

## Multiple Pilomatricomas: A Case Report

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**Received:** November 09, 2022; **Published:** November 16, 2022

### Abstract

Pilomatricoma is a benign tumor that arises from hair follicle matrix cells. Involvement of the upper limb and the lower limb are uncommon region. We report the case of a 9 year old girl, who presented at the age of 7 year old with a symptomatic painful firm nodule over the left arm and the right thigh and at the age of 9 year old with a painless firm nodule, associated with calcification discharge over the left leg whose histology was suggestive of Pilomatricoma.

**Keywords:** *Pilomatricoma; Shadow Cell; Ghost Cells; Basaloid Cells; Benign Neoplasms; Pilomatricoma*

### Introduction

Pilomatricoma was described by Malherbe and Chenantais in 1880 [1]. In 1961, Forbis and Helwig [2] proposed the term “pilomatricoma” instead of “calcifying epithelioma,” because the studies demonstrated the hair follicle matrix as the point of origin. These tumors are commonly present in the head and neck region, but less commonly in the upper limbs and the lower limbs and are rarely reported in other sites [3]. Pilomatricoma usually presents as asymptomatic, firm to hard, freely-mobile and slow growing, solitary mass t. In some cases, patients complain of pain symptom or discharge from the lesions [4,5]. Some patients still present multiple lesions [6].

Pilomatricoma is located in the deep dermis or in the subcutaneous layer. The size of the tumor mostly exceed no more than 3 cm. The overlying skin is a bluish discoloration or ulceration [7].

We report a case of multiple pilomatricomas over left arm, right thigh and left leg. We discuss about the clinical features, histopathological features regarding pilomatricoma in the upper extremity and the lower extremity.

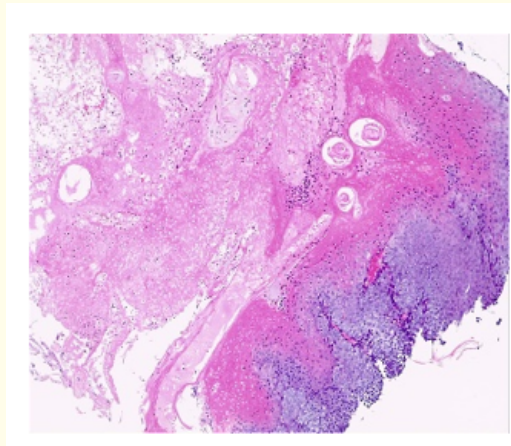
### Case Report

A 7-year-old girl presented with a 6 months history of insidious onset of two lesions -masse over left arm and right thigh. The mass on both sites were painful, progressively enlargement and not associated with itching and discharge. She denied any history of trauma over both sites and she had no previous history of fever, fatigue, weight loss, numbness or tingling.

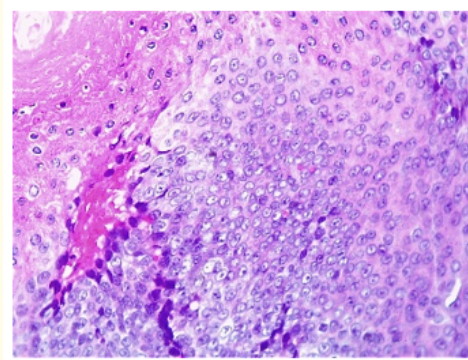
Physical examination revealed a 2 cm; mild tender, firm mass over the left arm. There was another palpable mass in the right thigh with a 1cm tender, firm mass. They were superficial and easily mobile. Excision biopsy was performed under local anesthesia. Grossly the

tumor were pale-yellow chalky in appearance and well circumscribed. Histopathology revealed formation of anucleated shadow cells, No evidence of malignancy cells.

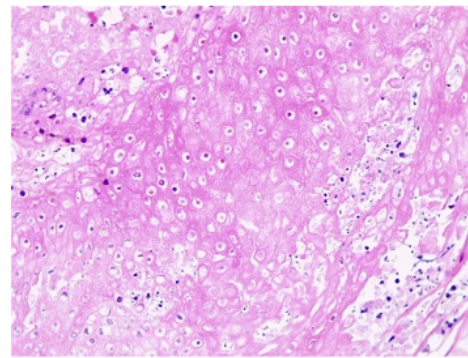
Two years later, she developed a firm mass with a 1 year history of insidious onset of the solitary mass over left leg. The mass is protruding from the skin, painless, progressively enlarging and associated with calcification discharge. She denied any history of trauma over the left leg. Physical examination revealed a 1 cm; non-tender, rubbery mass over the left leg. It was superficial and easily mobile. There were no palpable masses in the other extremities and no axillary adenopathy was present. Excision biopsy was performed under local anesthesia. The tumor was light-brown in grossly appearance. Histopathology revealed poorly-circumscribed mass composed of islands of basaloid cells and eosinophilic cells that was devoid of nuclei (Figure 1-3). Inflammatory cell infiltrate predominated by focal of lymphohistiocytes. The presence of prominent inflammatory reaction, foreign body granuloma, foci of calcification and fibrosis is also characteristic.



**Figure 1:** Histology of skin [H&E section, magnification 10x10] showing poorly circumscribed mass in the dermis composed of islands of basaloid cells and eosinophilic shadow cells.



**Figure 2:** Microscopic view [magnification 25x10] of pilomatricoma showing shadow cells and basaloid cells.



**Figure 3:** Microscopic view [magnification 40x10] of pilomatricoma revealing ghost cell, transitional cells and basaloid cells.

## Discussion

Pilomatricoma is a benign tumor occurring in the deep dermis or subcutaneous layer. It usually presents as a solitary lesion, but multiple lesions could be found in approximately 2% to 10% of cases [8]. In our case report presented with multiple lesions presentation. From previous studies, the cause of pilomatricoma is related to beta-catenin mutation [9]. Multiple lesions presentation is associated with familial inheritance or syndromic conditions, such as myotonic dystrophy, Gardner syndrome, Turner's syndrome, Steinert disease, Rubinstein-Taybi syndrome and sarcoidosis [10-12]. Malignant transformation of pilomatricoma is rare [13,14].

Pilomatricoma presents most commonly in children and young adults [3,7,12]. The majority of pediatric patients occurred before 10 years of age [8,15]. Pilomatricoma both solitary and multiple presentation had a female predominance [8,15]. The most common area of pilomatricoma was in the head and neck, the cheek or the palpebral area, followed by the upper extremities, trunk, and lower extremities [6]. Similarly in our case report, the patient is female predominance.

The histopathological features of a pilomatricoma shows a well defined tumor, which is surrounded by a connective tissue capsule. It is located in the dermis or subcutaneous layer. Composition of the tumor is from the islands of epithelial cells and varying amount of basaloid metrical cells. When the tumor matures, there is degeneration of these basaloid cells. It's characterized by formation of anucleated shadow cells. The tumors of recent origin, basophilic cells are usually present, as the lesion aging, there is decreasing number of basophilic cells because of their transformation into shadow cells [16]. The presence of prominent inflammatory reaction, foreign body giant cells, central calcifications and keratin debris is also characteristic.

The clinical differential diagnosis of these lesions should include sebaceous cyst, dermoid and epidermoid cysts, metaplastic bone formation, foreign body reaction, calcified hematoma, trichoepithelioma and basal cell epithelioma [13,14]. Surgical excision is recommended the treatment of choice. Recurrence is rare [8]. The cause of recurrence is mostly due to incomplete resection. Sufficient resection margins are recommended to reduce the recurrence rate [17].

Malignant transformation of the pilomatricoma is very rare [18,19]. All patients after surgical excision of tumor need dermatological evaluation with close long-term follow-up [11].

## Conclusion

We have presented this case report because of its multiple lesions presentation.

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