

PILOMATRIXOMA OF THE FOREARM: A CASE REPORT

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ABSTRACT

Pilomatrixoma is a benign neoplasm derived from hair follicle matrix cells. Involvement of the upper extremities is relatively uncommon and can be mistaken for malignancy. We present the case of a 52-year-old woman with a pilomatrixoma of the forearm, and we review the literature regarding pilomatrixomas in the upper extremity.

INTRODUCTION

Pilomatrixoma, also known as pilomatricoma or calcifying epithelioma of Malherbe, is a benign neoplasm that derives from hair follicle matrix cells. These lesions are typically found in the head and neck region, but also occur in the upper extremities and are rarely reported in other sites.¹⁻⁷ The largest case series in the literature includes 346 pilomatrixomas of which 15.3 percent were observed in the upper extremities.⁸ Despite the frequency of presentation of this lesion in the upper extremities, discussion of this lesion is essentially limited to the literature of otolaryngology, pathology, and dermatology.

We present a case of a forearm pilomatrixoma. Additionally, we discuss the clinical features and review the literature regarding pilomatrixomas in the upper extremity.

Case Report

A 52-year-old woman presented with a 5 month history of insidious onset of an isolated right forearm mass, located dorsally at the junction of middle and distal third

of the forearm. The mass was painless, slowly enlarging, and not associated with drainage. She denied any history of trauma, fever, chills, weight loss, fatigue, numbness, or tingling.

Physical examination revealed a 1.0 by 1.0 cm, non-tender, firm mass over the radial aspect of the distal one-third of the right forearm. It was superficial and easily mobile. There was no tenderness noted in the region of the first or second dorsal extensor tendon compartments. The neurovascular status of the right hand was noted to be intact, and Tinel's sign over the mass was negative. There were no other palpable masses in the extremities, and no epitrochlear or axillary adenopathy was present. Plain radiographs were unremarkable.

Excisional biopsy was performed under regional anesthesia. Grossly, the mass was white in appearance and well circumscribed. Histopathology revealed a pilomatrixoma, and the histology is presented in figures 1 and 2.

DISCUSSION

Pilomatrixoma, or calcifying epithelioma of Malherbe, is a benign skin neoplasm that arises from hair follicle matrix cells. In 1880, Malherbe and Chenantais first described this lesion, referred to as the calcifying epithelioma, though it was thought to derive from sebaceous glands.⁹ The term pilomatrixoma was introduced in a publication by Forbis and Helwig in 1961 to better convey the histological source.¹⁰ These lesions are typically found in the head and neck region, but they have also been described in various upper extremity locations. These lesions present most commonly in children and young adults, and they are noted more commonly in females.

A rare malignant counterpart, pilomatrix carcinoma, has been described, and approximately 90 cases have been reported in the literature. It is locally aggressive and can recur. In several cases, it has demonstrated metastases. Many key features are similar between these benign and malignant counterparts; the primary differentiating characteristics include a high mitotic rate with atypical mitoses, central necrosis, infiltration of the skin and soft tissue, and invasion of blood and lymphatic vessels.^{11,12}

In this patient's case, the definitive diagnosis was made only after histologic examination following excision

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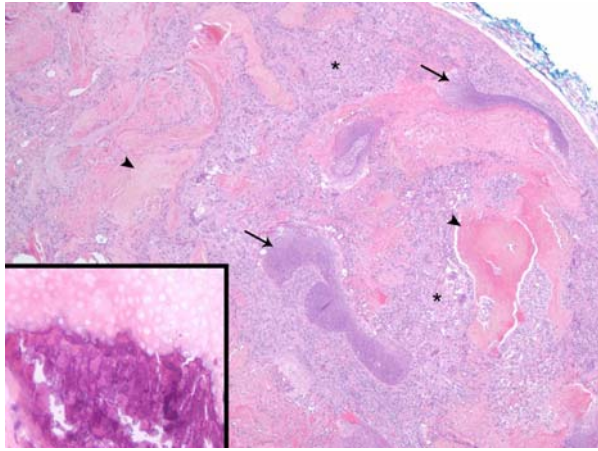


Figure 1. Forearm pilomatrixoma involving deep dermis and subcutis (original magnification 40X). The tumor is well-circumscribed with islands of basaloid cells (arrows) located both peripherally and centrally. Most of the tumor, however, is composed of eosinophilic keratin debris (arrowheads) and a mixed inflammatory infiltrate (asterisks). Focal areas of calcification (inset, 200X) were scattered throughout the tumor.

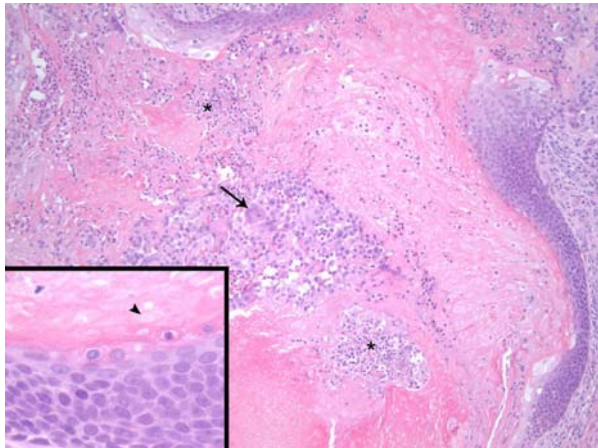


Figure 2. Basaloid matrical germinative cells (far right and top center) blend into keratinized ghost cells (original magnification 200X). Keratin debris elicits an inflammatory response including a mixture of acute inflammatory cells (asterisks) and a foreign body giant cell reaction (arrow). High power view (inset, 400X) showing transition from basaloid cells to ghost cells resulting from matrical keratinization of the basaloid germinative cells. Ghost cells characteristically retain their cell and nuclear borders, however, the nuclei lose their basophilic staining (arrowhead) leaving a "ghost-like" remnant.

of the mass. Pilomatrixomas are often misdiagnosed on preoperative evaluation. In a series of 51 histologically proven pilomatrixomas, Wells et al found that the referring diagnosis was incorrect in 94% of cases, and the preoperative diagnosis was incorrect in 57 percent.¹³ In a recent series of 346 pilomatrixomas, the preoperative diagnosis was accurate and consistent with the pathological diagnosis of pilomatrixoma in only 28.9 percent of cases.⁸ Finally, Kumaran et al. reported a correct preoperative clinical diagnosis in 46 percent following

retrospective review of 78 excised pilomatrixomas.¹ Incorrect preoperative diagnoses most commonly included unidentified masses, as well as epidermoid cysts, sebaceous cysts, dermoid cysts, nonspecified cysts, and foreign bodies.^{1,8}

On presentation, as in this case, palpation of a superficial firm nodule that is not painful or tender is characteristic; however, 32 percent in a series of 346 cases presented with pain and tenderness.⁸ Most commonly, the overlying skin is of normal color 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, often seen only by stretching the skin.¹⁴ 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.⁶ Although pilomatrixomas are usually solitary, multiple lesions have been reported in association with genetic disorders, such as myotonic dystrophy, Gardner syndrome, xeroderma pigmentosum, and basal cell nevus syndrome.^{6,8,15}

The histopathologic features of a pilomatrixoma include a well demarcated tumor which is often surrounded by a connective tissue capsule. Generally, it is located in the dermal or subcutaneous layer. The tumor is composed of islands of epithelial cells made up of varying amounts of uniform basaloid matrical cells and often shows cystic change. Centrally, there is degeneration of these basaloid cells as the tumor matures. This is characterized by formation of anucleated ghost (or shadow) cells due to the central unstained areas of these cells.¹⁶ It is important to note, however, that these ghost cells, though quite specific, are not unique to pilomatrixomas. There may be a variably prominent inflammatory reaction.⁹ Foreign body giant cells, keratin debris, and central calcifications are also characteristic. Calcification has been noted in 70 to 85 percent of cases.⁷

Diagnostic imaging is generally not obtained in the evaluation of pilomatrixomas as they are usually superficial, small, and well-circumscribed. Plain radiographs in this case were unremarkable, but pilomatrixomas may demonstrate foci of calcification. Computed tomography (CT) demonstrates a sharply demarcated, subcutaneous lesion of soft tissue density, with or without calcification. MRI may reveal a rim-enhancing lesion with small areas of signal dropout which may be consistent with calcifications.¹⁵ Ultrasound demonstrates a well-defined mass with inner echogenic foci and a peripheral hypochoic rim or a completely echogenic mass with strong posterior or acoustic shadowing in the subcutaneous layer.¹⁷

Wang et al. noted that 45 percent of cases of pilomatrixoma were incorrectly diagnosed by fine needle aspiration cytology based on their review of multiple case

reports and series.¹⁶ Nevertheless, in their study as well as other more recent studies, fine needle aspiration has been found to be quite accurate when two key components, basaloid cells and ghost cells, are visualized, as this has been found to be specific for pilomatrixoma.¹

As performed in this case, management of pilomatrixomas typically involves marginal excision. 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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