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CASE REPORT

Calcifying aponeurotic fibroma of the distal phalanx

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KEYWORDS
Calcifying aponeurotic fibroma;
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Summary Calcifying aponeurotic fibroma is a rare benign soft tissue tumor that primarily occurs on the distal portion of the extremities of children and adolescents. It appears like a firm, painless and slowly growing mass with high local recurrence rates. The lesion has characteristic histological features with areas of proliferative plumps of fibroblasts, chondrocytes and foci of calcification.

We present a case of calcifying aponeurotic fibroma of the sub-ungual area of the index finger distal phalanx with bone erosion, surgically treated. A 2 year follow up showed satisfactory functional result and no evidence of recurrence.

Introduction

Calcifying aponeurotic fibroma (CAF) is a rare benign soft tissue tumor that primarly occurs on the distal portion of the extremities of children and adolescents. However, it also affects other less common sites including knee, thigh, elbow, neck, abdominal wall and mandible.[1,2] It appears essentially like a firm, soft tissue mass which tends to infiltrate the surrounding fat, muscle tissue and rarely the bone and has a high rate of local recurrence after surgical treatment.[3,4] Histopathological features include the presence of proliferative fibroblasts, chondrocites and foci of calcification. We present a case of a 44 years old male with a calcifying aponeurotic fibroma of the sub-ungual area of the distal phalanx of the index finger with bone erosion.

Case report

A 44 years old male patient presented to our clinic with a nodular mass of the tip of his right index finger in the sub-ungual area (Figure 1). The patient reported that he had the lesion since two years. In the last month, according to the patient history, the lesion had grown up with pain and swelling. After a clinical examination, plain radiographs of the hand revealed a nodular mass with foci of calcification and bone erosion of the distal phalanx of the index finger (Figure 2). Surgical exploration under local anesthesia was performed. After a ring-block with lidocaine without adrenaline, an excision of the lesion was performed including 5 mm of the distal portion of the distal phalanx

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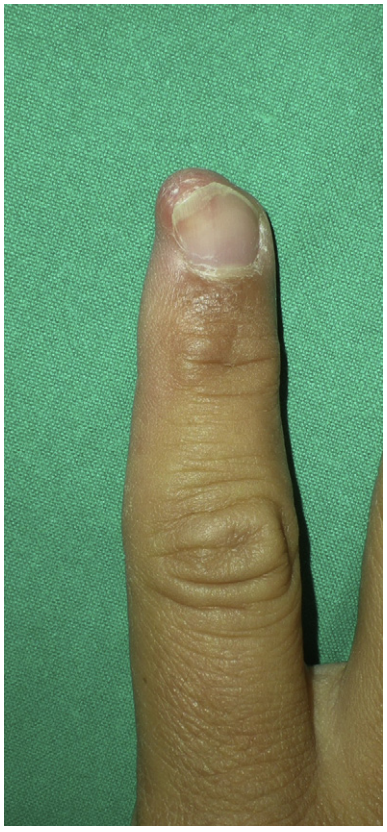

Figure 1 Presence of a firm nodule on the right index fingertip.

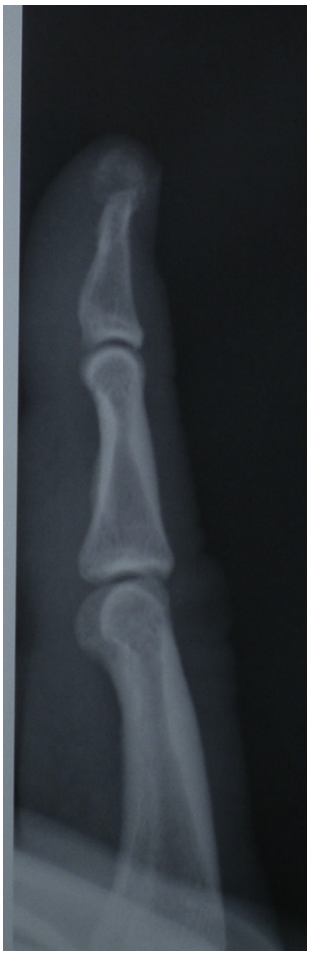

Figure 2 X-ray of the right index finger showing distal bone involvement.

that appeared clinically involved (Figure 3). Closure of the defect was obtained with advancement of an inferior fingertip skin flap. To allow re-growth of the residual nail, the nail bed was splinted with the previously removed nail. Histology revealed the presence of proliferative fibroblasts with the absence of cytologic atypia and mitotic figures in a myxoid stroma rich of calcification and concentric lamellar bone. Osteoclast-like cells were also present.

There wasn't any post-operative complication and a 2 year follow up showed satisfactory functional result and no evidence of recurrence (Figure 4).

Discussion

Calcifying aponeurotic fibroma was first described by Keasbey in 1953 as "Juvenile Aponeurotic Fibroma".[4] Since the first report, less than 100 cases have been published in the literature. CAF is mostly seen in children with the peak of incidence between the ages of 8-14[5] although few cases have been reported in adults. Most common sites include the palm of the hand and the sole of the foot. Other less common sites are neck, mandible, forearm, elbow, lower back, thigh and knee.[1–6] So far, no case of CAF on the subungual region has been described or reported. CAF has classically been described as a non tender, slow growing, soft tissue mass measuring usually less than 3 cm in diameter. Rarely this lesion can cause pain and discomfort. Surrounding normal tissue can be incorporated within the lesion and significant calcifications may be identified upon palpation.[2]

The radiographic features of CAF include the presence of a soft tissue mass of varying radiodensity with the presence of foci of calcification due of the mineralization of the lesion. However X-ray imaging is not useful for the diagnosis because it's not specific. Magnetic resonance and computerized tomography are the imaging modalities of choice for CAF. Computer tomography reveals a non-specific soft tissue mass with stippling of calcification and can show the infiltrative growth pattern of the lesion into the surrounding tissues. Magnetic resonance can contribute to preoperative differential diagnosis of calcifying aponeurotic fibroma from other fibrous tumors, giant cell tumor of the tendon sheat, or soft tissue sarcoma.[7]

Histological examination is necessary for the diagnosis. Microscopical features of the lesion are characteristic. There are foci of calcification and chondroid areas surrounded by proliferative plumps of fibroblasts with round or oval nuclei. Mitotic figures are rare. In some cases there are also multinuclear giant cells around calcified area. Ossification is rare. Keasbey and Fanselau[8] noted the relationship between the age of the patient and the characteristic of the tumor. These authors suggested the existence of two phases in the development of the tumor: an initial phase,

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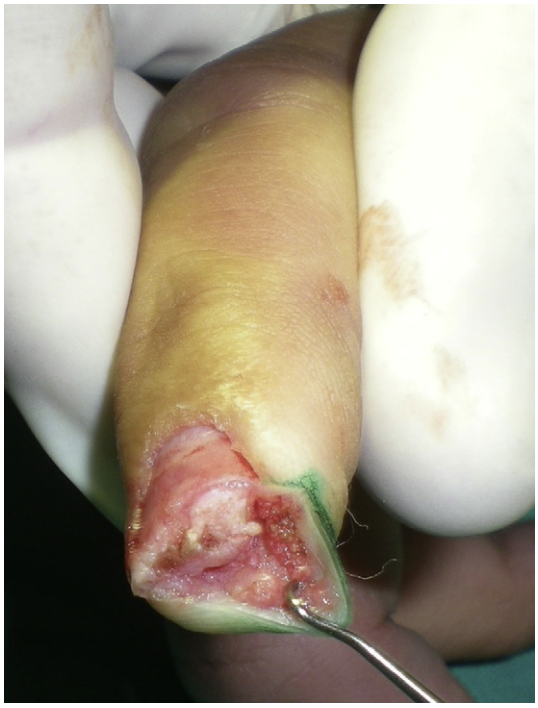

Figure 3 Peri-operative picture showing bone infiltration.

which is more common in infants, with few calcifications corresponding to a more aggressive tumor, and a late phase, in which the lesion shows a diffuse calcification and displays a more compact aspect with well defined borders.

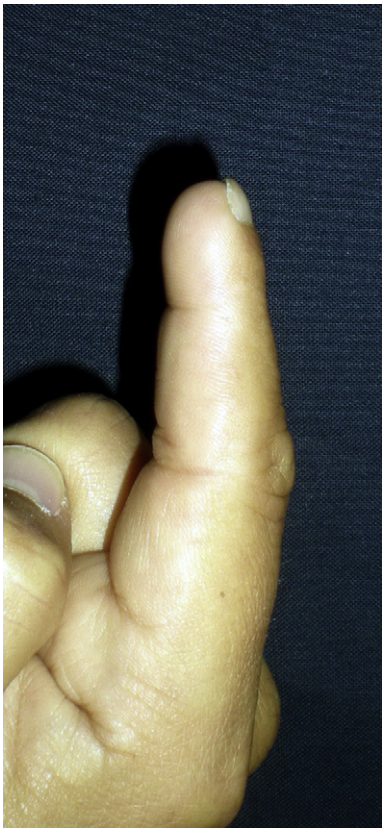

Figure 4 Follow up at 2 years.

Allen and Enzinger[9] noted a rather homogeneous pattern in all cases examined with no evidence of different phases in CAF maturation. Local recurrence is very high and occurs in more than half of the cases especially in infants and in cases of incomplete resection. Malignant transformation is very rare. Lafferty et al[10] reported a case of a calcifying aponeurotic fibroma with mestastatic fibrosarcoma of the lungs and the bones.

Conclusions

We report this case for its unusual localization. Keeping into consideration the literature, we should have requested MRI imaging preoperatively. In consideration of the high incidence of recurrence, radical surgical excision of CAF is recommended. However, considering the benign characteristic of the lesion and the rare metastases, a strong effort should be made, whenever possible, to preserve the function of the involved extremities. A strict follow-up is strongly recommended and in the case of local recurrence a complete surgical resection should be performed.

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Conflict of interest

None.

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