Case report

Calcifying epithelioma of Malherbe – a rare localization

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Calcifying epithelioma of Malherbe, also known as pilomatricoma or pilomatrixoma, mostly arises in the matrix hair follicle. It generally affects the head and neck, upper extremities, and trunk, with the lower extremities being a rare exception. We hereby present a case of a 31-year-old male patient who presented with a small, firm, subcutaneous mass over the left malleolus, which was provisionally diagnosed as lipoma. Surgical excision was performed, and the histopathology report revealed it to be pilomatricoma of the left malleolus.

Key words: hair matrix, Malherbe, pilomatricoma.

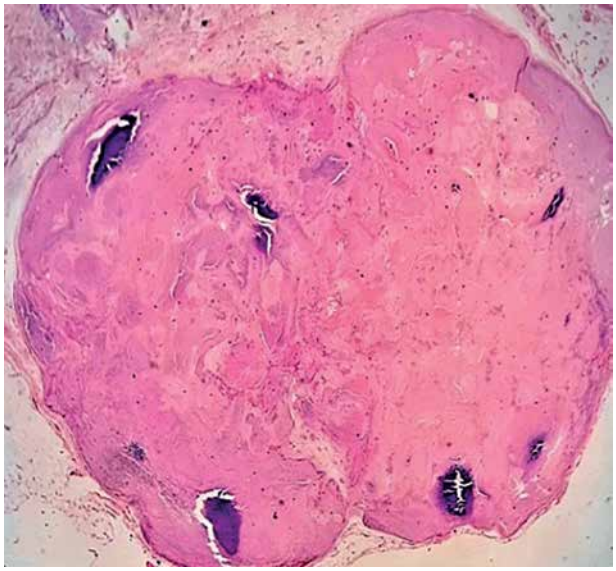
Introduction

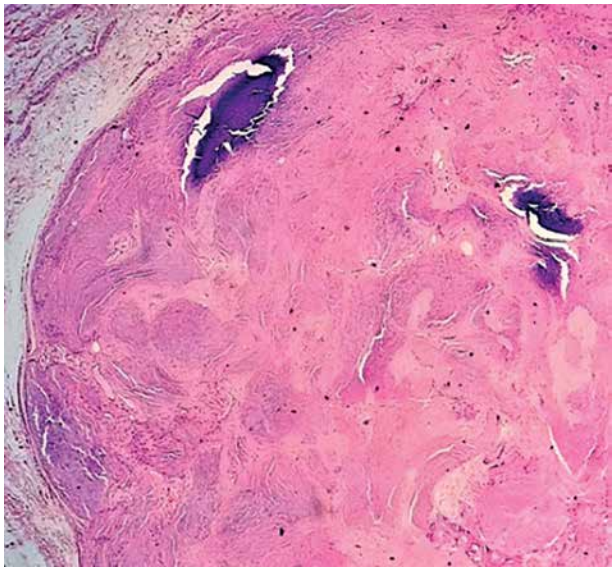
A benign appendageal tumour called a pilomatricoma develops from cells in the matrix of the hair follicle [1]. It was initially described by Malherbe and Chenantais in 1880; hence the term Malherbe's calcified epithelioma. The term "pilomatricoma" has been used since 1977 to describe benign ectodermal tumours that start in the germinal matrix of hair follicles [2]. It is frequently encountered in the paediatric age group and particularly affects women with a sex ratio of 1.5 [3]. It mainly represents as a solitary, firm, and subcutaneous mass with an overall incidence rate of 1 in 800 cutaneous tumour [4]. Many cases have been documented regarding the tumour arising from a common location such as the face (preauricular, periorbital), neck, upper limbs, and trunk. Rare localizations such as torso and lower extremities are less frequently reported [1]. Certain characteristic clinical features of the tumour imply a clinical diagnosis followed by histopathologic validation, because it is not frequently recognized prior to surgery. The purpose of this case report is to highlight an extremely unusual location of pilomatricoma in the lower extremities.

Case presentation

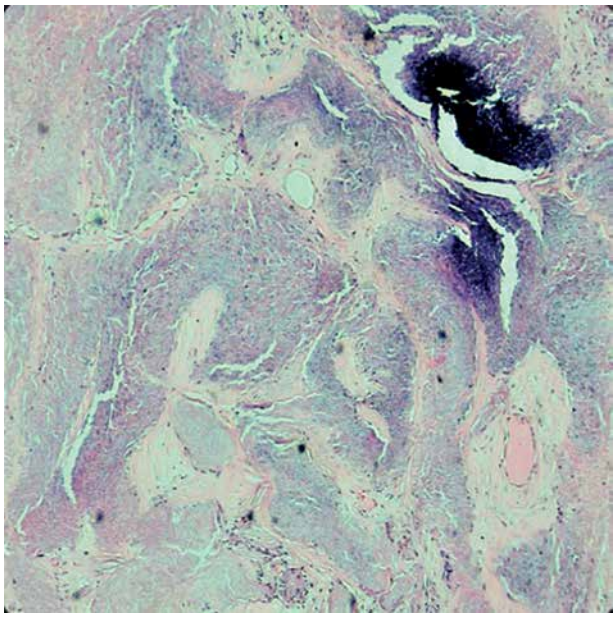
A 31-year-old male patient without substantial medical history came with complaints of a mass in his left malleolus of the left foot for the last 2 years. The onset was subtle and was gradually increasing in size. It was a solitary, skin coloured, firm mass, which was not associated with pain or fever. There was no history of trauma, weight loss, or previous inflammatory disease. On examination, a single, 2 × 2 cm, firm, well-circumscribed, non-tender, freely mobile mass with well-defined borders was present on the left malleolus of the left foot. Regional lymphadenopathy was absent. Superficial ultrasonography showed a well-defined, round to oval, heterogeneously iso- to hyperechoic lesion of size 13 × 12 × 16 mm in the subcutaneous plane with no vascularity on colour Doppler suggestive of lipoma. Surgical removal of the mass under local anaesthesia was performed, and it was sent for histopathological examination.

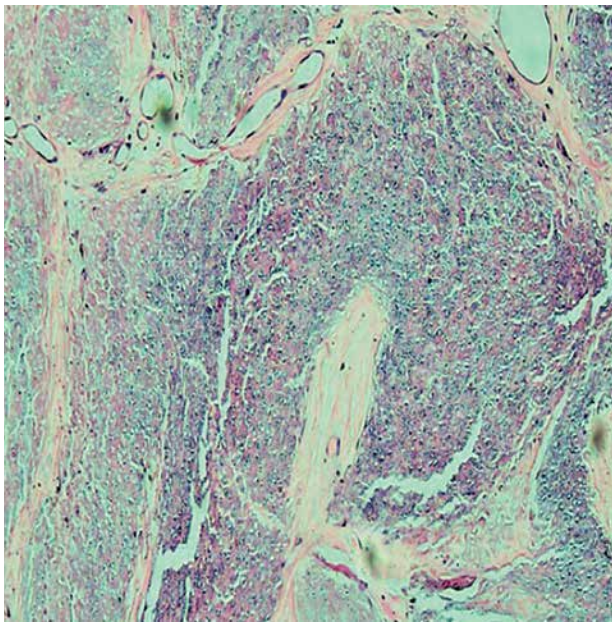
Gross examination of the specimen revealed a well circumscribed, grey, brown tissue piece along with skin flap throughout measuring 3 × 2 × 1.5 cm. Externally, a cyst like area was noted measuring 8 mm, and on cutting open 1 ml of thick reddish fluid oozed out. The sections were embedded in paraffin to form tissue blocks. Four-micron-thick sections were cut with the help of a microtome and stained by haematoxylin and eosin stain. Microscopic examination revealed stratified squamous lining epithelium. Subepithelium showed a solid nest of basaloid cells undergoing abrupt trichilemmal-type keratinization (Figs. 1, 2). Numerous ghost cells were noted, which





Figs. 1, 2. Photomicrograph showing stratified squamous lining epithelium. Subepithelium shows solid nest of basaloid cells undergoing abrupt trichilemmal-type keratinization with areas of calcification (HE, 40×]





Figs. 3, 4. Photomicrograph showing numerous ghost cells which hold their cell and nuclear borders (HE, 100×)

characteristically held their cell and nuclear borders; however, the nuclei lose their basophilic staining leaving a "ghost like" remnant (Figs. 3, 4). Dysplasia or malignancy was not seen. All these features pointed to a diagnosis of pilomatricoma. After monthly follow-up visits, no recurrence has been reported.

Discussion

Malherbe's calcifying epithelioma, sometimes called pilomatrixoma or pilomatricoma, is a benign skin tumour [3]. Its exact aetiology had not been clearly explained, although different studies suggest that a disruption in the hair follicle cycle, trauma, or infection can all lead to pilomatricoma development [5]. It is frequently reported in the paediatric age group as well as during the sixth decade of life. There are not many studies that go back and show a bimodal presentation. Many studies reveal a slight predominance of pilomatricoma in females rather than males. Pilomatricoma is frequently found in the head and neck, particularly in the cervical, frontal, and temporal regions, as well as the eyelids and preauricular regions, before spreading to the upper extremities and trunk [3]. Regarding lower extremity localization, only few occurrences have been documented in the literature.

A pilomatricoma is a single, tightly bound, painless, freely movable mass that is attached to the skin's under-

side but does not extend into the deeper planes. According to Kaddu *et al*., the histomorphological pattern goes through 4 stages: early, fully formed, early regressive, and late regressive [6]. Pilomatricomas can be categorised into 5 clinical types: projecting mass, pigmented, mixed type, ulcerated, and keloid-like. According to these criteria, our case fits the protruding mass and fully developed histomorphological stages. Besides its classical appearance, lesions can be keratotic or telangiectatic, resembling squamous cell carcinoma or basal cell carcinoma, and based on blue-black skin discoloration it can resemble haemangioma. Pilomatricoma size mainly ranges between 0.5–3 cm. Nodules larger than 5 cm are rarely seen [7]. When many lesions are present, it is important to consider the possibility of inherited or associated conditions such as Gardner syndrome, myotonic dystrophy, sarcoidosis, Steinert's disease, Turner syndrome, or xeroderma pigmentosum. Rarely, cases of malignant pilomatrix carcinoma have been recorded. According to Bremnes *et al*., in the literature only 55 pilomatrix carcinoma cases had been reported up to 2016 [8].

A beta catenin gene mutation, which is critical for the growth of hair follicles, is thought to be the cause of about 75% of pilomatricoma cases. Additionally, immunohistochemistry investigations linked pilomatricoma and proto-oncogene B-cell lymphoma antigen 2 overexpression. Foreign bodies, giant-cell tumours, epidermal cysts, dermoid cysts, sebaceous adenoma or carcinoma, calcified lymphadenopathy, ossified or calcified haematoma, and lipomas should all be ruled out as differential diagnoses [4]. Radiological modalities such as ultrasound, MRI, and CT scans play a minimal role in the diagnosis. Because it is readily available, ultrasonography is usually appropriate, with a positive predictive value of 95.5%. It can show the mass's superficial location, continuity with deeper structures, and degree of calcification [4]. Thus, definitive diagnosis of the tumour is only acquired through histopathological examination.

Standard treatment for pilomatricoma is surgical excision with 5–10 mm safety margins. Recently, Mohs micrographic surgery had been employed, especially in situations of suspected malignancy, to assure full margin-free excision [4]. This procedure leads to a low recurrence rate.

Conclusions

A diagnosis of pilomatricoma can be difficult, especially if the site is atypical. Radiological tools such as ultrasound and colour Doppler can assist the clinician towards the diagnosis, but only histopathological examination can give a definitive diagnosis.

The authors declare no conflict of interest.

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