Case Report

# An Unusual Presentation, Pilomatrixoma in the Buttock Area – A Case Report

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**ABSTRACT**

Pilomatrixoma is benign tumor of skin appendages it is rare tumor mainly involving head, neck face and upper extremities. [1]

A 16-year-old male developed a lesion, over a period of 2 year, at the middle of right buttock. The presumed diagnosis was a sebaceous cyst / pyogenic granuloma. An excision biopsy was performed and the histopathologic diagnosis was a pilomatrixoma.

**Key Words:** Pilomatrixoma, Buttock region.

## INTRODUCTION

Pilomatrixoma manifests clinically as a firm, deep-seated nodule, located mainly on the head, neck and upper extremities.Pilomatrixoma is commonly misdiagnosed preoperatively and is extremely rare at other than above mention sites. In reports of Wells at al[2] the referring diagnosis was improper in 94% of cases and the preoperative recognition in 57% cases.

## CLINICAL DATA

A 16-year old male patient presented to us with a painless lump of 10 cm x 06cm size, in the left buttock region for 02 years. There was brownish discoloration of over lying skin. A provisional clinical diagnosis of infected sebaceous cyst / pyogenic granuloma was considered. The rest of the surrounding examination yielded normal results.

The lesion was surgically excised by means of a total thickness comprising the lesion. There was an increase of small sized vessels and the overlying dermis and epidermis were atrophic. Histopathology of the excised lesion was done with a final diagnosis of Pilomatrixoma.

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Usually, the clinical development of pilomatrixoma is benign, although malignant transformations have been described[3]. Treatment is surgical excision and, if done adequately, recurrence is exceptional.

## DISCUSSION

Pilomatrixoma or calcifying epithelioma of Malherbe is a benign tumor of skin appendages with presumed differentiation towards hair cortical cells. The term pilomatrixoma is preferred to the former name of "calcifying epithelioma of Malherbe" which may give the unwary clinician a false impression that the lesion is malignant.[4,5] It manifests clinically as a firm, deep-seated nodule, located mainly on the head, neck and upper extremities.[3] The lesion presents as a slow-growing, bluish, solitary, and well-circumscribed nodule within the dermis.

It may arise in persons of any age, but about 40% arise in children younger than 10 years and 60% in the second decade of life. Most studies report a slight preponderance in females.[1] In this case, patient is male and presented at the age of 16 year. Pilomatrixoma can have a history of several years. Growth can arrest spontaneously. In our case, history only goes back to two years.

Although pilomatrixoma is a benign skin tumor, in the literature there are reports of pilomatrixoma carcinoma.[6,7,8] Recurrence and histologically increased mitotic figures with atypical cells favor malignant changes.

Plain film radiography can be useful in pilomatrixoma because it can reveal areas of

prominent stippled calcification.This is referred as 'tent sign,' which indicates several facets and irregular angles of pilomatricoma. Positive tent sign should alert the diagnosis of PMX[9,10]Diagnosis is only possible if we consider pilomatrixoma in the differential diagnosis at sites even not typical.

Histology reveals irregular islands of progressively degenerating epithelial cells separated by fibrous stroma. The examination reveals 2 main cell types: basaloid cells at the margin of epithelial islands and nucleated shadow cells in the inner core. Calcium deposits are seen as fine basophilic granules. The key feature is the presence of abrupt keratinization of these cells, leading to the formation of ghost or shadow cells as confirmed in histopathology report.

Pilomatrixoma is associated to mutations in the betacatenine gene (CTNNB1) and it has been confirmed that this mutation does not only occur in pilomatrixoma but also in hair follicle carcinomas. The betacatenine dysfunction is the main cause of tumor growth in the hair follicle.[11]

Complete surgical excision is recommended for pilomatrixoma. A good prognosis is noted, but malignant transformation rarely occurs and is associated with multiple local recurrences.

## CONCLUSION

Despite the clinical picture and benign character of pilomatrixoma, the histopathology is essential for confirmation of diagnosis and benign nature of the lesion.

Complete surgical excision, including the overlying skin is the treatment of choice

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**Figure Legends:**

**Figure 1: Gross specimen after excision, showing brownish discoloration of overlying skin.**

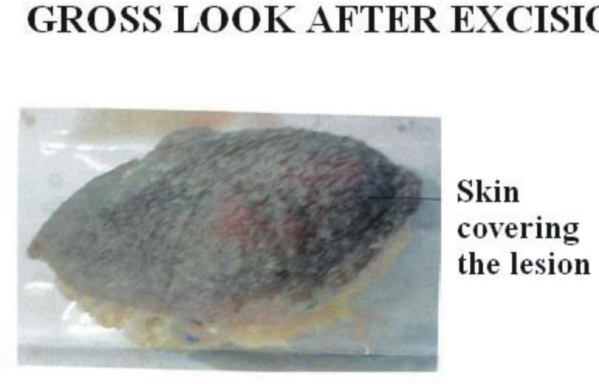
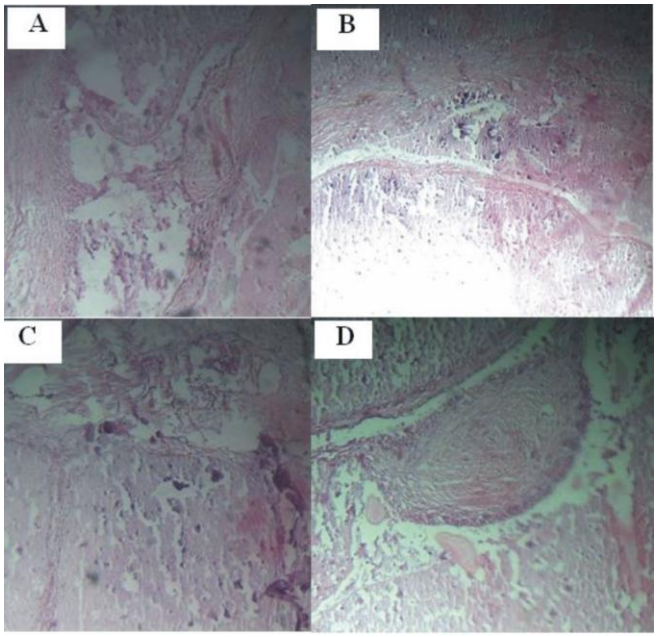


**Figure 3: Histological slides (A, B, C, D), magnification 40X, Hematoxiline Eosin staining. Showing rounded nodules, lined by stratified squamous epithelium with many ghost like cells and focal areas of calcifications**

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| Submitted for Publication: | 28-05-2014 |
| Accepted for Publication: After minor revisions | 30-05-2014 |

**Figure 2: Cut surface of the specimen, showing gray to while nodules.**