

## Painful calcifying aponeurotic fibroma

Sir,

Calcifying aponeurotic fibroma is a rare soft tissue tumor that presents as a painless mass on acral sites in children and adolescents.<sup>[1]</sup> Conservative surgical excision is both diagnostic and therapeutic. We describe a case of painful calcifying aponeurotic fibroma that developed after a burn injury on the left foot.

A 22-year-old man presented with a firm, tender, 2.5 × 2.5 cm skin-colored nodule with well-defined margins on the mid-plantar aspect of his left foot [Figure 1a]. The overlying skin was unremarkable and non-adherent to the mass which was fixed to the underlying soft tissue structures. The lesion had gradually enlarged over the last 14 years and was painful. There was history of a burn injury at the same site, two months prior to the onset of the lesion. The differential diagnoses of nodular fasciitis and post-traumatic neuroma were considered.

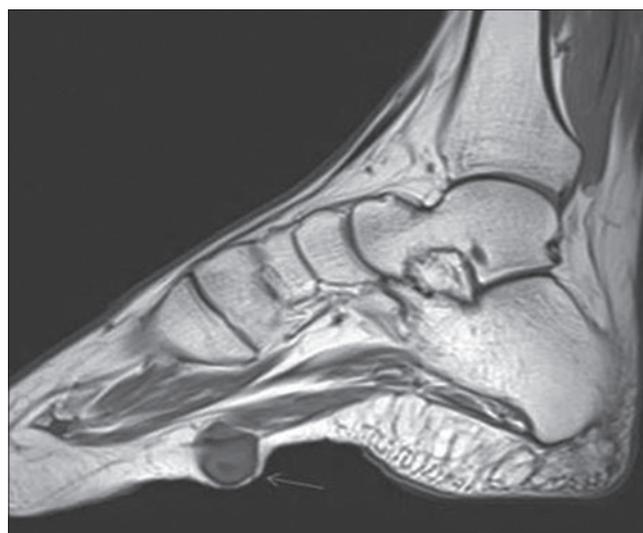
Fine needle aspiration cytology revealed fascicles and scattered spindle-shaped cells embedded in an eosinophilic matrix. The cells had plump, elongated nuclei with vesicular chromatin and scant to moderate cytoplasm. The features were suggestive of a benign spindle cell tumor such as a fibroma or schwannoma. Contrast enhanced magnetic resonance

imaging demonstrated a 1.8 cm × 1.7 cm sized well-defined lesion (T1 hypointense, T2 hyperintense) in the subcutaneous plane on the plantar aspect of the mid-foot [Figure 1b]. Some T2 hypointensities were seen within the lesion. It also showed mild post-contrast enhancement. Superiorly, the tumor indented the plantar muscles and fascia with loss of fat planes while the bones were normal. The patient was referred to the plastic surgery service for wide local excision and reconstruction. The defect was reconstructed using a local hatchet flap from the non-weight bearing instep area of the sole. Excisional biopsy revealed a spindle cell tumor arising from the aponeurosis in the deep subcutaneous tissue. The tumor cells were arranged in fascicles and had bland spindle-shaped nuclei and moderate eosinophilic cytoplasm. Foci of calcification and ossification were observed [Figure 2a and b]. Focal areas of increased mitosis were seen with a mitotic count of 1/10 high-power fields [Figure 3a]. However, the Ki-67 index was below 1% [Figure 3b] and there was no significant nuclear atypia. Based on these findings, a diagnosis of calcifying aponeurotic fibroma was made. No recurrence was evident at follow up one and a half years after excision.

Calcifying aponeurotic fibroma, classified under superficial musculoskeletal fibromatosis, is an uncommon, locally aggressive, soft-tissue tumor originating from fibroblasts.<sup>[2]</sup> Initially designated as juvenile aponeurotic fibroma, the name was changed as it affects older age groups as well.<sup>[1]</sup> The peak incidence of this tumor occurs between 8 and 14 years



**Figure 1a:** Well-defined skin-colored nodule measuring 2.5 cm × 2.5 cm on the mid-plantar aspect of left foot

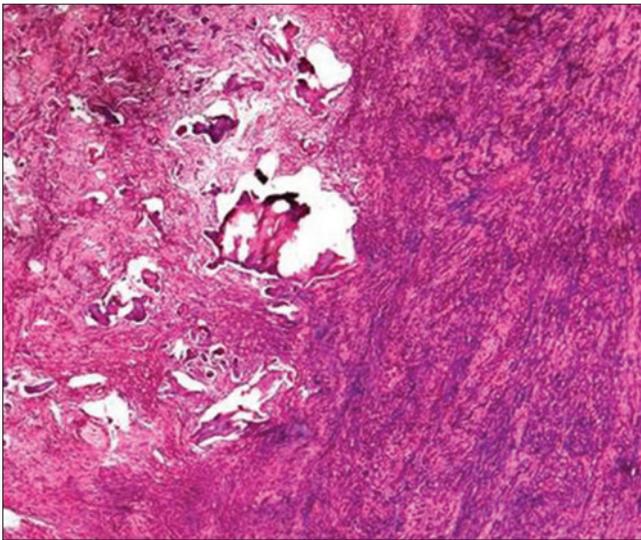


**Figure 1b:** Magnetic resonance imaging (T1-weighted, sagittal) of the foot showing well-defined soft tissue mass on the plantar surface (arrow)

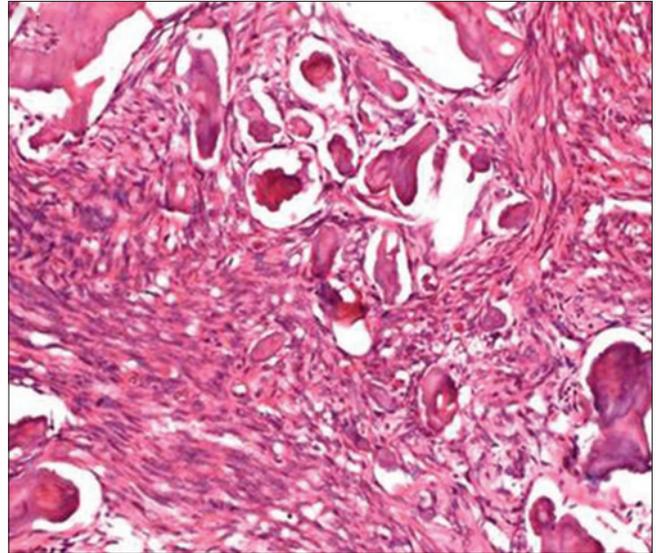
of age.<sup>[3]</sup> The sites of predilection include the hands and feet with almost 77% of reported cases involving the palms and fingers and 13% involving the feet. Other sites may also be affected.<sup>[3-5]</sup> There is usually no preceding history of trauma. The typical clinical presentation is as an ill-defined, slowly progressing, firm, subcutaneous mass, measuring <3 cm in diameter, free from the overlying skin but adherent to the underlying soft tissue. Adjacent bone involvement may rarely occur. Although usually asymptomatic, significant pain has been associated with the tumor.<sup>[6]</sup> Two growth phases are described: in the early diffuse phase, it is infiltrative and devoid of calcification and the late compact phase is characterized by well-defined borders, calcification and cartilage formation.

Magnetic resonance imaging and computed tomography are the preferred imaging modalities for diagnosis and delineating the extent of the lesion. Magnetic resonance imaging findings include an ill-defined soft-tissue mass with speckled calcification, iso- to low signal intensity on T1-weighted images, heterogeneous high signal intensity with minor areas of iso- to low signal intensity on T2-weighted images and heterogeneous post-contrast enhancement.<sup>[7]</sup>

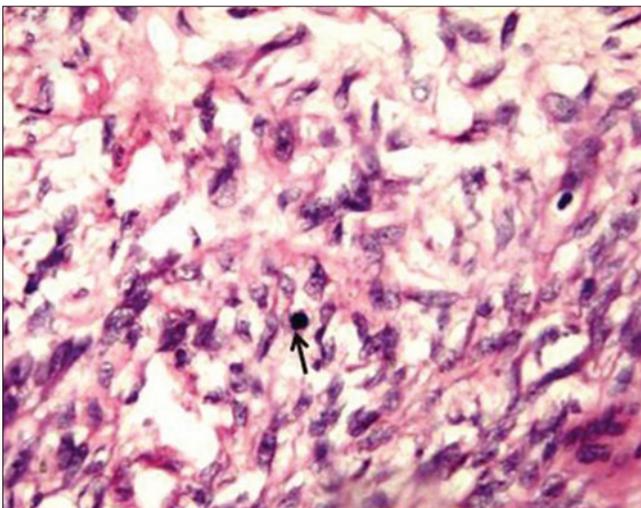
The definitive diagnosis of calcifying aponeurotic fibroma is made on histopathology which demonstrates a poorly encapsulated tumor composed of parallel-oriented spindle-shaped fibroblasts with indistinct cytoplasm and round to ovoid nuclei arranged in fascicles around central



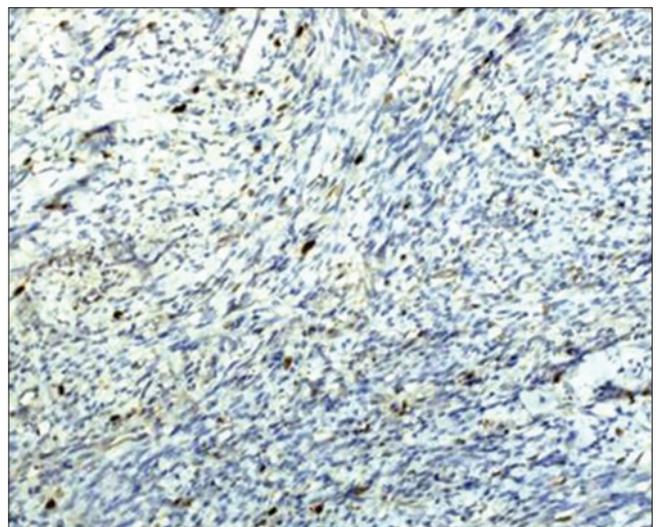
**Figure 2a:** Short fascicles of spindle cells in a sclerotic background with interspersed foci of calcification and ossification (H and E, x40)



**Figure 2b:** Tumor cells showing elongated nuclei, moderate eosinophilic cytoplasm along with intervening foci of calcification and ossification (H and E, x200)



**Figure 3a:** Higher magnification depicting spindle-shaped fibroblasts with one mitotic figure (arrow) (H and E, x400)



**Figure 3b:** Nuclear positivity in the spindle cells (Ki-67, x40)

**Table 1: Differential diagnoses of calcifying aponeurotic fibroma in an adult**

Entity	Clinical features	MRI findings
Calcifying aponeurotic fibroma	Asymptomatic, ill-defined, slowly progressing, firm, subcutaneous mass, free from overlying skin but adherent to underlying soft tissue, on hands/feet	Ill-defined mass with speckled calcification, iso- to low intensity on T1W1, heterogenous high intensity with minor areas of iso- to low intensity on T2W1 and heterogenous post-contrast enhancement
Giant cell tumor of the tendon sheath <sup>[8]</sup>	Asymptomatic slow-growing mass on the fingers in second to fourth decades	Lobulated and well-defined margins with hemosiderin deposits, close association with the tendon sheath, uniform contrast enhancement, pressure erosions in adjacent bones, absence of calcification
Soft tissue chondroma <sup>[9]</sup>	Slow-growing painless lesion on the fingers in adults more than 30 years of age	Metaplastic cartilage is more well-developed compared to CAF. Low intensity on T1W1 and very high intensity on T2W1. Intra-lesional low intensity may reflect calcification
Fibroma of the tendon sheath <sup>[9]</sup>	Well-defined slow-growing hard nodular mass	Well-defined mass with decreased signal intensity on all sequences and some or no contrast enhancement
Plantar fibromatosis <sup>[1]</sup>	One or more nodules which may become painful and ulcerate, mostly on the medial half of mid-foot, after 20 years of age	Absence of calcification
Nodular fasciitis <sup>[9]</sup>	Non-neoplastic rapidly growing reactive lesion usually on the forearm in the second to fourth decades	Non-specific. Intensity slightly greater than or equal to muscle in T1W1, intermediate to hyper intense on T2W1, diffuse or peripheral post-contrast enhancement, linear extensions along the superficial fascia and mild surrounding edema
Myositis ossificans <sup>[10]</sup>	Painful swelling that occurs mainly on the proximal aspect of limbs such as elbow, thigh, shoulder or buttocks usually after trauma	Early stage: heterogeneous mass with poorly defined margins, high intensity on T1W1; fluid levels corresponding to hemorrhage may occur. Intermediate stage: rim of mineralization appears as a curvilinear hypointensity on all imaging sequences. Mature lesions: high intensity due to fatty proliferation with sharp hypointense margin on T1W1 and low intensity on T2W1 due to hemosiderin deposition
Fibro-osseous pseudotumor <sup>[11]</sup>	Painful swelling usually involving the fingers and less commonly the toes in young females, history of antecedent trauma in 40% cases	Subcutaneous well-circumscribed soft-tissue mass, heterogenous appearance with areas of high and intermediate intensity on T2W1, intermediate intensity on T1W1, signal void foci that represent calcification or blood products within the mass

CAF: Calcifying aponeurotic fibroma, MRI: Magnetic resonance imaging, T1W1: T1-weighted images, T2W1: T2-weighted images

areas of calcification. The cells are enmeshed in a dense collagenous stroma. Foci of chondroid differentiation and osteoclast-like giant cells are also observed. Mature bone formation has also been described in a long-standing lesion.<sup>[5]</sup> Occasional mitotic figures may be detected, but nuclear atypia is not seen.

The differential diagnoses of calcifying aponeurotic fibroma in adults are discussed in Table 1.

The treatment of choice for the tumor is local excision including a margin of surrounding normal tissue. The probability of recurrence is 50%, due to its poorly circumscribed growth pattern. Malignant transformation, though rare, has been described.<sup>[12]</sup> Strict follow-up is recommended and repeat surgical excision is required in case of recurrence.

The present case was unusual as the tumor was extremely painful and developed after a burn injury to the affected site. This is in contrast to the asymptomatic nature of the tumor in the majority of cases and its *de novo* occurrence described in previous reports. Further, the tumor demonstrated prominent foci of

calcification and ossification that may be attributed to its prolonged duration.

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#### Conflicts of interest

There are no conflicts of interest.

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