Case Report

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# Pilomatricoma of the calf: a case report and review of literature

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**Introduction and importance:** Pilomatricoma (PMC) is a benign adnexal dermal or subcutaneous tumor, which is derived from immature hair matrix cells. It makes up around 20% of all tumors related to hair follicles in most series and is therefore the most common hair-follicle neoplasm. Nevertheless, diagnosing it remains intricate due to the prevalence of more frequent pathological conditions in soft-tissue. Anatomopathological examination proves to be a valuable asset, offering a definitive and certain diagnosis.
**Case presentation:** The authors hereby present a case of a 17-year-old patient with no medical history, who was referred to our medical unit subsequent to the emergence of swelling in the right calf. MRI results highlighted the presence of a subcutaneous nodule situated on the right calf. Following a percutaneous biopsy, the diagnosis of PMC was definitively confirmed. A successful surgical excision of the tumor was performed, and the postoperative progress demonstrated positive outcomes.
**Clinical discussion:** PMC usually appears as flesh-colored to white, firm papules or papulonodules that may have an overlying pink to blue hue. MRI plays a crucial role in diagnosis, as it delineates the tumor's extent in relation to the skin and muscle compartments. Preoperative histological confirmation is essential to rule out other potential diagnoses and precisely establish the required resection margins.
**Conclusion:** PMC is an infrequent occurrence in general surgery departments. General surgeons should, however, be well-acquainted with this benign tumor while assessing soft-tissue masses.

**Keywords:** calcified epithelial carcinoma, case report, pilomatricoma, skin, tumor

## Introduction

Soft-tissue tumors are a relatively rare and diagnostically challenging group of neoplasms that can have varying lines of differentiation. The vast majority of soft-tissue tumors are benign lesions such as lipomas, haemangiomas, and fibrohistocytic lesions. Soft-tissue sarcomas are rare, accounting for less than 1% of malignancies.

Pilomatricoma (PMC) is categorized among benign superficial soft-tissue tumors. It was first described by Malherbe and Chenantais in 1880 who hypothesized that the lesion originated from a sebaceous gland and therefore they called the tumor '*calcifying epithelioma of sebaceous glands*'[1]. The term pilomatrixoma, to denote origin from hair matrix cells, was suggested by Forbis and Helwig in 1961[2]. This was later corrected to PMC, as more etymologically correct[3]. Most occur in children and adolescents, but they can rarely occur in elderly patients[4]. The most frequent locations are the head and neck, the reach of the limbs remain exceptional. These tumors have a wide variety of signs, which often causes misdiagnosis. Fortunately, the overall prognosis is favorable. The cure without recurrence is the rule after complete surgical excision. We report a rare case of a young girl with PMC in an unusual location in the calf.

This work has been reported in line with the Surgical CAse REport (SCARE) 2023 criteria[5].

### HIGHLIGHTS

- Pilomatricoma is a rare benign tumor that originates from the matrix cells of hair follicles.
- It is rarely considered a differential diagnosis of benign masses and imposes a diagnostic challenge preoperatively.
- Although rare, pilomatricoma can affect the calf.
- Histology remains the gold standard for diagnosis.

## Case presentation

A 17-year-old patient, without any significant personal or family medical history, presented at our outpatient department with a subcutaneous mass that had gradually increased in size over the past month. The clinical examination unveiled a subcutaneous mass that appeared multinodular with a firm consistency. The mass was well-demarcated, measuring ~3 cm along its longest

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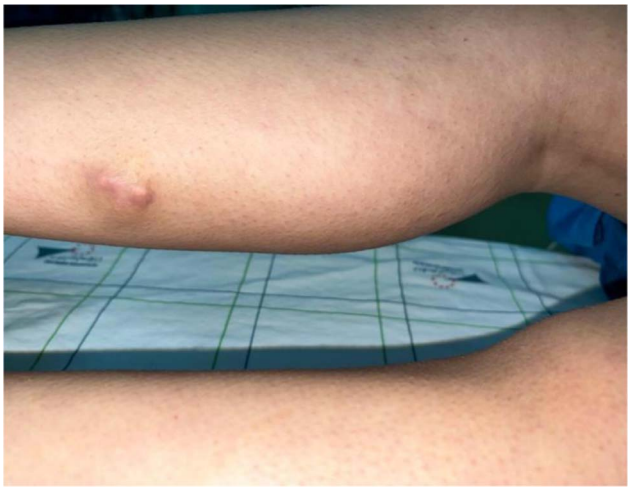

**Figure 1.** Image showing a subcutaneous nodule located on the right calf.

axis. It was observed to be adherent to the skin while maintaining mobility relative to the deeper tissue planes (Fig. 1). Notably, there were no signs of regional lymphadenopathy. In terms of the initial assessment, the differential diagnosis considered dermoid cyst, adenopathy, epidermal cyst, hemangioma, lipoma, as well as malignant soft-tissue tumors, including basal cell carcinoma, squamous cell carcinoma, and malignant melanoma.

Ultrasonography unveiled a well-defined, oval-shaped, heterogenous hypoechoic lesion with a posterior acoustic shadow situated within the subcutaneous layer of the right calf (Fig. 2A). It does not extend deep to the fascia. MRI indicated the presence of a vascularized subcutaneous nodule on the posterior-medial aspect of the right leg (Fig. 2B-C).

The patient underwent an incisional biopsy that allowed for establishing the diagnosis. The surgical intervention was made by a senior surgical oncologist. During the procedure, the lesion was infiltrated with a mixture of 2% lidocaine and bicarbonates. An elliptical incision was meticulously made over the mass and the skin was excised as the tumor was removed in bloc (Fig. 3A). Adequate hemostasis was made using bipolar electrocautery. The wounds were closed by direct suture without tension. The patient was discharged from the hospital the next day and had a follow-up consultation after 15 days, showing good progress. She was satisfied with the treatment and the postoperative results. The 3-month follow-up did not reveal any recurrence.

## Results

Gross examination of the skin excision specimen showed a whitish nodular lesion, partially calcified, measuring $2 \times 1.8 \times 1$ cm. Microscopically, the tumor was composed of sheets of basaloid cells. These cells vary in size, with some being small, highly basophilic, and exhibiting a central, hyperchromatic, and nucleolated nucleus. Others appear mummified, anucleated, with faintly visible cellular borders. Rare mitotic figures are observed. The adjacent connective tissue is fibroinflammatory, characterized by lymphocytes and multinucleated giant cells, suggestive of a foreign body reaction. Some areas of calcification were noted (Fig. 3B).

## Discussion

PMC is a rare disease; studies report the incidence to be between 0.001 and 0.0031% of all dermatohistopathologic materials submitted for examination[6]. Table 1 compiles published studies on PMC that include more than 100 tumors. The largest series available is the one by Moehlenbeck *et al*., dated back to 1973, which includes a total of 1569 PMCs. Head, upper extremity, neck, trunk, and lower extremity are affected with decreasing frequency. Localization in the upper extremity is greater than in the lower extremity in a ratio of about 3:1[7].

It mainly affects the child before the age of 10 years with a female predominance (sex ratio of 1.5). It would also be more common in patients with Steinert myotonic dystrophy and in Gardner syndrome, which associates rectocolic polyposis with extradigestive signs[4]. Concurrent disorders were not identified in our patient. The exact etiology and pathogenesis are still unknown. Several mechanisms of development have been proposed. Most accepted theories involve the inclusion of epidermal elements into abnormal locations. Dubreuilh and Cazenave proposed that PMCs originate from branchial clefts and are of ectodermal origin, while others have described the tumors as epithelial hamartomas[13]. These tumors express human hair keratin basic protein 1, a marker of normal cortical cells of the hair shaft. Differentiation of progenitors into cortical cells may therefore play a role in development of these tumors. Most of them express a β-catenin abnormality, such as mutation in

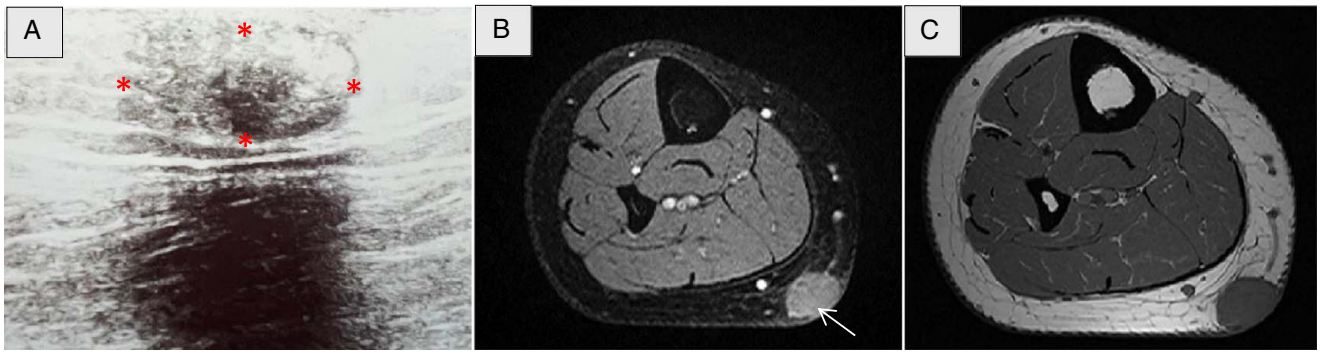


**Figure 2.** (A) Gray scale US image revealing a well-circumscribed heterogeneous, hypoechoic mass (calipers), with a posterior acoustic shadow. The MRI findings indicate that the lesion exhibited hyperintensity (arrow) in T1 injected sequences (B) and hypointensity in T1 sequences (C).

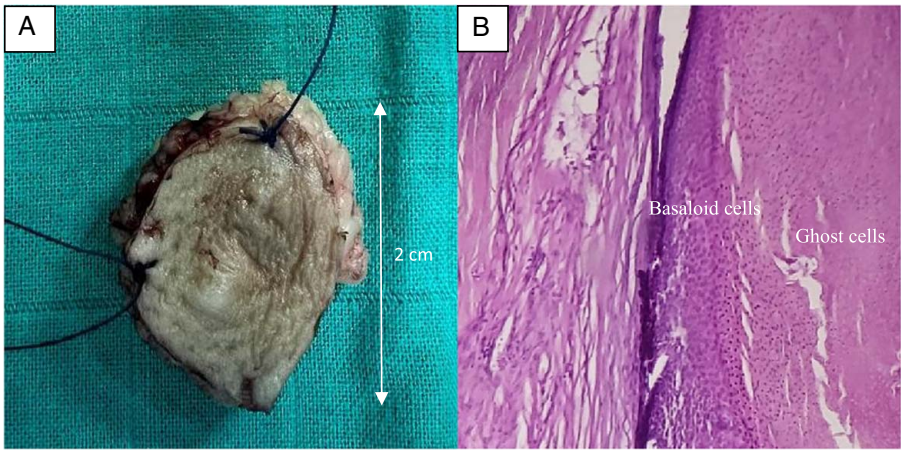


**Figure 3.** A: Composite operative picture. B: Microscopic examination shows alternating bands of small basophil cells with very high nucleocytoplasmic ratio with significant mitotic activity, and anucleated ghost cell ranges.

CTNNB1, which plays an important role in the regulation of normal cell growth and function[14].

PMC typically occurs as an asymptomatic subcutaneous nodule, round or oval, of hard or firm consistency, adhering to the superficial plane while being mobile to the deep planes[4,15], the skin next to the lesion is often bluish. The usual size is less than three centimeters, but cases of giant PMCs exceeding five centimeters in diameter have been reported. It is generally unique, but some patients have already developed, simultaneously or successively, several PMCs[15]. The 'tent sign' is characteristic of this injury and refers to the palpation of a firm and lobulated consistency when the overlying skin is stretched between the fingers. Recently, the 'wrinkle or skin fold sign' has also been described as the formation of a wrinkle or fold that occurs when slight pressure is applied to the tumor margins with both thumbs perpendicular to the skin tension lines[16].

The primary diagnostic tool when evaluating a soft-tissue mass is US, which typically reveals a PMC as a small subcutaneous mass characterized by a 'target' appearance, consisting of a central echogenic region surrounded by a hypoechoic outer rim. The existence of a posterior shadow cone will indicate the presence of calcifications. Intermediate internal vascularity can be seen in 50%. MRI allows the study of the anatomical relationships of the tumor with the adjacent structures for optimal planning and guides biopsy; therefore, the first diagnostic step is to classify the tumor as either 'deep' or 'superficial', depending on the location of the investing fascia. Standard radiography is only useful when faced with the suspicion of a PMC when it is significantly calcified. Histology remains the gold standard for diagnosis by identifying basaloid cells with or without ghost cells most often associated with calcifications[17]. Given the risk of tumor cell dissemination along the biopsy tract with local recurrence, it is necessary to accurately identify this path to enable its excision during tumor resection. Additionally, it is recommended to perform the biopsy along the axis of the limb and to avoid reaching the muscle fascia to prevent any potential dissemination.

The reference treatment consists of a complete surgical excision taking a skin spindle, especially if the lesion is adherent to the dermis[4]. This is because malignant transformation is possible and spontaneous regression has not been observed[18]. The prognosis of PMC is generally good; carcinomatous degeneration is extremely rare and has been reported in elderly subjects with a history of multiple excision attempts[10]. The incidence of recurrences after surgery has been reported to be between 0 and 6%[4].

**Table 1**

**Series of more than 100 cases of PMCs cited in the literature.**

| Authors | Number of cases | Number of years | Lower extremity's location |
|---|---|---|---|
| Mohlenbeck[7] | 1569 | – | 77 |
| Pirouzmanesh *et al.*[8] | 346 | 11 | 7 |
| Guinot-Moya *et al.*[9] | 205 | 33 | 29 |
| O¿Connor *et al.*[10] | 201 | 9 | 0 |
| Lan *et al.*[11] | 179 | 11 | 0 |
| Gay Scoda *et al.*[12] | 179 | 17 | 25 |

## Conclusion

PMC is a skin tumor of rare occurrence that should not be disregarded. Despite its rarity, it stands as the most prevalent among hair-follicle tumors. The localization of PMC in the limbs remains exceptional. Diligent clinical assessment coupled with a heightened sense of suspicion leads to precise diagnosis, suitable treatment, and the prevention of undue extensive surgical interventions. While the prognosis is favorable, continuous monitoring is imperative to detect any potential recurrence.

## Ethical approval

A written consent was received from the patient. In such case, the anonymized presentation of case report does not require a separate approvement by the ethics committee.

## Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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## Author contribution

S.B.: wrote the article and did the review and editing; A.S. (oncology surgery professor) supervised the writing of the paper; N.R.: helped in the interpretation of histological data; O.B., O.A., and Z.E.: have helped in data collection.

## Conflicts of interest disclosure

The authors declare that they have no competing interests.

## Research registration unique identifying number (UIN)

Not applicable.

## Guarantor

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## Data availability statement

Not applicable.

## Provenance and peer review

Not commissioned, externally peer-reviewed.

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