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Periungual Pilomatrixoma – a Case with Very Rare Localization

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A 54-year-old woman with a slowly growing periungual tumor mass, pathologically verified as pilomatrixoma, is described. A multi-aspect overview of the clinical, diagnostic, and therapeutic features of this hair matrix-derived benign neoplasm are presented to accentuate the importance of a proper clinico-pathological approach and raise the suspicion index, even on such an anecdotally rare predilection.

Key-words: pilomatrixoma, periungual localization

Introduction

Pilomatrixomas are subcutaneous benign neoplasms of hair matrix origin, usually localized on face, neck, and upper extremities [15]. The versatile morphology and unusual clinical appearance, as well as the low prevalence, extremely deteriorates the diagnostic verification of the tumor, challenging even the most experienced clinicians.

Herein, a very rare periungual localization of pilomatrixoma is presented.

Case report

A 54-year-old Caucasean woman with a three-year history of slowly enlarging flesh coloured mass on the lateral margin of the left upper thumb is presented. She did not recall traumatic impact or any pre-existing nail abnormality. Keratolytic and anti-mycotic topical agents proved ineffective. Physical examination showed an oval tumor with firm consistency and a diameter of 7mm, with no change of the overlying skin. The presumptive clinical differential diagnosis included a myxoid cyst, ossifying hematoma, fibroxanthoma, glomangioma, and foreign-body granuloma. X-ray analysis showed microcalcifications in the subcutaneous mass. Surgical excision with histological examination was performed. The pathological picture revealed a well-circumscribed

nodulo-cystic tumor with islands of eosinophilic cells derived from basaloid proliferations **(Fig. 1)**, centrally forming anucleated ghost cells **(Fig. 2)** and zones of calcification. The diagnosis of pilomatrixoma was made. The patient was free of recurrences till the 7th month after surgery.

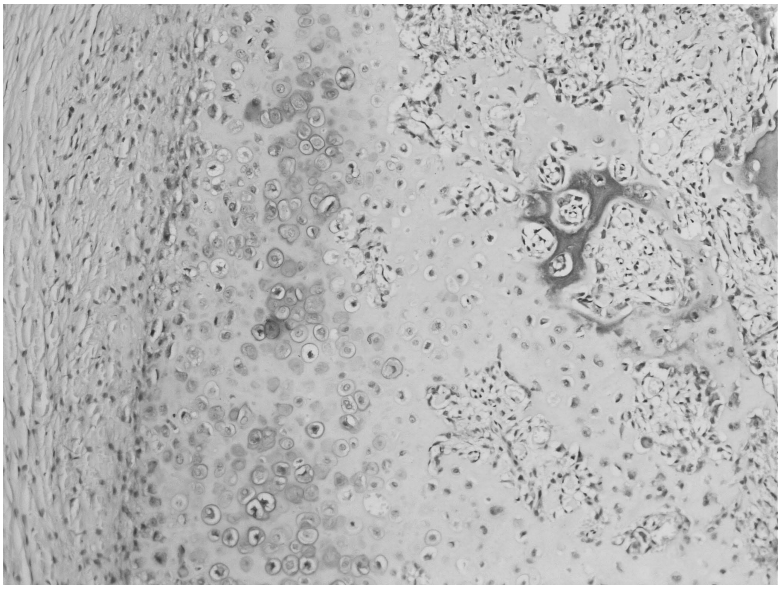

Fig. 1. Basaloid proliferations with central degeneration towards anucleated cells. HE x 100

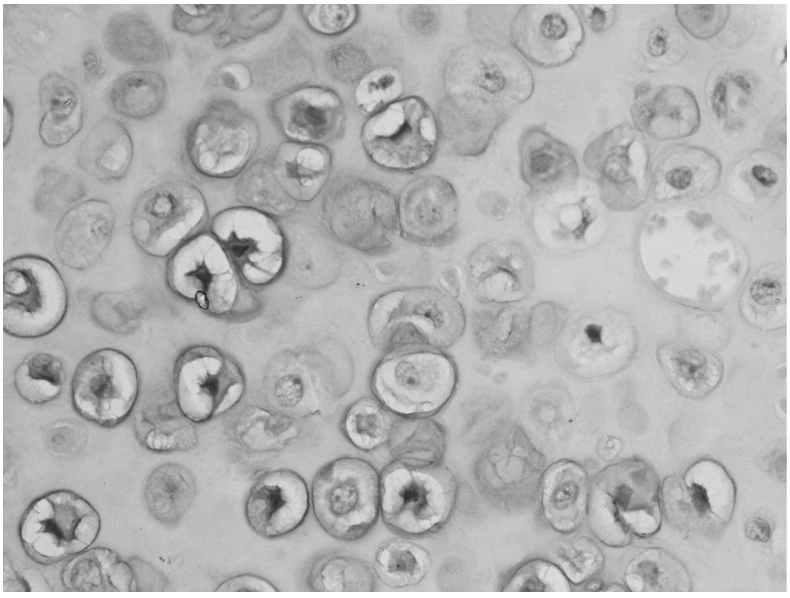

Fig. 2. Ghost cells. HE x 400

Discussion

Pilomatrixomas were first described by Malherbe and Chenantais in 1880 [10]. Although Jones and Campbell coined the term, when discovered a subcutaneous tumor in a pediatric population, it was later fully recognized that the tumor also affects adults [6]. Moreover, despite its low incidence in the elderly population, the only malignant transformations have been described only in this age group [8]. It seems that the pilomatrixoma carcinomas appear on head and neck of middle aged patients. The exact rate of malignant transformation is not clear. No more than 20 cases have been described worldwide, and it is still unknown whether this is a malignant case arising de novo, or represents a degeneration of pre-existing pilomatrixoma [7].

The diagnostic verification of the tumor is extremely challenging [9]. First, there are no specific clinical features. Usually a subcutaneous mass has been growing slowly for some years. The overlying skin can be unchanged, erythematous, slightly livid, or with yellowish discoloration [11]. The tumor is well-circumscribed and very firm on palpation.

The anatomic localization is usually on the head and neck region, less commonly - on the upper extremities [14]. To our knowledge, no periungual localization has been described to date. It seems rather unusual since no hair follicles are presented in close proximity to the nail bed. The differential diagnosis of tumors, arising around the nail apparatus, includes periungual fibroma, pyogenic granuloma, superficial acralfibroxanthoma, periungual melanoma and squamous cell carcinoma [12]. Some inflammatory reaction patterns can also show such localization - periungual warts, foreign body granuloma, residual haemangiomas [13]. The case presented showed an extremely rare variant of periungual tumor, probably arising from an ectopic stem follicle in conjunction with the nail martix. Such a predilection is exceptional, and makes this observation highly anecdotal and extremely difficult for diagnosis.

Imaging diagnosis has proven not to be highly specific. Conventional X-ray, shows milliar micro-calcifications, which can be seen in all cases of dystrophic calcification [5]. Ultrasound is not very discriminating, either. Heterogeneous echotexture, internal echogenic foci in scattered-dot pattern, and posterior shadowing with hypoechoic rim are not considered informative, since they can be seen in any long-standing subcutaneous tumors [1,4]. Sebaceous cysts, foreign body reactions and metastatic bone formations share the same computer-tomographic features as pilomatrixomas – uniform, homogenous on T1 weighted signal with varying results on T1 with contrast and T2 imaging [16].

The golden standard for reliable diagnostic verification remains pathological evaluation 3. It is defined by monomorphous basaloid proliferations with degradation towards amorphic ghost (shadow) cells representing hair cortex differentiation. The foci of calcification originate secondarily due to dystrophic calcium deposition [2].

The proper therapeutic approach acquires surgical excision. A low recurrence rate of 2.6% is expected, and radical excision margins of 2 cm are, therefore, approved. Patients' population that has been follow-up showed free of recurrence interval of 3 to 37 months [17]. Re-appearance of the tumor raises a suspicion of malignancy evoking aggressive surgical excision, which is supposed to be sufficient and curative treatment with excellent prognosis.

Conclusions

The periungual case presented here is a rare example of pilomatrixoma with unusual localization, unpredictable clinical evolution and extremely difficult surgical access. It

demonstrates the importance of complex clinico-pathological correlation and surgical skills, needed to precisely cure, monitor, and prevent such anecdotal hair matrix proliferation.

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