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CASE REPORT
Hand

Calcifying Aponeurotic Fibroma in the Deep Space of the Hand

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Summary: Calcifying aponeurotic fibroma is a rare tumor that can involve the upper extremity, but deep hand space involvement is exceptionally uncommon and presents unique diagnostic and treatment challenges. We present a 9-year-old girl with a progressively enlarging mass on the ulnar side of the left palm. Magnetic resonance imaging revealed an ill-defined tumor with fatty infiltration spanning the third and fourth metacarpal spaces, suspicious of an aggressive tumor. The delay in diagnosis, likely stemming from the location in the deep space of the palm and lack of functional impairment, allowed this lesion to become atypically large and expansive. Given the child's age, a marginal excision was performed with careful preservation of the adjacent tendons, bone, and neurovascular structures. Although long-term monitoring for recurrence is ongoing, the patient had no evidence of recurrence at 1-year follow-up. *(Plast Reconstr Surg Glob Open 2024; 12:e6358; doi: 10.1097/GOX.0000000000006358; Published online 3 December 2024.)*

Calcifying aponeurotic fibroma (CAF) is a rare, benign, and locally aggressive tumor predominantly found in the extremities of children.[1] Since its initial description in 1953 by Keasbey et al, more than 150 cases have been documented with varying presentations. The upper extremity is a common site for these lesions, but deep space involvement in the hand is extremely rare and necessitates magnetic resonance imaging (MRI) to delineate the boundaries of the tumor[2] and confirmatory biopsy.[1] We present the rare case of a 9-year-old girl with a large CAF embedded in the deep mid-palmar space of the hand.

CASE PRESENTATION

A White 9-year-old girl presented to our clinic with subtle swelling of her left palm that was noticed by her father 3 months before her initial consultation. There was no history of trauma, and her medical history was unremarkable. A physical examination revealed a 3 × 3 cm diffuse soft-tissue swelling to the ulnar aspect of her left palm. She had no functional deficits and minimal discomfort. MRI showed an ill-defined 3 × 2.5 × 1.5 cm fat-containing mass in the left hand, extending within the third and fourth intermetacarpal spaces and displacing the ring and little finger flexor tendons (Figs. 1–3). Urgent surgical excision was planned.

Brunner-type zigzag incisions on the palmar-ulnar side of the hand exposed the tumor beneath the subcutaneous fat. The gritty, off-white mass was grossly distinct from the surrounding normal tissues. The tumor was meticulously dissected away from the flexor tendons and neurovascular structures. The common digital vessel and nerve to the ring and little fingers were encased in the lesion and required dissection from unaffected areas both proximally and distally. The deep areas of the tumor were adherent to the fascia overlying the interossei muscles and the periosteum of the fourth and fifth metacarpals, and each of these structures was elevated with the tumor to ensure a clear margin. After the mass was resected, the wound was closed, a compressive dressing was applied, and the arm was casted to protect the incisions given concerns about compliance with postoperative activity limitations in a patient of this age. Histopathology confirmed a benign spindle cell tumor with calcification, consistent with a CAF (Fig. 4). One year postoperatively, the patient reported no pain or nerve deficits, and the wound was well healed. There has been no clinical evidence of recurrence.

DISCUSSION

CAF, previously known as juvenile aponeurotic fibroma, is an uncommon benign soft-tissue tumor

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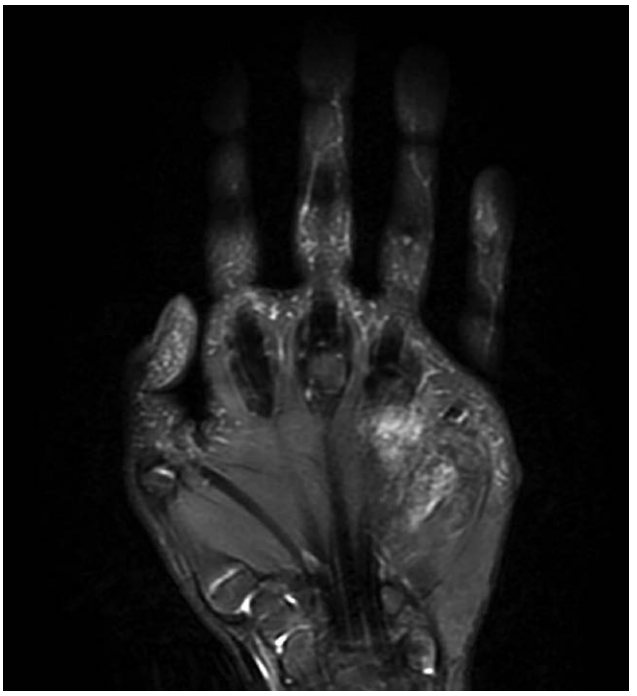

Fig. 1. Contrast T2-weighted MRI coronal view of the left-hand ill-defined heterogeneous soft-tissue mass.

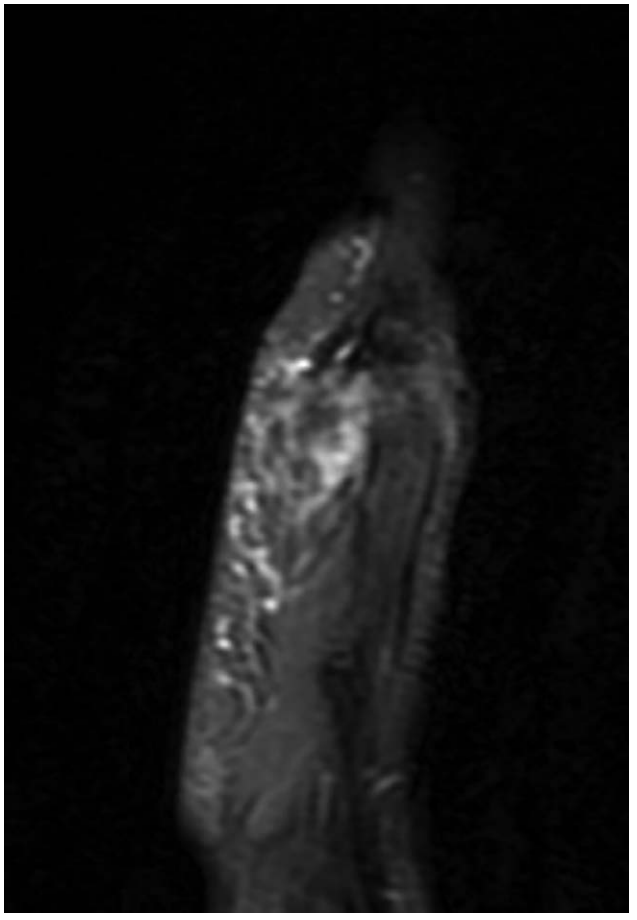

Fig. 2. Contrast T2-weighted MRI sagittal view of the left-hand ill-defined heterogeneous soft-tissue mass.

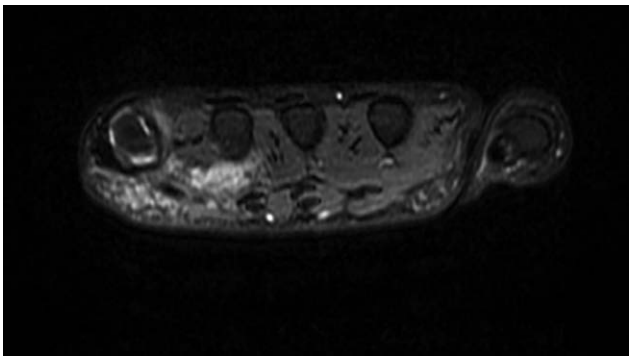

Fig. 3. Contrast T2-weighted MRI axial view of the left-hand ill-defined heterogeneous soft-tissue mass.

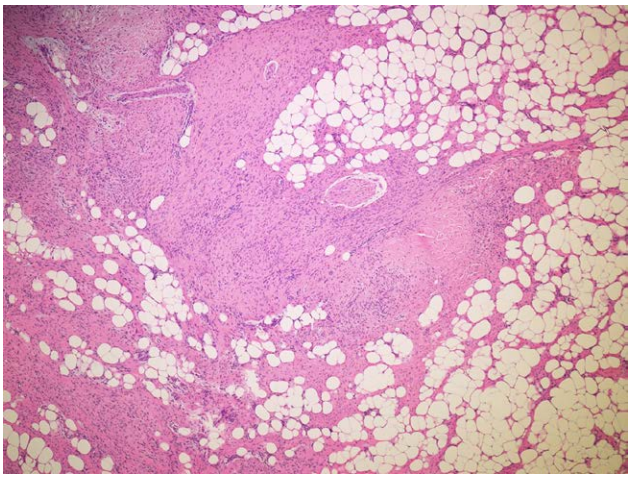

Fig. 4. Calcifying aponeurotic fibroma histopathology.

predominantly affecting children and young adults.[1] CAF often presents as a slow-growing, painless mass infiltrating surrounding tissue with a predilection for the distal extremities. However, it has also been described in the proximal extremities, back, trunk, and neck.[1,2] Pain and tenderness have been reported in some cases.[3] Most reports on pediatric hand CAF are isolated to the fingers, wrist, or palm.[1–9] Involvement of the deep palmar space with extension to the bone and interosseous muscles, is particularly rare. Although a few similar cases have been reported, the lack of detail on the depth or involvement of the tumor makes comparisons difficult. (**See table, Supplemental Digital Content 1,** which displays the summary of palmar hand CAF cases, **http://links.lww.com/PRSGO/D672**.) Nevertheless, the size and expansiveness of the CAF seen in our patient is extremely rare, and we hypothesize that the delay in diagnosis and treatment was likely due to the deep palmar location and the absence of functional impairment. In fact, the slow, subtle swelling of the palm went largely unnoticed by the patient's parents until the patient complained of occasional discomfort while holding objects.

Accurate diagnosis of CAF poses a significant challenge due to its nonspecific presentation and similarity to

potentially malignant soft-tissue tumors.[10] MRI can help assess the tumor's composition and anatomic involvement,[10,11] which was particularly extensive in our case, but definitive diagnosis relies on histopathology after surgical excision.[1] In our case, the biopsy revealed the characteristic features of CAF, including fibroblasts, chondroid areas, and calcifications within a collagenous stroma.[11] Surgical excision is the preferred treatment for CAF,[10] though it carries a high local recurrence rate of up to 50%, particularly in younger patients, and 1–3 years postoperatively.[1] Given this risk, close long-term follow-up is recommended despite the lack of standardized treatment guidelines.[10] CAF has been reported to involve the neurovascular structures and, in very rare cases, invade down to the periosteum and joint space. In our patient, the lesion was intricately involved with all vital hand structures (ie, tendons, nerves, muscle, bone, and vessels), which required a more conservative and painstaking approach to preserve function. Fortunately, the patient's hand remains fully functional, with no sensory loss or recurrence a year postoperatively.

Although the malignant transformation of CAF is exceedingly rare, it must be considered. There have been reports of CAF metastasizing years after excision, emphasizing the importance of regular follow-up. In 1 study, a 3-year-old girl's CAF in the palm metastasized as fibrosarcoma to the lungs and bones nearly 5 years after local excision.[3] Benichou et al[12] described a 7-year-old boy who experienced local recurrence of CAF for 7 years, leading to hand amputation and subsequent death due to the extension of fibroblastic processes to the lungs and pleura. Although our patient shows no evidence of recurrence, our oncology team will continue to follow up with her.

CONCLUSIONS

Our patient underscores the diagnostic and management challenges of pediatric soft-tissue tumors in the deep palmar spaces. Although benign, the high recurrence rate warrants considering CAF in the differential diagnosis of rapidly growing pediatric soft-tissue lesions. Successful management of hand CAF hinges on balancing adequate excision to reduce recurrence while preserving function. The necessity of long-term follow-up in young patients cannot be overstated.

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DISCLOSURE

The authors have no financial interest to declare in relation to the content of this article.

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