**Case Report**
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# Pilomatrixoma of the Thigh - A Case Report

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## Abstract

Pilomatrixoma is a benign skin tumor derived from hair follicle matrix cells. They typically present as a superficial, firm, solitary, slow-growing, painless mass in the dermis. Pilomatricomas are frequently misdiagnosed and are not usually considered in differential diagnoses, either in clinical set-up or during cytological reporting. These lesions are typically found in the head and neck region. Involvement of the lower extremities is relatively uncommon and can be mistaken for malignancy. We present the case of a 55-year-old male with a pilomatrixoma of thigh which is a rare site of occurrence of the lesion and we review the literature regarding pilomatrixomas.

**Keywords:** Hemorrhagic Stroke, Ischemic Stroke, Risk Factors, Socioeconomic Status

## 1. Introduction

Pilomatrixoma also known as a calcifying epithelioma of Malherbe[1] and is a benign skin appendage tumor derived from the hair matrix[2]. Although it can occur at any age most cases of pilomatrixoma occur in children. It is most common on the head, face and neck, but lesions can also occur on the upper and lower extremities and trunk[3] and are rarely reported in other sites[4]. Pilomatrixoma is associated with high levels of beta-catenin caused by either a mutation in the APC gene or beta-catenin gene. These high levels of beta-catenin can aid cell proliferation and inhibit cell death. Patients usually present with a single, firm, stony, hard nodule.

## 2. Case Summary

A 55 year old male presented with complaints of a single swelling in the posterior aspect of right thigh since 1 year. The swelling was painless and gradually progressed in size. There was no history of trauma or fever. On palpation there was 5x3 cm single hard non tender swelling which was freely mobile in vertical and horizontal directions with no signs of inflammation. There were no other palpable masses in the extremities Local ultrasound showed a well defined elliptical heterogenous lesion measuring 10.4x2.9 cm in size with diffuse calcification noted in the intramuscular plane in posterior aspect of right upper thigh. Underlying bony cortex and vessels were normal. On colordopplerno vascularity was present. X-ray of right thigh was done which showed calcified radio opaque mass in the upper thigh level.

FNAC of the swelling was inadequate and inconclusive. Routine blood investigation was within normal limits.

Decision was taken to do excisional biopsy of the swelling.

## 3. Intra-Operative Findings

A vertical incision was taken over the swelling. Fascia incised. Intramuscular plane reached by blunt dissection. A well circumscribedbony hard encapsulated swelling

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was noted in the intramuscular plane which was delivered out. Its capsule was also removed. Suction drain was kept. Muscles and fascia approximated. Skin closed.

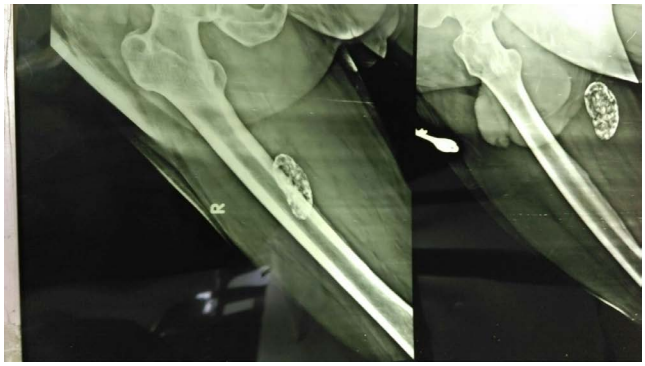

**Figure 1.** X ray of Right thigh showing a radio opaque mass in the subcutaneous tissue.

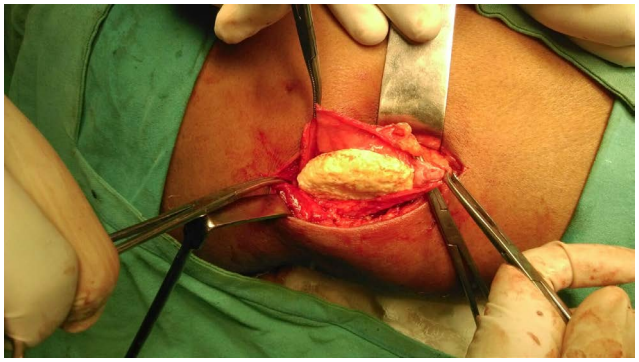

**Figure 2.** Intra operative photograph showing a calcified mass.

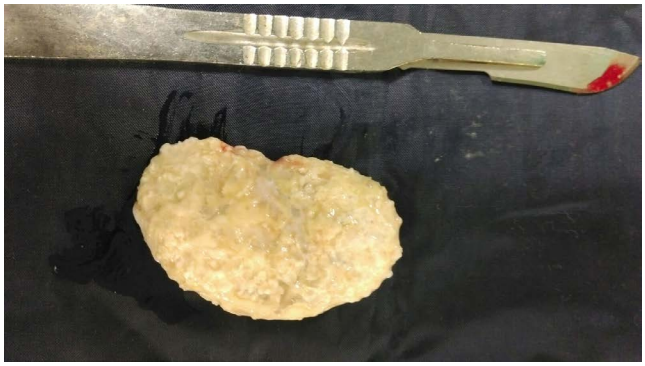

**Figure 3.** Post operative photograph of Pilomatrixoma.

The specimen and capsule was sent for histopathological examination.

Post op recovery of patient was good and was discharged on 4th day after drain removal.

Histopathology examination revealed mass composed of basaloid cells and showed ghost cell appearance suggestive of pilomatrixoma. No evidence of malignancy. The capsule showed fibrocollagenous tissue.

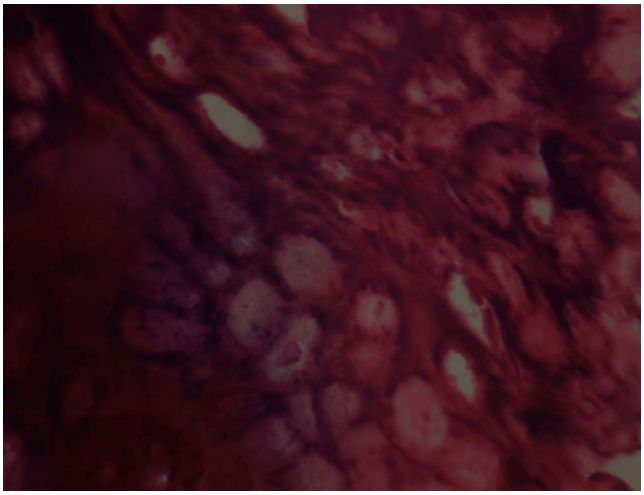

**Figure 4.** Pilomatrixoma with prominent basaloid cells.

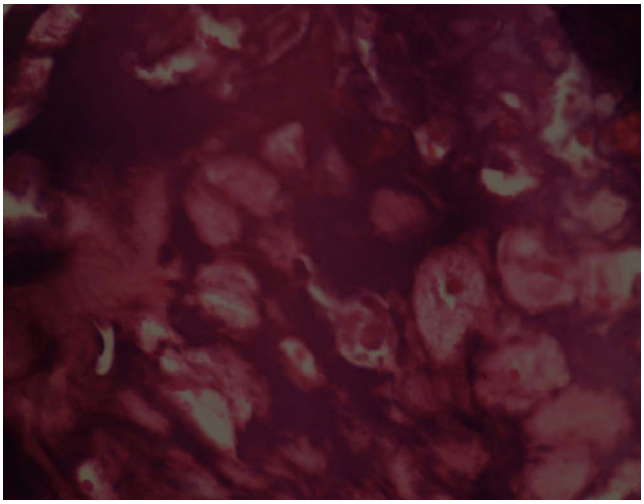

**Figure 5.** Pilomatrixoma with Ghost cells (shadow cells) and basaloid cells, associated with a granulomatous reaction.

## 4. Discussion

Pilomatrixoma, is a benign skin appendage tumor. It is derived from the hair matrix. It is slow-growing (grows over several months or years). It presents as a hard mass beneath the skin, usually not associated with any other signs and symptoms.

In 1880, Malherbe and Chenantais first described this lesion, referred to as the calcifying epithelioma, though

it was thought to derive from sebaceous glands[5]. Forbis and Helwig in 1961 introduced the term pilometrixoma do describe the histological source of the swelling[6]. The swelling is present as superficial nodule. It is usually non tender. In a series of 346 cases, 32 percent presented with pain and tenderness[7]. Most lesions are small, measures 0.5-3 cm. Rarely, giant lesions up to 15 cm are reported. Most commonly, the overlying skin is of normal in colour and texture; however, the examiner may observe the Tent sign, consisting of flattening of some portion or the entire surface of the tumor with angulation resembling the side of a tent. This sign is sign seen only by stretching the skin[8]. This has been attributed to attachment of the tumor to the overlying epidermis, and the associated bluish or reddish discoloration is due to the growth of blood vessels into the overlying skin. Pilomatrixom as are usually solitary. Multiple pilometrixoma have been reported in association with genetic disorders, such as myotonic dystrophy, Gardner syndrome, xerodermapigmentosum, and basal cell nevus syndrome[9]. A review of the literature in 1999 showed that 25 cases were associated with myotonic dystrophy, a rare neuromuscular disorder transmitted as an atypical autosomal dominant trait with variable expression[10]. Patients tended to have multiple pilomatrixomas, the maximum lesions reported are 31. Multiple pilomatrixomas may be a marker for myotonic dystrophy. Investigators have also reported isolated cases in association with Rubinstein-Taybi syndrome, Turner syndrome, Goldenhar syndrome, Churg-Strauss syndrome, sternal cleft and mild coagulative defect, and sarcoidosis[11].

The exact underlying cause is not well understood; however, somatic changes (mutations) in the CTNNB1 gene are found in most isolated pilomatrixomas. These data directly implicate beta-catenin/LEF misregulation as the major cause of hair matrix cell tumorigenesis in humans[12]. In one study of 10 pilomatrixoma lesions, all immunostaining results were strongly positive for BCL2[13]. This is a proto-oncogene that helps suppress apoptosis in benign and malignant tumours; these data suggest that faulty suppression of apoptosis contributes to the pathogenesis of these tumours.

The lesion is usually found in the lower dermis and subcutaneous fat. It is sharply demarcated and is usually surrounded by a connective tissue capsule. In this patient's case, the definitive diagnosis was made only after histologic examination following excision of the mass. Irregularly shaped islands of epithelial cells are seen; they can be recognized as either basophilic cells or shadow cells. Basophilic cells are usually arranged either on one side or along the periphery of the tumour islands. The shadow cells have a central unstained area, corresponding to the lost nucleus. The tumor is composed of ghost cells, basaloid cells, and giant cell, in addition to keratin debris and intracellular and stromal calcifications[9]. Calcium deposits are seen in 75% of lesions with von Kossa staining. Pilomatrixomas are often misdiagnosed on preoperative evaluation most commonly included unidentified masses, as well as epidermoid cysts, sebaceous cysts, dermoid cysts, nonspecified cysts, and foreign bodies.

The following imaging studies may be helpful:

- Radiography: P lain radiography often shows nonspecific calcification of the lesion.
- Ultrasonography: On ultrasonography it consistently appeared as a target lesion, with a hypoechoic rim and an echogenic center. It also shows calcification.
- Magnetic Resonance Imaging: MRI may be diagnostic if further reports can confirm the correlation between the high-signal bands in T2-weighted images and the bands formed by basaloid cells evident upon histologic examination.
- CT Scan: Shows a well-delineated subcutaneous tumor containing microcalcifications.

As performed in this case, management of pilomatrixomas typically involves marginal excision. The treatment of choice is surgical excision[15]. Lesions are mostly poorly delineated, but encapsulated forms have been observed; these are less likely to recur because complete resection is easier. Incomplete resections have been followed by local recurrence; wide resection margins (1-2 cm) have been recommended to minimize the risk of recurrence. Secondary lesions after surgery are rare; this risk decreases progressively with age. Lesions on the extremities may be left untreated unless they become large or symptomatic, however in many cases these are excised for definitive diagnosis. If the tumor adheres to the dermis, the overlying skin may be excised. The recurrence rate is low, ranging from 0 to 3 percent[15].

If a lesion recurs after excision or rapidly enlarges, it should be excised due to malignant potential or possible misdiagnosis.

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