We present an unusual case report of an aggressive, recurrent calcifying aponeurotic fibroma of the thumb in an adult man with invasion into the distal and proximal phalanges, the skin, the radial and ulnar neurovascular bundles, and the tendons, treated with amputation and an immediate toe-to-thumb transfer. (J Hand Surg 2011;36A:110–115. © 2011 Published by Elsevier Inc. on behalf of the American Society for Surgery of the Hand.)

Key words  Calcifying aponeurotic fibroma, thumb, recurrence.

A  CALCIFYING APONEUROTIC FIBROMA was first described in 1953 by Keasbey.1 It is typically described as an indurated, painless, locally aggressive, soft-tissue neoplasm that involves the hands and feet of children. It often infiltrates into surrounding fascia, muscle, and tendons with a 50% predilection for recurrence after surgical excision.2 Bone involvement is very rare, with few case reports in the literature.3,4 Conservative surgical resection as both a therapeutic and a diagnostic intervention is the only treatment recommendation published in the literature.5 There have been no reports regarding treatment of recurrence or reconstruction if the tumor involves critical tissues.

We present a case report of an aggressive, recurrent calcifying aponeurotic fibroma of the thumb in an adult man with invasion into the distal and proximal phalanges, the skin, the radial and ulnar neurovascular bundles, and the tendons, treated with amputation and an immediate toe-to-thumb transfer.

CASE REPORT

History

A 44-year-old man presented with a large indurated mass circumferentially involving the region overlying the distal and proximal phalanges of his nondominant right thumb. Approximately 7 to 8 years before his presentation, the patient had a biopsy-proven calcifying aponeurotic fibroma excised from the ulnar aspect of his right thumb interphalangeal (IP) joint. The patient reported that after a 2-year period a new mass appeared in the same location. The mass did not affect him symptomatically or functionally, therefore he did not seek medical attention for 6 years. He eventually developed symptoms and presented to our institution with complaints of a chronic open sore, numbness and tingling to the thumb pulp, pain over the dorsum of the distal and proximal phalanges, and pain with IP joint motion.

Physical examination

Examination of the right thumb revealed a circumferentially enlarged thumb, relative to the contralateral side, with an indurated, lobulated mass present over the distal and proximal phalanges (Fig. 1). The patient had good capillary refill and a 2-point discrimination of 5 mm. He could flex the IP joint 15° passively and actively. The contralateral normal thumb dimensions were measured as a template by which a great toe could be reduced to create an aesthetically pleasing reconstructed thumb. Preoperative measurements were taken of the thumbs bilaterally and the great toe.

Imaging

Radiographs revealed a large calcified mass surrounding the IP joint of the right thumb with erosion into the underlying proximal and distal phalanges (Fig. 2). There was swelling of the surrounding soft tissues.
Magnetic resonance imaging (MRI) revealed a multilobulated soft tissue mass surrounding the IP joint with circumferential scalloping and invasion of the proximal and distal phalanges. The mass demonstrated a heterogeneous decreased T1 and increased T2 signal with minimal enhancement after administering gadolinium. It measured approximately 3.0 cm in the greatest anteroposterior dimension and 2.5 cm in the greatest proximal-distal dimension. The mass encased the extensor mechanism at its insertion and the neurovascular bundles (Fig. 3).

**Histology**

The intraoperative preliminary frozen sections confirmed the diagnosis of calcifying aponeurotic fibroma.
made in the initial surgery 7 to 8 years previously. The margins were clear of any tumor, and there was no evidence of malignancy.

**Treatment options**

Given the size of the right thumb mass and the aggressive nature of this recurrent lesion, the treatment options included continued observation, lesion debulking, amputation and closure, and amputation with immediate or delayed reconstruction. Observation would permit continual invasion of the mass into surrounding soft tissue and bone, becoming increasingly more symptomatic and debilitating with time.

Considering that this aggressive lesion had recurred after an initial debulking procedure, recurrence would be probable with a second debulking procedure. In addition, debulking such a large mass circumferentially might threaten the vascularity to the thumb.

Because of the extensive involvement of the lesion with the surrounding soft tissue and osseous structures, an amputation would ensure excision of the lesion in its entirety. Intraoperative frozen sections would confirm that the margins were free of residual tumor and if there were evidence of malignancy. After the amputation, the amputation defect could be covered with a soft tissue flap in preparation for a later thumb reconstruction. Alternatively, an immediate free toe-to-thumb transfer could be performed.

We opted to treat this recurrent calcifying aponeurotic fibroma of the thumb with an amputation through the base of the proximal phalanx, and provided that the margins on intraoperative frozen sections were negative, to proceed with an immediate reconstruction using a free trimmed great toe-to-thumb transfer, as previously described by Wei et al.² (Fig. 4). The patient tolerated the procedure well. There were no complications perioperatively.

One year postoperatively, the patient was assessed. He had a healed thumb wound and evidence of nail regrowth and subjective return of sensation. At this time, the 2-point discrimination was 8 mm. He had near normal motion at the metacarpophalangeal joint (MCP) and the carpometacarpal joint. The patient’s newly reconstructed thumb had opposition to the small finger, 0 to 45° of MCP joint movement, 0 to 5° of IP joint motion, 25° to 60° palmar abduction, and 22° to 57° of radial abduction. Radiographs demonstrated union of the toe phalanx to the proximal portion of the thumb phalanx (Fig. 5). The toe donor site had healed completely without residual dysfunction.

**Final pathology**

The final histopathology confirmed a calcifying aponeurotic fibroma with clear margins. There was a multinodular proliferation of mononuclear and giant cells. The individual mononuclear cells had an ovoid to spindle nucleus with finely dispersed chromatin and small to inapparent nucleoli. There were areas of central cartilaginous metaplasia and dystrophic calcification (Fig. 6).

**DISCUSSION**

Calcifying aponeurotic fibromas are painless soft tissue neoplasms common in children, although there are documented cases from birth⁷ to 64 years.⁸ They typically infiltrate into surrounding soft tissue and rarely into bone. Although the literature has characterized this lesion radiologically, definite diagnosis is confirmed with a histopathological examination.

Radiologically, a calcifying aponeurotic fibroma typically shows a soft tissue mass with no associated osseous lesions and a fine stippling of focal calcification. Occasionally scalloping of the cortex and thickening of the bone has been reported.⁹,¹⁰

Morii et al.¹¹ identified several MRI features seen in a calcifying aponeurotic fibroma. In addition to being distributed subcutaneously, being ill-defined, and having a tendency to infiltrate into or adhere to the surrounding tissue, the masses were of isointensity to low intensity on T1-weighted images. The T2-weighted images showed heterogeneous high signal intensity with minor areas of isointensity to low signal intensity. These findings are consistent with the radiologic findings in our case patient.

Histopathology of a calcifying aponeurotic fibroma confirms the diagnosis. Onak-Kandemir et al.¹² found these lesions to have calcified and chondroid central areas and surrounding areas of fibroblastic proliferation. Plump fibroblasts with round and oval nuclei are separated with dense collagenous stroma. Mitotic figures are rare.

Cytological features of such lesions were described by Tai et al.⁵ Fine-needle aspiration biopsy revealed highly cellular, abundant, benign-appearing spindle cells with small oval nuclei, fine chromatin, smooth nuclear contours, and small chromocenters. Chondroid cells, multinucleated giant cells, fragments of myxoid stroma, several small blood vessels, and coarse calcified debris were identified.
The recurrence rate after surgical excision is generally reported as 50%. Carroll documented a recurrence in 8 of 13 patients (62%), whereas Allen and Enzinger reported that 10 of their 25 patients (40%) had a recurrence. The tumor-free interval ranged from 6 months to 23 years.

This case report was unusual in that it involved a large (2.5 to 3.0 cm), aggressive, recurrent calcifying aponeurotic fibroma invading the surrounding soft tissue and bone of an adult man’s thumb. The patient presented with functional and cosmetic complaints. After a thorough discussion of his treatment options, the surgeons in conjunction with the patient elected to perform an amputation of the affected thumb with confirmation of negative margins on intraoperative frozen sections followed by an immediate trimmed great toe-to-thumb transfer.

An amputation of the thumb ensured that the lesion was completely excised and provided the best attempt at preventing recurrence. The advantage of an immediate free toe-to-thumb transfer is that in a one-stage procedure the great toe can be trimmed and tailored to reconstruct the amputated thumb, giving a good cosmetic and functional outcome. After the amputation of the thumb through the base of the proximal phalanx, the surrounding tissue and neurovascular bundles had fresh-cut edges and were free of scar, thus making the immediate reconstruction technically more straightforward.

The disparity of the large great toe compared with the smaller-sized thumb was accommodated for by trimming the great toe to the circumference of the contralateral normal left thumb. This provided a cosmetically acceptable reconstruction of the thumb. Given that the MCP joint was preserved, the trimmed great toe was affixed to the remaining base of the proximal phalanx, thereby preserving the range of motion of the MCP joint and maintaining function.

**FIGURE 4:** Postoperative result after a trimmed toe-to-thumb transfer. A Volar abduction of the thumb. B Opposition of the thumb.
Ray et al.\textsuperscript{14} reported on 6 patients who underwent an immediate reconstruction of a traumatic, nonreplantable thumb amputation by a great toe transfer. They stated that the donor site was acceptable,\textsuperscript{6,15} the rehabilitation was faster,\textsuperscript{16} and an immediate reconstruction may be economically and psychologically better when compared with delayed reconstruction algorithms.\textsuperscript{14}

Given the complexity of such a reconstruction, the disadvantages of the procedure should be addressed. These disadvantages include the requirements of a trained microsurgical team, high patient compliance in a dedicated postoperative rehabilitation regimen, and the potential for serious complications of surgery such as microvascular toe failure and suboptimal function and cosmesis. Considering the multifaceted aspects of the surgery, patient factors such as age, hand dominance, occupation, medical comorbidities and concomitant injuries need to be considered.

When the calcifying aponeurotic fibroma is large enough to encase critical structures and its debulking has the potential to compromise the vascularity and function of the thumb, an amputa-}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{image1.png}
\caption{Radiographic evidence of bony healing after a trimmed toe-to-thumb transfer.}
\end{figure}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{image2.png}
\caption{There is multinodular proliferation of mononuclear and giant cells. Individual mononuclear cells have an ovoid to spindle nucleus with finely dispersed chromatin and small to inapparent nucleus. There are areas of central cartilaginous metaplasia and dystrophic calcification.}
\end{figure}

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