Magnetic Resonance Imaging of a Cystic Pilomatricoma in an Ankle Joint

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abstract

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Pilomatricoma, also known as calcifying epithelioma of Malherbe, is an asymptomatic, slow-growing, benign dermal neoplasm that originates from hair cortex cells. The most common sites are the head and neck (68.2%), followed by the trunk (14.4%) and the upper (15.3%) and lower (2%) extremities. The majority (60%-70%) of tumors develop in the first 2 decades of life. A typical pilomatricoma presents as a firm, calcified, dermal nodule appearing as a solid mass in subcutaneous tissue on computed tomography or magnetic resonance imaging.

This article describes the case of a 42-year-old man who presented with nontender soft tissue swelling in the right ankle joint. Radiographs and magnetic resonance imaging findings showed an inflamed cystic mass with calcification in the lateral malleolar area of the right ankle joint. These atypical magnetic resonance imaging features around the ankle joint led to confusion with other disease entities, including chronic bursitis, pigmented villonodular synovitis, rheumatoid nodules, and gout. After complete excision of the mass, histological examination revealed a cystic pilomatricoma.

Although rare, a cystic pilomatricoma should be considered in the differential diagnosis of soft tissue masses with calcification.

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Pilomatrixoma, also known as calcifying epithelioma of Malherbe, is an asymptomatic, slow-growing, benign dermal neoplasm that originates from hair cortex cells.\textsuperscript{1} The head and the neck are the most common sites (68.2%), followed by the trunk (14.4%) and upper (15.3%), and lower extremities (2%).\textsuperscript{1} The majority (60%-70%) of tumors develop in the first 2 decades of life.\textsuperscript{2} More than 50% of cases preclude correct diagnosis preoperatively. Factors that contribute to misdiagnosis include cystic lesions of various consistencies, punctum-like appearance, atypical location, and absence of clinically recognizable calcification.\textsuperscript{3} A typical pilomatrixoma presents as a solitary firm and calcified dermal nodule appearing as a solid mass in subcutaneous tissue on computed tomography (CT) or magnetic resonance imaging (MRI).\textsuperscript{4} To the current authors’ knowledge, no reports have been published in the English literature describing the MRI appearance of cystic pilomatricoma, although the cystic variant of the tumor may represent up to 12% of pilomatricomas.\textsuperscript{3} This article describes the case of a 42-year-old man with pilomatricoma presenting as a cystic mass in the ankle joint.

**CASE REPORT**

A 42-year-old man presented with a 6-month history of soft tissue swelling in his right ankle. The patient reported no history of trauma or major surgery in the ankle area. Physical examination revealed a small, potato-sized, nontender, fixed mass in the right lateral malleolar area. Laboratory findings were normal. Radiographs of the right ankle showed nodular clustered calcifications with soft tissue swelling in the lateral malleolar area (Figure 1). No evidence existed of a bony abnormality in the fibula or joint space narrowing in the right ankle. Magnetic resonance imaging (MRI) was performed with a 3.0T scanner (Sigma HDxt; GE Healthcare, Milwaukee, Wisconsin). T1-weighted MRI revealed a lesion with intermediate- to slightly high-signal intensity at the lateral aspect of the lateral malleolus of the right fibula (Figure 2A). On T2-weighted MRI, the lesion appeared as an elongated mass with central heterogeneously high-signal intensity and a peripheral rim of low-signal intensity surrounded by an area of diffuse high-signal intensity (Figure 2B). In addition, the lesion mass had a broad base on a thickened dermis. On plain radiographs, the lesion appeared as a nodular mass with low-signal intensity at the lateral margins in all sequences. Contrast-enhanced T1-weighted MRI revealed diffuse reticular and peripheral enhancement (Figures 2C, D). No joint effusion existed in the right ankle, and no signal intensity change was evident in the bone marrow of the distal fibula. The initial radiological differential diagnosis included chronic bursitis with dystrophic calcification, pigmented villonodular synovitis, gout, and rheumatoid nodule. The patient underwent complete excision of the mass lesion.

Macroscopic examination of the excised lesion demonstrated grayish, amorphous, gelatinous masses that contained 1.5×0.5-cm subpidermal cysts and calcifications (Figure 3A). Microscopic examination revealed that the subpidermal cyst was partially lined by abundant keratinous debris; the inferior–peripheral aspects were lined by ghost cells that had a distinct border, eosinophilic cytoplasm, and a central clearing that represented the outline of an absent nucleus. No internal content existed in the subpidermal cyst (Figures 3B, C).

**DISCUSSION**

Pilomatrixoma is a common skin neoplasm in the pediatric population and is often misdiagnosed as one of several dermal disorders.\textsuperscript{5} Although this lesion was originally referred to as a calcifying epithelioma, the etymologically more accurate terms pilomatrixoma or trichomatrixoma have been in general use since 1977.\textsuperscript{6} A pilomatrixoma usually presents as a solitary, superficial, firm, slow-growing, painless nodule in the dermis and ranges in diameter from 0.5 to 3 cm.\textsuperscript{7} Histopathological features include the presence of basoloid and squamoid epithelia and cornified eosinophilic masses containing shadow cells in association with multinucleated histiocytic giant cells, calcification, or ossification.\textsuperscript{8} In 30% of cases, basoloid cells are converted into ghost cells, also called shadow cells, which are more centrally located. These cells have lost their nuclei, are filled with keratin, and stain as eosinophilic, with an unstained area in the center appearing as a shadow due to the loss of the nucleus. The lesion stroma is collagenous and contains dilated blood vessels. Chronic inflammation with a foreign body reaction can be seen in the stroma adjacent to the ghost cells.\textsuperscript{7} Calcium deposition often occurs in the ghost cell region and the stroma, with an incidence of 69% to 85%.\textsuperscript{7}
Various types of calcification, including nodular, diffuse amorphous, and rim-like shapes, were seen on radiographs in 6 (55%) of 11 cases of pilomatricoma. By sonography, pilomatricoma frequently appears as a subcutaneous mass containing internal calcification, internal echogenic foci, and a hypoechoic rim. In addition, internal vascularity exists in 50% of these masses. On MRI, pilomatricoma usually shows isointense-signal intensity in muscle on T1-weighted MRI and inhomogeneous intermediate-signal intensity on T2-weighted MRI. Furthermore, Hsieh et al suggested that the presence of low-signal intensity on T2-weighted MRI corresponds with the distribution of keratin and calcification.

Although the initial report for imaging of pilomatricoma described no enhancement on contrast-enhanced T1-weighted MRIs, most reported cases have shown inhomogeneous enhancement. According to Lim et al, rim enhancement may be seen in all cases. This enhancement difference can be explained by variations in the ratio of the components in a pilomatricoma. In addition, diffuse enhancement and peritumoral strands around the cystic tumor were demonstrated by MRI in the current case. Lim et al reported that MRI findings such as peritumoral edema or inflammation may reflect chronic inflammation with a foreign body reaction.

Kakarala et al reported the cystic nature of pilomatricoma in the preauricular area by contrast-enhanced CT and demonstrated homogeneous low density without contrast enhancement. They reported that the appearance of a cystic lesion on CT or MRI could lead to the misdiagnosis of pilomatricoma as another benign cystic tumor, such as a dermoid or epidermoid cyst.

However, the mass in the current case was not initially considered to have the typical cystic appearance because of the high-signal intensity lesion on T2-weighted MRI, which was interpreted as a cystic portion with a thick wall occupying less than half of the tumor.

Kakarala et al also reported the ultrasound and CT findings of a cystic pilomatricoma with a similar nature located in the infra-auricular area; the cystic portion of the tumor was seen as irregular anechoic areas on ultrasound and as an irregular, poorly enhanced, central area on contrast-enhanced CT. However, the solid portion showed inhomogeneous iso- to high echogenicity in the surrounding tissue on ultrasound and marginal enhancement on contrast-enhanced CT.

Kaddu et al proposed that the natural course of this neoplasm is a chronological process in which the lesion presents as an infundibular matrix cyst at an early stage and that the rapid proliferation of matrix and supramatrix cells in fully developed pilomatricomas results in an increase in the size of the lesion presents as a subcutaneous mass containing internal calcification, internal echogenic foci, and a hypoechoic rim. In addition, internal vascularity exists in 50% of these masses. On MRI, pilomatricoma usually shows isointense-signal intensity in muscle on T1-weighted MRI and inhomogeneous intermediate-signal intensity on T2-weighted MRI. Furthermore, Hsieh et al suggested that the presence of low-signal intensity on T2-weighted MRI corresponds with the distribution of keratin and calcification.

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the lesion with time. This is accompanied by the obliteration of the original cystic configuration so that they are cystic neoplasms rather than true cysts due to the absence of a cyst lining. In the early regressive stage, the tumor appears to have a moderately dense inflammatory lymphohistiocytic infiltrate with histiocytic giant cells. At the late regressive stage, the neoplasm no longer appears cystic.

Because no cystic portion dominated the tumor in the current case, the tumor was probably at the early regressive stage, which is probably the reason the tumor presented with an atypical cystic nature on MRI.

In contrast to reported cases of cystic pilomatricoma with typical clinical presentations, the diagnosis of a cystic pilomatricoma when it occurs in an atypical location or in older patients may be more challenging for musculoskeletal radiologists and orthopedic surgeons who are unfamiliar with the imaging findings of cystic pilomatricoma, which may lead to unnecessary clinical and radiological studies. In the current patient, these atypical MRI features around the ankle joint led to confusion with other diseases, including chronic bursitis, pigmented villonodular synovitis, rheumatoid nodules, and gout.

**CONCLUSION**

The identification of radiological features, such as the presence of calcification in the tumor and absence of joint involvement, could be helpful in differentiating cystic pilomatricoma from other diseases. Due to the relative rarity of calcification in other diseases included in the differential diagnosis, the presence of calcification in the tumor could enable radiologists to establish a definitive diagnosis. In addition, MRI can be useful for differentiating gout and rheumatic arthritis by confirming the absence of bone marrow abnormalities and erosive changes around the ankle joint. Cystic pilomatricoma, although rare, should be considered in the differential diagnosis of soft tissue masses that contain calcification.

**REFERENCES**


