

Calcifying epithelioma of Malherbe

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Abstract

Pilomatricoma is a common and benign cutaneous tumor of the hair follicle in children. It is an annexial tumor often unknown and confused with other cutaneous lesions. The usual locations are the head and the neck. The aim of this work is to report a series of 14 cases with unusual forms collected in the dermatology department of the CHU Hassan II of Fez over a period from December 2015 to April 2018. The study concerned 9 women and 5 men. The average age was 32 years (8-60 years). The cervico-facial localization was observed in 2 cases, the other localizations were represented by: The upper limbs (9 cases), the trunk (one case) and the scalp (2 cases). The clinical aspect was typical in all cases with subcutaneous nodules of stony consistency, ulcerated in three cases. The dermoscopic aspects described are: whitish structures, homogeneous erythematous zones, irregular linear vessels, point and hairpin vessels, blue-gray zones. All patients underwent nodule resection under local anesthesia. The histological study was in favor of a mummified epithelioma of Malherbe complete excision without signs of malignancy. No patients had a recurrence. The originality of our study lies in the presence of exceptional localizations in the limbs, the trunk and the scalp, the age of unusual occurrence in 5 patients between 15 and 60 years, the occurrence during pregnancy in a patient and the giant character arriving at 10cm in 1 case, and also in the description of the dermoscopic signs of this tumor.

Keywords

pilomatricoma; unusual forms; dermoscopy

Introduction

Pilomatricoma or calcifying epithelioma of Malherbe is a benign cutaneous tumor of the hair follicle. It is the most common adnexal tumor in children. It is an annexial tumor often unknown and confused with other cutaneous lesions. The usual locations are head and neck, the involvement of members remains exceptional.

Objective

The purpose of this work is to report a series of 14 cases with unusual shapes.

Methods

This is a descriptive study collecting all the cases of pilomatricomas confirmed on histological study of the operative specimen, followed in the dermatology department of the CHU Hassan II of Fez, between December 2015 and April 2018. We recorded for each patient epidemioclinical, histopathological data, treatment and evolution.

Results

Of the fourteen observations collected, nine were female (64%) and five male (sex ratio F / H: 1.8). The average age was 32 years old with extremes ranging from 8 to 60 years old. Cervicofacial localization was observed in two cases, in the internal canthus appeared in a pregnant woman (Figure 1) and in the cheek appeared in a 13-year-old girl (Figure 2), the other locations were represented by: the upper limbs (9 cases), (Figure 3, 4 and 5) the trunk (a case) (Figure 6) and the scalp (2 cases) (Figure 7). The evolution was on average 21 months (2 months-13 years). The clinical aspect was typical in all cases with subcutaneous nodules of stony consistency, ulcerated in three cases. The dermoscopy performed in eight patients revealed the appearance of white or yellowish-white areas (100%), a vascularization of irregular linear vessels type and point vessels (37.5%), of arborising vessels in crown on erythematous background and homogeneous red areas (25%) (Figure 1, 2, 3, 4, 6 and 7). All the patients had benefited from an excision of the nodules under local anesthesia, with histological confirmation. No patient had recurred with a mean follow-up of 12 months.

Discussion

Pilomatricoma was described by Malherbe Chenantais in 1880 as a benign, calcified tumor of the sebaceous glands, and its origin was confirmed later by Forbis and Helwing [1,2]. Indeed, these two authors demonstrated that the origin of the tumor was the cells of the hair matrix. Pilomatricoma typically occurs as an asymptomatic round or oval irregular subcutaneous nodule with a hard or firm consistency. The skin in view is often bluish. The tumor adheres to the superficial level, whereas it is mobile with respect to the deep levels. The sign of the tent described by Graham and Meruim is very evocative of the diagnosis [2]. Pilomatricoma can take different clinical forms and be perforating, ulcerated as in three of our patients, anestodermal with erythematous or pigmented skin [1,2]. This explains the diagnostic errors found in the literature. In most published cases, preoperative diagnosis is reported in only one third of cases [3]. The difficulty of the clinical diagnosis lies in the variable clinical appearance of the pilomatricoma and the ignorance of this tumor by certain clinicians. Some authors have tried to improve the tools of the clinical diagnosis of the pilomatricoma by the dermoscopy however this is not enough for the diagnosis of certainty [4]. The diagnosis of pilomatricoma should remain clinical, assisted by dermoscopy and confirmed by histology which eliminates some differential diagnoses mainly epidermoid and pilaris cysts

but especially the malignant pilomatricoma. The immunohistochemical study facilitates its distinction [3]. The carcinomatous degeneration of pilomatricoma remains controversial [5,6]. Some authors, however, have proposed additional imaging tests such as ultrasound to aid diagnosis. MRI has little interest in pediatrics [7,8]. Standard radiography is useful only in the presence of suspicion of a significantly calcified pilomatricoma. However in most cases pilomatricoma was diagnosed postoperatively. Most authors report the occurrence of pilomatricoma in children especially before ten years [2,9]. However it can appear at any age, congenital forms have also been reported [10]. Pilomatricoma is more common in women in the majority of published series with a sex ratio of 1.5 women for one man [11]. In our series the sex ratio was 1.8 women for a man. Regarding the different locations of pilomatricoma, our results do not agree with data from the literature with less frequent involvement of the cervicofacial region [12]. Only some exceptional localizations at the level of the members, were observed contrary to our series [7,13]. No morbid association is reported in our series. A combination of pilomatricoma with myotonic steinert dystrophy has been found with a predominance of multiple localizations and familial forms [3]. The treatment of pilomatricoma is complete surgical excision with a skin spindle, especially if the lesion is adherent to the dermis. This is the reference treatment to avoid the majority of recurrences [11]. The prognosis of the pilomatricoma is good. Healing without recurrence is the rule after total surgical excision.

Conclusion

The originality of our study lies in the presence of exceptional localizations in the limbs, the trunk and the scalp, the age of unusual occurrence in 5 patients between 15 and 60 years, the occurrence during pregnancy in a patient and the giant character arriving at 10cm in 1 case.

The dermoscopic aspects described are: whitish structures, homogeneous erythematous zones, irregular linear vessels, point and hairpin vessels, blue-gray zones. Dermoscopy could thus provide useful arguments for improving the diagnosis of pilomatricoma, but the confirmation is histological. Total surgical excision is the treatment of choice, allowing a cure without recurrence.

Authors contribution

All the authors contributed for the interpretation of data for the work; and drafting the work or revising it critically for important intellectual content; and the final approval of the version to be published; and the agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Figures

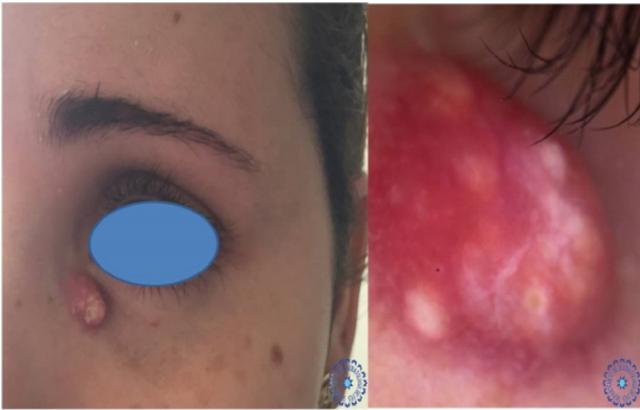


Figure 1: An internal canthus pilomatricoma and dermoscopy showing yellowish-white areas and vessels disposing in a crown



Figure 2: Cheek pilomatricoma and dermoscopy showing yellowish-white areas and arborizing vessels disposing in a crown

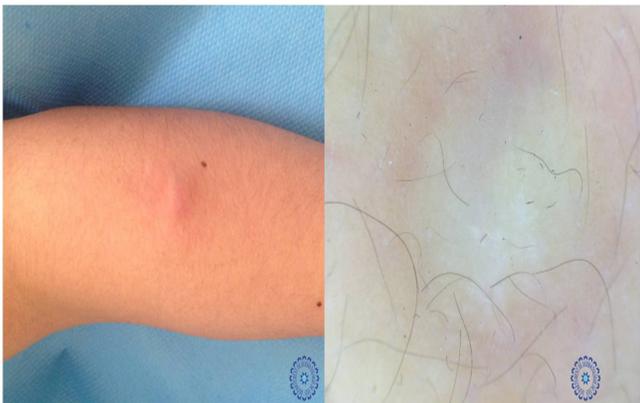


Figure 3: Pilomatricoma of the forearm and dermoscopy showing white-yellowish areas and whitish streaks



Figure 4: Ulcerated pilomatricoma of the arm and dermoscopy showing white-yellowish areas and irregular linear vessels



Figure 5: Pilomatricoma encapsulated in the arm during surgery



Figure 6: Truncal pilomatricoma and dermoscopy showing whitish areas and dotted vessels



Figure 7: Pilomatricoma of the scalp and dermoscopy showing whitish areas, homogeneous red areas and irregular linear vessels

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