CASE REPORT

Calcifying aponeurotic fibroma of the elbow : a case report

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Abstract : Calcifying aponeurotic fibroma is a rare soft tissue tumor that typically occurs in the distal extremities of children and adolescents. Because a calcifying aponeurotic fibroma is an ill-defined mass and has a tendency to infiltrate the surrounding tissue, local recurrence rates are 50%. We report a case of calcifying aponeurotic fibroma occurring in the elbow region, an uncommon site. The patient is followed up for 6 years without local recurrence of the tumor. J. Med. Invest. 58 : 159-162, February, 2011

Keywords **:** *calcifying aponeurotic fibroma, elbow, MRI, CT*

INTRODUCTION

Calcifying aponeurotic fibroma (CAF) is a rare benign soft tissue tumor that typically occurs in the distal extremities of children or adolescents. The most common sites for CAF are the palms and soles of the feet. Until date, only two cases of CAF of the elbow region have been reported (1). Here we report CAF arising in the elbow region of a 10-year-old female without local recurrence after an operation.

CASE REPORT

A 10-year-old girl was referred to our hospital for a tumor on her left elbow. She had been aware of this slowly growing mass since the past 5 years. Physical examination showed an indolent and firm mass measuring 10 mm×10 mm overlying the flexor aspect of the left elbow joint. The overlying skin did not show a dimpling sign fixed to the mass. The patient had no complaints of motor dysfunction or sensory disturbance.

A computed tomography scan showed that the mass extended from the subcutaneous tissue to the radius (Fig. 1). The density of the mass was equal

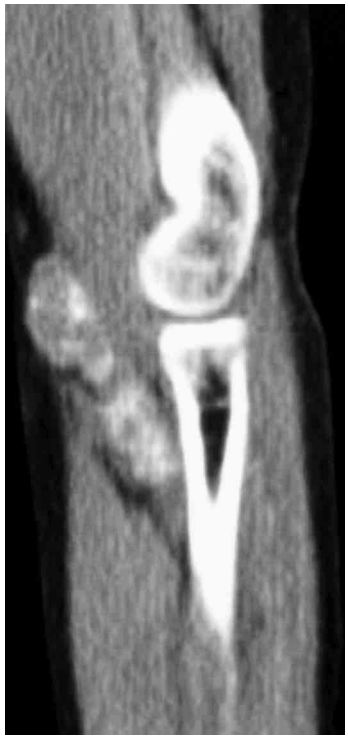

Fig. 1 Preoperative computed tomography images of the left elbow region. Stippled calcification of the mass was obvious. The mass extended from the subcutaneous tissue to the radius.

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to surrounding muscles with diffuse calcification. Contrast-enhanced magnetic resonance imaging (MRI) showed a heterogeneously enhanced soft tissue mass measuring 10 mm×10 mm×50 mm (Fig. 2). This tumor seemed to be attached to the distal part of the biceps brachii tendon and to be relatively well defined. A T1-weighted image showed low signal intensity, whereas a T2-weighted image showed low- and high-signal intensity areas of the mass. The diagnosis supported by these radiological findings was a hemangioma. In contrast, color Doppler ultrasound findings showed avascularity of this mass.

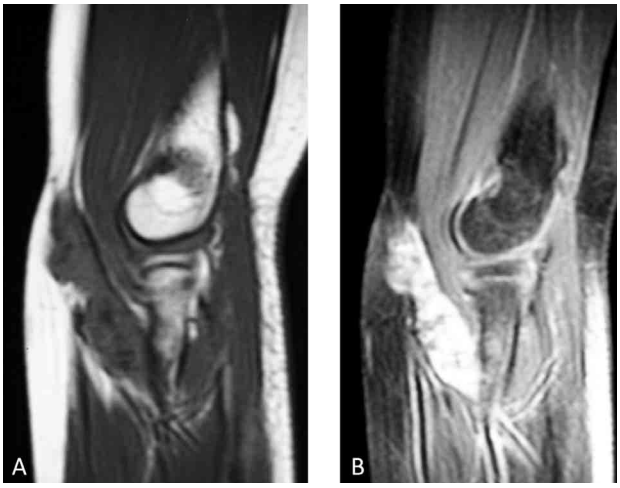

Fig. 2 Magnetic resonance imaging of the left elbow region. T2-weighted image demonstrated heterogenously low-intensity area of the mass.
A, Sagittal T1-weighted image. B, Sagittal T2-weighted image.

The tumor was resected for a definitive diagnosis (Fig. 3). The tumor extended to the biceps brachii tendon, the periosteum of the radius, and the brachial artery but was easily dissected from the surrounding tissue except for the biceps brachii tendon.

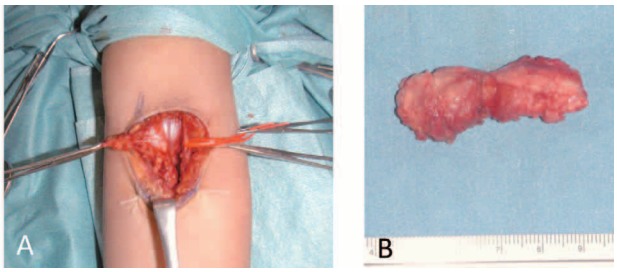

Fig. 3 Intraoperative view of the resected material. A. The tumor was attached to the biceps brachii tendon. Dissection from the radial nerve branch and radial artery was easy. B. The biceps brachii tendon was partly excised with the tumor.

This tendon was resected partially together with the mass. On intraoperative observation, the mass was relatively well defined, firm, and whitish.

Microscopical section revealed a fibroblast-like spindle cell neoplasm with a configurated palisaded sequence surrounding scattered calcification foci (Fig. 4A). The tumor cells had invaded the surrounding fatty tissue and muscle (Fig. 4B). No necrotic area was observed in the tumor. The cells showed no atypical nuclei or mitotic activity (Fig. 4C). These findings supported a pathological diagnosis of CAF.

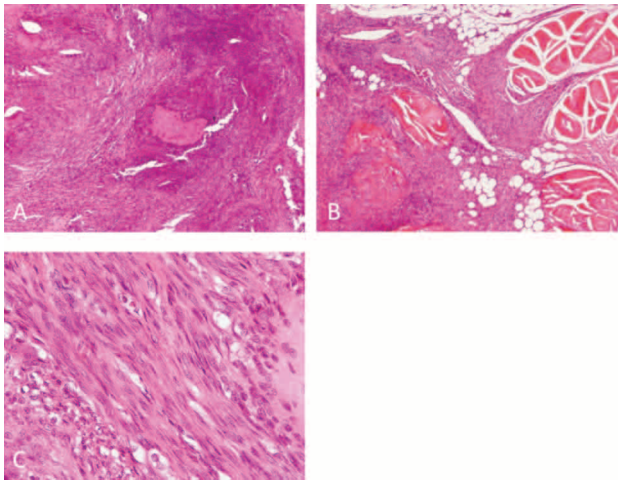

Fig. 4 Light microscopy (hematoxylin and eosin, 40 x). A, Nuclear palisading surrounding the calcified foci was seen. B, Fibroblastic spindle cells invaded the surrounding muscle and fat tissue. C, At higher magnification (200 x), dense spindle cells are aligned linearly. Mitotic figures are not obvious.

We have been observing the patient twice a year for 6 years after the operation, and no recurrence has been found on palpation or on MRI scans.

DISCUSSION

CAF, which was first described in 1953 by Keasbey (2) as juvenile aponeurotic fibroma, is a rare benign soft tissue tumor. Since the first report, fewer than 150 cases have been published in the literature. CAF typically occurs in the first or second decades of life, although cases have been reported for ages ranging from birth to 64 years of age. The median age at the time of diagnosis is 12 years. This tumor typically occurs on the fingers and the palmer and plantar aspects of the foot. Enzinger

and Weiss (3) stated that 90% of CAFs occur on the distal portion of the extremities. Rarely, case reports have shown that CAF arises on the back, knee, thigh, forearm, elbow, and maxillofacial region (4-9). To our knowledge, only two cases of a CAF elbow lesion have been reported (4).

The characteristic clinical findings of CAF are that it is a slow growing, painless, and presents as a firm mass that is not fixed to the overlying skin. Our patient had these distinctive clinical findings. The size of a CAF is typically less than 3 cm in diameter, but the tumor in our case was having an elongated shape as if it had enlarged along the intermuscular space ; it was 5 cm longitudinally.

Radiograph findings of this tumor usually show calcification. Morii *et al*. (10) reported that T1-weighed images of CAF showed isointensity or low intensity, and that T2-weighed images showed heterogeneously high signal intensity with minor areas of isointensity to low signal intensity. CAF and a giant cell tumor of the tendon sheath (GCTTS) usually arise close to the fascia and tendons. However, GCTTS shows a lobulated and well-defined margin with uniform enhancement after the administration of contrast agent, whereas CAF shows an ill-defined, speckled calcification, and heterogeneous gadolinium enhancement on MRI (10, 11). A soft tissue chondroma is also included in the differential diagnosis of calcifying tumors. These tumors have a well defined, lobulated, and diffuse calcification (10). In our case, the MRI findings of a relatively well-defined margin, no lobulation, diffuse calcification, and heterogeneous gadolinium enhancement were not typical findings of a CAF, GCTTS, or soft tissue chondroma.

Histopathologically, CAF is composed of plump fibroblasts with round and ovoid nuclei. The tumor cells have a tendency to extend into the surrounding fatty tissue and muscle. Scattered calcification foci surrounded by tumor cells with nuclear palisading occur, and osteoclast-like multinucleated giant cells are occasionally adjacent to the calcified foci (1, 2, 12). Despite dense cellularity, mitotic activity is unremarkable. Our case presented all of these pathological characteristics of CAF. The nuclear division count was 1 per 20 high-power fields.

Local recurrence rates are more than 50% (2, 12), which could be caused by the tendency of this tumor to infiltrate the surrounding tissue. In fact, we found tumor cells at the margin of our specimen, but no local recurrence has occurred in 6 years postoperatively.

Some authors have described the biphasic features of CAF (2, 12). In the initial phase, the tumor has infiltrative and destructive growth and often lacks calcification. In the later phase, the tumor becomes more compact and nodular with well-defined borders, and diffuse calcification becomes obvious. The tendency for a decrease in cellularity and an increase in collagen stroma with age may cause this tumor maturation. No apparent local recurrence had occurred despite an incomplete resection in our patient, who was 10 years old at the time of the operation, which may have been associated with maturation of the tumor cells.

We herein reported a case of CAF in the elbow region. The patient is still followed up at our hospital because the local recurrence rate of CAF is very high. To our knowledge, only one case of malignant transformation of CAF has been reported till date (13). If the local recurrence occurs, surgical resection and definitive histopathological diagnosis may be necessary.

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