficielle très mobile par rapport aux plans profonds, nous avons donc opté pour une chirurgie sans avoir recours à d'autres explorations. L'excision chirurgicale a eu lieu sous anesthésie locale via une petite incision. L'anatomopathologie a objectivé un pilomatrixome de la paupière supérieure gauche.
Les pilomatrixomes sont des tumeurs bénignes retrouvées souvent dans les 2 premières décades avec comme localisation préférentielle le canthus médial. Souvent mal diagnostiqué cliniquement, le diagnostic correct n'est établi qu'après excision et examen histologique. Le diagnostic pathologique repose sur la découverte de grandes masses de cellules fantômes, associées à des cellules basophiles, une inflammation, des cellules géantes à corps étranger, une calcification et une ossification.
Les pilomatrixomes sont des tumeurs bénignes dont il faut envisager le diagnostic avant une exérèse chirurgicale chez un enfant, surtout lorsqu'il s'agit d'une tumeur indurée mais mobile, d'aspect crayeux située au niveau du canthus médial.

I. INTRODUCTION

Named by Forbis and Helwig in 1961, pilomatrixoma, a benign tumour of the hair follicle, was first described by Malherb in 1880 as a "calcifying epithelioma" and was thought to originate from the sebaceous gland.(1) It is an adnexal skin tumour arising from the matrix cells at the base of a hair and is often mistaken for a dermoid cyst. (2)

There are few reports in the literature of eyelid pilomatrixomas. It can occur in men and women of all ages. However, it is most common in the first two decades of life with a female preponderance. It has been

reported as one of the most common skin appendage tumors in patients aged 20 years or younger, accounting for approximately 75% of cases.(3) (4)

Pilomatrixoma can occur in most hair-bearing parts of the body, but has a predilection for the skin of the head and neck. It is considered an uncommon lesion of periocular tissues, (4) and presents as painless subcutaneous nodules that can be mistaken for a dermoid cyst. (5) (3)

The diagnosis of pilomatrixoma is usually not suspected clinically due to its similarity to a variety of cystic lesions, (6) (7) and the correct diagnosis can only be made after histological study. (8)

Clinically, the lesion presents as a slow-growing, irregularly shaped nodular, mass. The skin usually has a reddish to blue colour due to dilated blood vessels and chalky white nodules may be visible through the skin. There is usually no history of inflammation or trauma. (9) (1)

The exact pathogenesis is still unknown but is thought to be related to molecular genetic mutations in the Wnt signalling pathway in basophilic and umbilical cells. (10)

We report an unusual appearance of a pilomatrixoma of the eyelid mimicking the clinical features of a dermoid cyst.

II. OBSERVATION :

Mrs. D.R., 25 years old, with no previous pathological history, presented with a cystic swelling of the left upper eyelid that appeared three years ago and was progressively increasing in size. The visual acuity was 10/10. Clinical and ophthalmological examination was normal. The tumor was well limited, located in the tail of the eyebrow. (fig.1) We therefore thought it was a dermoid cyst despite the fact that the patient thought it had appeared in adulthood. The tumor was superficial and very mobile in relation to the deep planes, so we opted for surgery without the use of CT. Surgical excision took place under local anaesthesia via a small incision. We removed a tumor measuring 14 mm by 5 mm, bilobed and fleshy, with limited contours and a consistency very different from dermoid cysts. (Fig. 2) Pathology revealed fibrous tissue containing clumps and masses of squamous cells, often mummified or calcified. These clumps are surrounded by a granulomatous reaction rich in giant cells. This was a pilomatrixoma of the left upper eyelid.

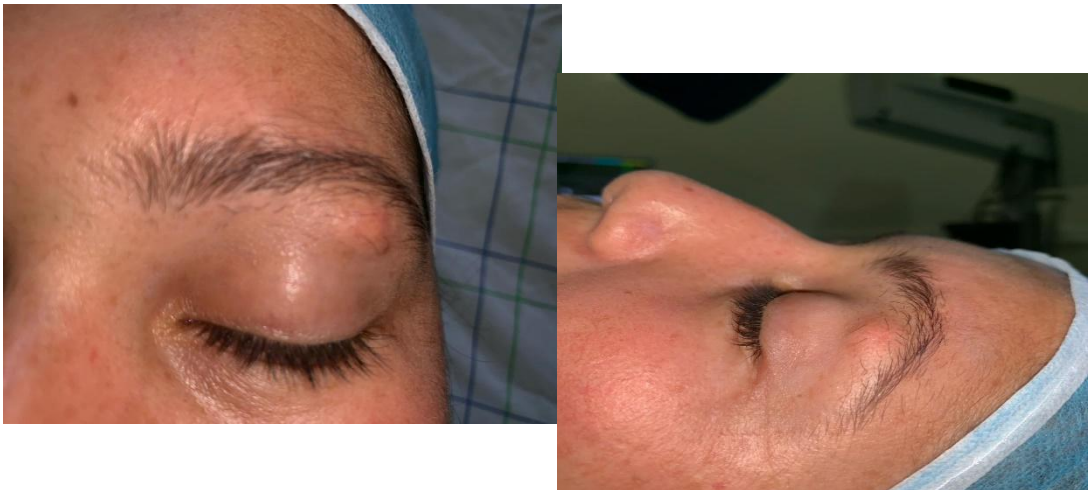

Figure 1: Eyelid mass in a 25 years old woman

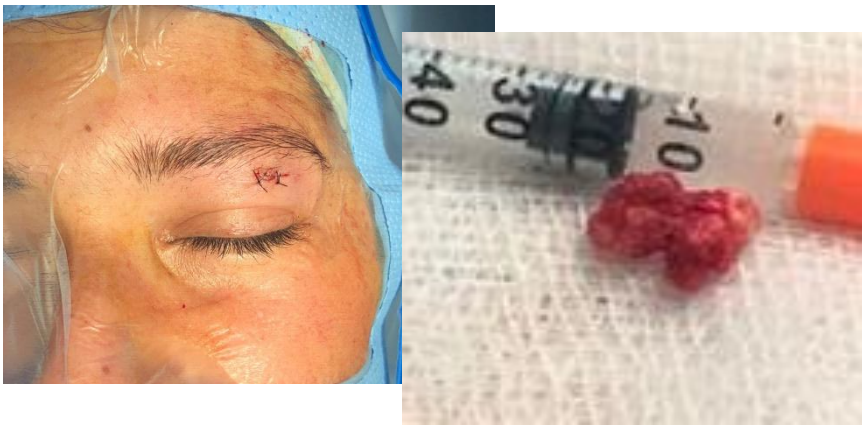

Figure 2 : Pilomatrixoma after surgical excision

III. DISCUSSION

Pilomatrixoma is a relatively rare benign tumor arising from the hair root matrix.

It can occur almost anywhere in the body, but tends to develop in the cervical and facial region, often involving the eyebrow. (8) In contrast, pilomatrixoma of the eyelids is less common and often misdiagnosed clinically, as it is confused with cyst, chalazion, sebaceous carcinoma and other tumors. (11)

It is most common in the first two decades of life, and tends to affect young women. (12) (2) Among case reports and "case series" of periocular pilomatrixoma worldwide, the youngest patient we are aware of was a 1-year-old female. (4)

This type of tumor typically presents as a solitary, superficial, slowly growing, irregularly shaped, nodular and painless mass, easily mobilized from the subcutaneous tissue. The nodule size is variable (less than 3 cm) but never larger than a hazelnut and may be characterized by a stony hardness. However, a few cases of giant pilomatricomas have been reported. (13) It is usually covered by skin with red to blue coloration due to dilated blood vessels. Rarely, there is ulceration. Foreign-body giant cells, inflammatory cells, calcifications and hemorrhages may also be seen. (4) It is usually located near the lateral aspect of the eyebrow or upper eyelid, and is frequently first diagnosed as a dermoid cyst. Pilomatrixoma sometimes shows rapid growth and may resemble a keratoacanthoma. In some cases, trauma can play a definite role in enlarging an existing lesion by causing hemorrhage into the tumor. (14) There are rarely ulceration or drainage. (8)

Initially, the lesion is well limited and contains two distinct zones: a peripheral hyperbasophilic zone, composed of small basal-like epithelial cells; a central eosinophilic zone composed of lighter cells called shadow cells. The latter may contain calcium or even ossify, causing a giant cell granulomatous reaction.

There is histochemical evidence of a dysembryoplastic origin of the tumour from the primitive cells of the hair matrix. The term pilomatricoma should therefore be preferred to "calcification" as it is a benign lesion. (5)

In our patient case, pathology exam revealed fibrous tissue containing clumps and masses of squamous cells, often mummified or calcified. These clumps were surrounded by a granulomatous reaction rich in giant cells.

Pilomatrixoma should be differentiated from the malignant form of this tumor, pilomatrix carcinoma, which occurs more often in middle-aged and older individuals, more frequently in men than in women. Microscopically, it is characterized by an exuberant proliferation of basaloid cell masses arranged in a disordered fashion throughout the tumor, varying degrees of cytological atypia, frequent mitoses and areas of necrosis. Keratinization with ghost cell formation is less extensive than in benign pilomatrixomas. The infiltrative growth pattern that extends to deeper soft tissues can lead to vascular and perineural invasion. (1)

No specific preoperative examination is helpful in diagnosing this benign tumor. Imaging is often non-specific. (15) However, ultrasound is the most commonly reported modality for identifying pilomatrixoma, which shows well-defined, ovoid, hypoechoic heterogeneous masses with posterior shadowing and echogenic internal foci. (2) Hoffmann et al (16) were the first to suggest that MRI could be a diagnostic tool for pilomatrixoma. Recently, pilomatrixoma has been described as a well-defined mass with intermediate homogeneous signal intensity on T1-weighted images and high signal intensity on T2-weighted images. (17)

Core biopsy is preferable to fine-needle aspiration, as it is more likely to show the classic findings of calcification, foreign-body giant cell reaction, basaloid squamous cells and pathognomonic anucleate squamous cells ('ghost cells'). (15) However, as spontaneous remission has not been documented, the therapeutic approach requires surgical excision of the lesion with wide margins for histological diagnosis. (18) As the tumor may be

strongly adherent to the skin, it may be necessary to remove the overlying skin. However, the tumor never adheres to the subcutaneous tissue and separation from the underlying skin is always easy. Recently, good cosmetic results with endoscopic excision of pilomatrixomas of the forehead and eyebrow have been reported. (8) The prognosis is generally good. (13)

There is an estimated 3-4% risk of recurrence. (18) Incomplete resection leads to high recurrence rates. (2) Multiple pilomatrixomas or pilomatrixoma recurrences are rare and automatically raise suspicion of related disorders such as myotonic dystrophy, Churg-Strauss syndrome, Gardner syndrome, trisomy 9, Rubinstein-Taybi syndrome, xeroderma pigmentosum, and sarcoidosis. Familial occurrences are considered as a chance event or related to the syndromes mentioned above. (4)

IV. CONCLUSION

Pilomatrixoma is an uncommon tumor of the eyelid that is most common in children and adolescents. It is often misdiagnosed preoperatively, although there are clinical features of the lesion that can help clinicians differentiate it from other tumors seen in childhood. A pilomatrixoma should be considered in the differential diagnosis of a painless, firm to hard, nodular mass of the eyelid that progressively increases in size. Complete surgical excision is always curative and confirm the diagnosis.

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