CASE REPORT
Giant Calcifying Aponeurotic Fibroma of Web Space: Case Report with Review of Literature
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Abstract:
Calcifying aponeurotic fibroma is an uncommon, benign fibroblastic tumor which has a preference for neighborhood invasion and a high relapse rate. Henceforth, precise preoperative analysis and complete extraction are imperative to counteract the recurrence of the tumor after careful surgical removal. We report a case of 75 year old man with left hand first web space calcified aponeurotic fibroma with distinct radiological and MRI features.

Keywords: Calcifying Aponeurotic Fibroma, Fibroblast, Dense Calcification, Palisading, Web Space

Introduction:
Keasbey in 1953 first described Calcifying Aponeurotic Fibroma (CAF) and alluded it as Juvenile Aponeurotic Fibroma [1]. CAF characteristically appears in Palmer region of hands and plantar aspect of feet and is an infrequent, benign, regionally destructive fibroblastic soft tissue tumor, usually affecting young and adolescent age group. [1]. Pathologically, the condition is characterized by fibroblast proliferation with calcification [2]. The tumors show invasive growth; therefore, they are often difficult to distinguish from malignant tumors. The tumour has a tendency for local relapse after surgical resection [3-4]. We report a case of long duration swelling in the first web space of the left hand with characteristic radiological and Magnetic Resonating Imaging (MRI) findings.

Case Report:
We are presenting a case of 75 yrs. an elderly patient with a slow-growing painless mass in the first web space of left hand since last 30years. The physical examination uncovered a firm to hard non-tender, discernable mass occupying first web space calcified aponeurotic fibroma with distinct radiological and MRI features. Radiographs demonstrated a relatively well-defined soft tissue mass with areas of dense calcifications (Fig.2). MRI (Fig.4a-b) revealed a well-defined heterogeneous intensely enhancing mass lesion measuring 71 × 54 mm on the palmar aspect of left hand between 1st and 2nd digit causing abutting and splaying of adductor pollicis brevis and adductor pollicis muscles and abutting flexor tendon at its terminal part. The lesion is appearing iso to hypointense on T1WI, heterogeneous on T2WI. The mass was excised and was revealed to be a pale yellow to gray colored, firm lesion (Fig.3). On histopathological assessment, tumor mass displayed cellular, plump fibroblastic cells palisading around a hyalinized collagenous zone, which sequentially surrounds the calcification in distinctive zonation pattern without any mitotic figure or cellular atypia (Fig.4c), confirming the diagnosis of calcifying aponeurotic fibroma.
Fig. 1: Preoperative Image of Calcifying Aponeurotic Fibroma involving Left Hand

Fig. 2: Lateral and AP X-ray of Left Hand Showing Well Defined Lesion with Areas of Dense Calcification

Fig. 3: Cut Section of Calcifying Aponeurotic Fibroma Left Hand

Fig. 4: Coronal MR Images (a-b) Demonstrate Well-Defined Subcutaneous Soft Tissue Mass, Showing Hypointense Signal Intensity on T1WI (a), Heterogeneous Intensely Enhancing on T2WI (b). Microscopy shows Fibroblastic Cells Arranged in A Palisading Fashion around Foci of Calcification (c).
Discussion:
A calcifying aponeurotic fibroma is non-metastasizing locally aggressive tumor ordinarily happens in the first two decades of life, with a predilection for male sex. CAF ordinarily happens in the distal limits of the body, most usually in the fingers, palms, and soles [5].

The tumor has been typically described as a small (usually <3cm in diameter) slow growing firm, non-tender mass with an inclination to invade the contiguous tissue, good prognosis but high recurrence rate of roughly 50% following surgical resection [1]. CAF albeit locally relapsing, for the most part, does not metastasize. Lafferty et al. [6] reported a case of CAF which has metastasized to bone and lung and metastatic lesions were histopathologically confirmed as fibrosarcoma, five years after the removal of CAF.

Histologically, the lesion revealed characteristic palisading of rounded, chondrocyte-like cells around dispersed chondroid or calcific nodular foci. CAF typically has biphasic growth pattern as reported by Enzinger and Weiss [7] in early phase tumor shows infiltrative and destructive growth and usually lacks calcification and most often seen in young patients but tumour becomes increasingly nodular and smaller with notifiable calcification and cartilage formation in the late phase. The variable presentation of CAF on imaging largely depends upon patient age, bony association and existence of calcifications. Based on clinical presentation and radiological features our case corresponds to the late phase of CAF. Emphasizing features of calcifying aponeurotic fibroma on radiographs include a not well characterized or inadequately delineated edge of the tumor and an inconsistent degree of fine, stippled calcifications inside the lesion. Outward disintegration of the nearby bone is in all respects once in a while seen [3]. For our situation lesion was well demarcated with dense calcification. Most common benign lesions that ought to be separated from CAF are giant cell tumors of the tendon sheath (GCTTS), and fibromatosis [8]. GCTTS classically presents as a well-defined mass with or partially/completely enveloping a tendon and displays a low signal on T1WIs and T2WIs due to the presence of hemosiderin [9]. Fibromatosis at times very difficult to differentiate from CAF because of its resembling imaging and clinical features. (Hypointense signal on T2W1 imaging sequence with distinct border and invasion to the adjoining tissue)[10].

In conclusion, this case is worth reporting because of its unusually large size, presence of calcification and characteristic findings on MRI.

References

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