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RECURRENT PILOMATRIXOMA

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Abstract

Pilomatrixoma is a rare tumor of the matrix cells of the hair follicle, it commonly occurs in young male patients and the commonest site is the head and neck region. Here is an odd case at which the tumor developed in the lower limb of a 65 year old male patient.

Introduction

ilomatrixoma, also known calcifying epithelioma of Malherbe or pilomatricoma. It is a benign neoplasm that is derived from hair follicle matrix cells. These lesions are typically found in the head and neck region, but less commonly in the extremities 1-7. It occurs at 1st & 2nd decades of life, but can develop in old people. The lesion is usually less than 10 mm in diameter but a giant form can occur in rare cases. Recurrence after surgical excision is rare, but could occur especially in aggressive type^{8,9}.

Case Report

A 65 year old man presented with 6 months history of gradual onset of a painless left thigh mass. He denied any history of trauma and he had no fever, weight loss, fatigue, paraesthesia or any other associated symptoms. Physical examination showed a 10x10 cm, nontender, firm mass over the anterolateral aspect of the middle of the left thigh. It was superficially attached to the skin but not attached to the deep structures and was easily mobile in all directions. It had smooth surface, well defined edge, and not pulsatile. The overlying skin was normal and there was no lymphadenopathy. The neurovascular

examination of the left lower limbs was normal.

Plain x-ray of the left thigh was normal apart from soft tissue shadow. Ultrasonic examination showed a well-defined solid mass at the subcutaneous tissue of the left thigh. Hematological & biochemical investigations were unremarkable.

Excision of the mass was done under local anesthesia and it was sent for histopathological examination.

Grossly, it was a nodular mass, 10x7x4 cm, covered by skin ellipse. The mass is white-yellow in color, hard, granular and gritty.

Microscopic examination reveals nests of basal cells with abundant abrupt keratinization forming ghost cells with no evidence of malignancy, the diagnosis was giant pilomatrixoma (Figure 1).

After eight and a half month, the patient presented with a recurrent mass at the previous site of tumor, it was 5x5x2 cm. Excision was done with the overlying skin under local anesthesia (Figures 2-5). Histopathological examination showed aggressive pilomatrixoma with cellular atypia with no malignancy. Postoperatively, the wound recovered well without any complications. He had no evidence of recurrence in his one-year follow-up.

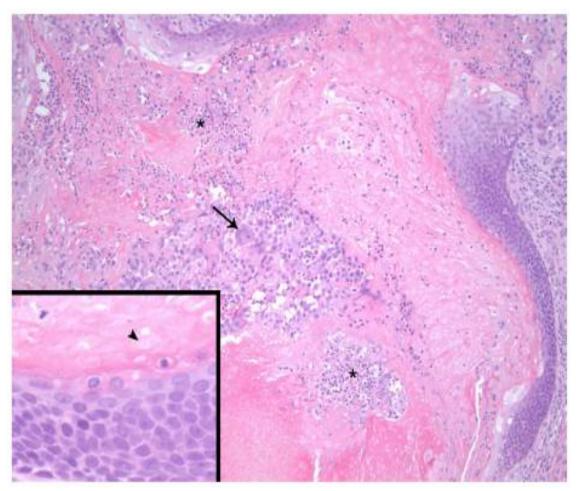


Figure 1: Pilomatrixoma of the left thigh

Asterisks: basaloid epithelium with acute inflammatory cells, Arrow: foreign body giant cells. Arrow head: Ghost cells characteristically retain their cell and nuclear borders, however, the nuclei lose their basophilic staining leaving a "ghost-like" remnant in eosinophilic keratin debris.



Figure 2: Recurrent mass



Figure 4: Skin defect after excision



Figure 3: Excised tumor



Figure 5: Wound closure

Discussion

In 1880, Malherbe and Chenantais first described this benign lesion and referred to as the calcifying epithelioma, though it was thought to derive from sebaceous glands¹⁰. The term pilomatrixoma was used in a publication by Forbis and Helwig in 1961 to better describe the histological source¹¹. These tumors are usually found in the head and neck region, but they have also been described in extremities. They present most commonly in children and young adults, and more in females^{12,13}.

Α rare malignant form, pilomatrix carcinoma, has been described 12-14, and about 90 cases have been reported in the literature. It is locally aggressive tumor and can recur. In several cases, it has demonstrated metastases. Many features are similar between these benign and malignant lesions; include a high mitotic atypical mitoses, necrosis, infiltration of the skin and soft

tissue, with invasion of blood lymphatic vessels^{12,13}.

This patient was an adult male and presented with a lesion in the lower limb unlike the usual presentation of such lesions in children and young adult female patients. In addition, pilomatrixoma is a giant type (more than 5 cm) & aggressive recurrent disease unlike the common presentation of such lesions in small size (less than 3 cm). Recurrent aggressive disease is rare (0-3%)¹⁵⁻¹⁹. Recurrence may be due to incomplete excision^{8,9}. It is excised because of malignant potential of such lesions^{20,21}.

Pilomatrixomas are often misdiagnosed on preoperative evaluation. In a series of 51 pilomatrixomas, Wells et al found that the referring diagnosis was incorrect in 94% of cases²². Incorrect preoperative diagnoses most commonly included unidentified masses, as well as epidermoid cysts, sebaceous cysts, dermoid cysts, nonspecified cysts, and foreign bodies^{1,23}.

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