

Case Report

Calcifying aponeurotic fibroma around Achilles tendon: A case report $^{ᡵ, k \times k}$

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ABSTRACT

Calcifying aponeurotic fibroma is an uncommon benign fibrous tumor, typically develops around the fascia and tendons and commonly observed in children and adolescents. It usually occurs in the distal portion of the extremities. Presented as slow growing painless mass, treated with complete surgical excision with high recurrence tendency. We report a case of calcifying aponeurotic fibroma in a 4-year-old girl located posterior to Achilles tendon. Clinical, radiographic, and magnetic resonance imaging findings are described. To the best of our knowledge, this is the first reported case of CAF located posterior to Achilles tendon.

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Introduction

Calcifying aponeurotic fibroma (CAF) was first time described by Keasbey [1]; where it was referred as juvenile aponeurotic fibroma in 1953. It is one of the unusual benign soft tissue masses. It usually occurs at the children and adolescent age group, typically at the 1st and 2nd decades of life, and predominantly in males [2]. Although it has been reported at varying ages and it can arise from different anatomical location as dense fibrous connective tissue mass [3,4]. CAF generally occurs at the distal upper or lower extremities, most commonly involving the deep volar fascia, tendons, and aponeuroses of the hands [4,5].

During literature review, it was found that these CAFs can arise from the neck, abdominal wall, back, knee, thigh, forearm, elbow, and arm connective tissues [2,3,6]. Clinically, CAF usually presents with painless, hard, and slow growing soft tissue mass with high recurrence tendency [7].

Treatment of CAF is usually by complete surgical excision although couple of studies show approximately 50% recurrence rate [8].

We report a case of CAF arising from the soft tissue surrounding the Achilles tendon of a 4-year-old girl. In the

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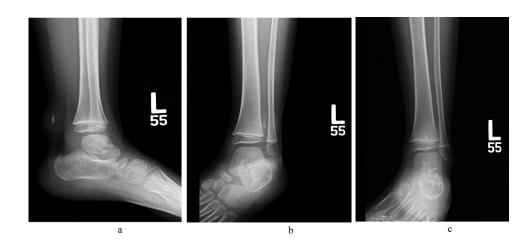


Fig. 1 – Lateral (a), oblique (b), and AP (c) views of the left ankle show there is an ill-defined oval-shaped focal soft tissue lesion along the course of the Achilles tendon associated with central calcifications, probably representing an underlying soft tissue tumor of the Achilles tendon.

present study, a case of CAF arising posterior to Achilles tendon is presented, as well as a review of the literature.

Case report

A 4-year-old, healthy girl, referred to the oncology orthopedic clinic from a secondary hospital with a 1-year history of posterior distal left leg lump, just above the left ankle joint, which gradually over a 2 months period. No history of pain or trauma. No other lumps identified in the body.

On the clinical examination, the lump was hard, slightly mobile, and nontender. The mass approximately measured 2×2 cm.

Ankle radiograph (Fig. 1) shows, ill-defined, oval-shaped focal soft tissue opacity along the course of the Achilles tendon associated with central calcifications.

The ankle MRI showed a well-defined oval-shaped soft tissue mass encasing the posterior aspect of the Achilles tendon. The mass measured $2.4 \times 1.6 \times 1.7$ cm in craniocaudal, transverse, and anteroposterior dimensions, respectively. The mass demonstrates low signal on both T1- and T2-weighted images, with mild heterogeneous enhancement (Fig. 2).

US-guided biopsy was carried out (Fig. 3) for further histopathology assessment.

The anatomical pathology gross specimen was received as multiple fragments of pale tan tissue that measured in aggregate $0.5 \times 0.1 \times 0.1$ cm. The histopathological examination demonstrated a benign fibrous lesion with calcification (Fig. 4), suggestive a CAF. The fibroblasts were positive for smooth muscle actin (SMA) and negative for beta-catenin immuno-histochemical stains.

The patient followed up in the pediatric orthopedic clinic and surgical excision option was provided, but the patient's family refused any surgical intervention and agreed on regular follow-up.

Discussion

CAF typically occur in young age group, "peak incidence, age 8-14 years" [2] Murphey. Most of CAF are usually sorted as a benign fibroblastic tumor however, they have an aggressive behavior with local invasive nature and increased in local recurrence rate after surgical resection is common [1].

In literature, the most common sites mentioned for CAF are the palms and soles of the feet. However, it also affects other less common sites, including the neck, forearm, elbow region, popliteal area, lower back, thigh and knee, which all are areas that are closely related to aponeuroses, tendons or fascia [3].

The usual clinical manifestation is a slow growing painless soft tissue mass, which does not affect the adjacent joint motion [2,9]. In our case, the mass (lump) was present for more than a year with minimal change.

There are only limited reports in the radiology literature describing the imaging appearances of these lesions [2]. On radiography, CAF appears as a soft tissue mass, with fine or course calcification. Signs of bone involvement can be seen, such as periosteal reaction and cortical defect [2]. CT scan is a good modality to visualize the associated calcification. Magnetic resonance (MRI) is the most accurate tool in the evaluation of soft tissue tumors, mainly for preoperative planning and to evaluate the lesion extent and the margins. The tumor signal intensity is likely related to the amount of calcification and the fibrous components [9], according to Cho "CAF was reported to present like heterogeneous mixtures of high and low signal intensity on T2-weighted images, and intermediate on T1-weighted images, with intense contrast enhancement". In our case, the lesion shows typical fibrous imaging characteristics with low T1, T2 signal intensities and mild enhancement in the postcontrast images.

However, it is nonspecific in evaluation of CAF [2]. CAF appearance can be variable from one case to another, depending on the soft tissue involvement and calcification [6]. This

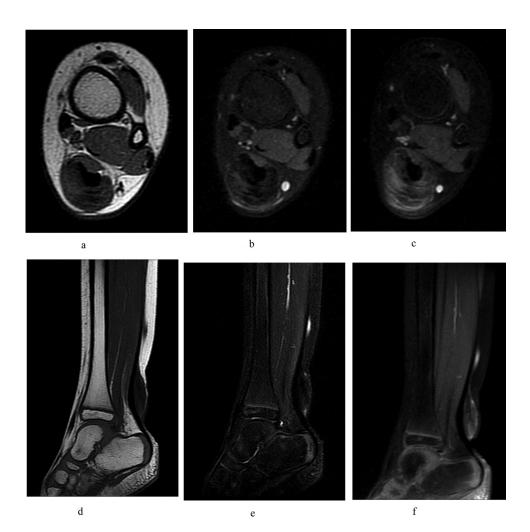


Fig. 2 – Axial T1, T2 fat sat and T1 postcontrast fat sat images of the lesion show in a good advantage the center of the lesion and the posterior encasement of the left Achilles. Low signal intensity on both T1 (a, d) and T2 (b, d) sequences. Mild heterogeneous enhancement in postcontrast images is evident (c, f).

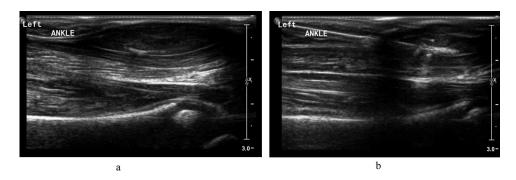


Fig. 3 – (a) Pre core needle insertion, gray scale ultrasound image shows oval-shaped soft tissue mass with small comet tail artifact which suggests presence of calcification. (b) Post core needle insertion, gray scale ultrasound image confirms the presence of the needle within the soft tissue mass.

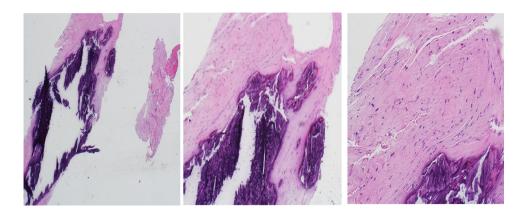


Fig. 4 – Three different magnified histopathological images of the lesion that show benign fibroproliferative process with spotty calcification, suggestive of CAF.

kind of tumor is usually diagnosed by histopathology sampling and only a histopathological examination can provide a definitive diagnosis of CAF [10]. Unfortunately, histopathological examination of CAF can be mistaken for a fibroma or dermatofibroma, mostly due to abounded fibrous tissue components [8] and it is crucial to differentiate it from other soft tissue tumors, mainly synovial sarcomas [2].

CAF has an ill-defined contour, varying from rubbery to firm consistency. It is gray-white mass in gross pathology. The lesion usually has abundant calcification surrounding areas of fibrous tissue containing spindle-shaped fibroblast [2].

Resection is the initial treatment of choice with close follow-up [9]. However, local recurrence usually is seen in more than half of the cases.

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