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Research Paper

Chondromyxoid Fibroma of Scaphoid: a Rare Case

Medical Science

KEYWORDS : Chondromyxoid fibroma , aneurysmal bone cyst, Scaphoid, wrist arthodesis

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ABSTRACT

Chondromyxoid fibroma is a rare benign cartilage-forming tumor, counting for 0.5% of all primary bone tumors . The development of Chondromyxoid fibroma in the carpal bones is extremely rare. This article describes a rare case report of Chondromyxoid Fibroma in Scaphoid bone of a 15 year male. To our knowledge, this is the first report of Chondromyxoid fibroma in Scaphoid. The rarity of the lesion, together with its misleading clinical -radiological and histopathological features, prompted us to report our case.

Indroduction:

Chondromyxoid fibroma, a rare chondroid tumor, consisting of a mixture of fibromyxoid tissue and cartilage tissue in variable proportions and accounting for 0.5% of all primary bone tumors and 2% of benign bone tumors, occurs predominantly in adolescents and young adults and more commonly in males than in females (2:1) [13]. This tumor has a predilection for the bones of the lower extremities, usually the proximal tibia. Small bones of hand and feet including vertebrae are rare sites [15,16,17,18]. Only a few isolated cases of chondromyxoid fibroma affecting carpal and metacarpal bones have been reported [1, 3, 11, 12].

CASE

A 15-year-old boy presented with a 7-month history of intermittent vague pain and progressive increasing swelling in the region of the wrist of his left hand with inward deviation of hand and weakness of grip strength for last 4 month. No history of trauma, or complaints suggestive of infection.

Physical examination showed a eccentric swelling of about 7x 5 cm size (fig 1) localised on radial aspect of left wrist with overlying skin normal, fixed tender and variegated consistency, overlying tendons free .Movement at wrist was restricted and painful. No distal neurovascular deficit. No involvement of other bones. Laboratory tests were unremarkable.

A radiograph of his left hand showed a expansile bony growth arising from distal part of scaphoid with multiple lobular, septed, intact cortex, marginal sclerosis, cortical thinning and without obvious matrix mineralization and no new bone formation. There was associated osteopenia of other bone(fig 2). It was concluded that the tumor was likely to be benign and the differential diagnosis included enchondroma, aneurismal bone cyst, giant cell tumor and osteoblastoma. Fine needle aspiration cytology showed features of acute inflammatory lesion. MRI revealed multiple cystic lesion associated with fluid –filled level involving Scaphoid causing its expansion and no obvious area of cortical breach, confirming to Aneurysmal bone cyst(fig 3a,3b) .We planned for Scaphoidectomy and limited wrist arthodesis

At surgery , a radiodorsal incision given, bulging bony mass from distal part of Scaphoid with surrounding fibrosis encroaching lunate making hard for smooth removal of Scaphoid(fig 4 a). As a result Lunate was also sacrificed and scapholunatectomy was performed. All precaution was taken to avoid damage to volar carpal ligaments. Gross examination of Scaphoid showed replacement of entire medullary cavity with spongy, gelatinous material (fig 4 b, c). There was breech of cortex at capitate articular surface without involvement of capitate. Lunate was firmly adhered to Scaphoid .There was no change in radial and ulnar articular surface. Finally total wrist arthodesis with 3.5 mm recorn plate and auto iliac cancellous bone grafting was done (fig 5).

Histopathology reported band-like peripheral cellular condensation surrounding and dividing the pale-staining matrix-rich central parts of tumor in low power (fig 6a). Large lobulated areas of spindle-shaped or stellate cells distributed within abundant myxoid or chondroid intercellular material in high power (fig 6b). A characteristic finding was the increased cellularity of the tissue near the septa. These findings concluded the diagnosis of Chondromyxoid Fibroma.

Fig 1. Eccentric Swelling of about 7x 5 cm size localised to radial aspect of left wrist with overlying skin normal

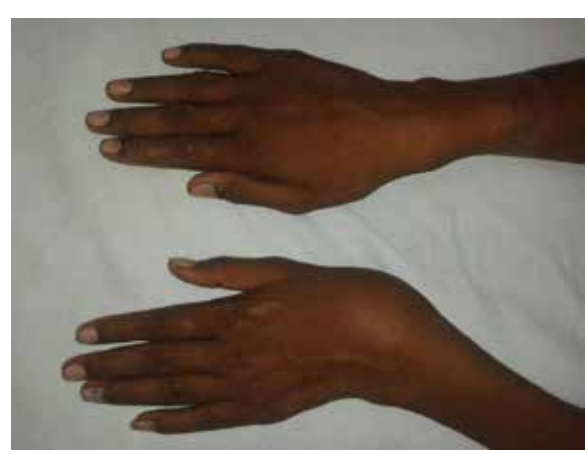

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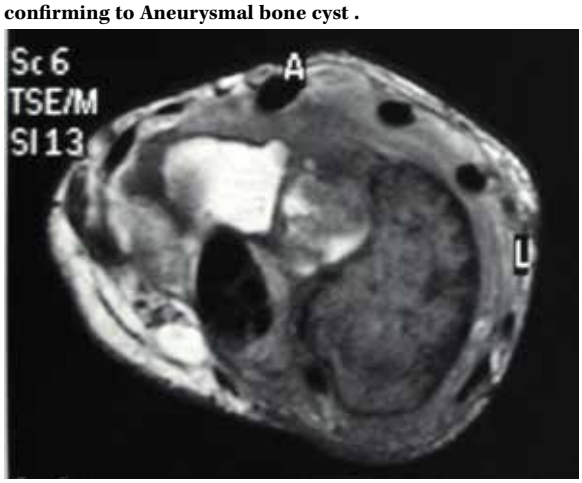
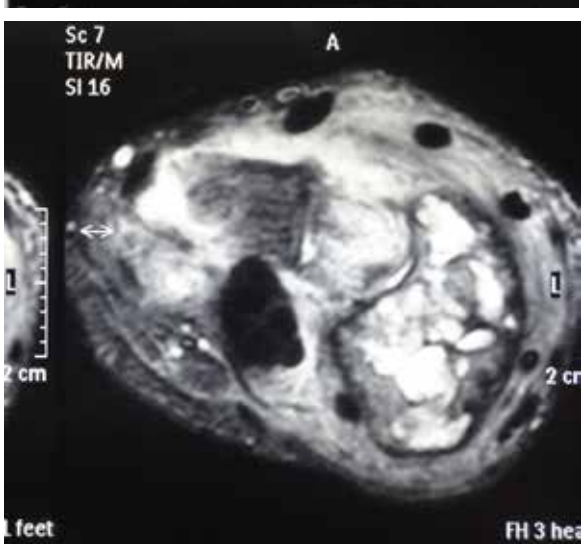
Fig 2: A expansile bony growth arising from distal part of scaphoid with multiple lobular, septed, intact cortex, marginal sclerosis, cortical thinning and without obvious matrix mineralization and no new bone formation. There was associated osteopenia of other bone.

Fig 3a,3b: transverse section showing multiple cystic lesion associated with blood –filled level involving Scaphoid causing its expansion and no obvious area of cortical breach, confirming to Aneurysmal bone cyst .

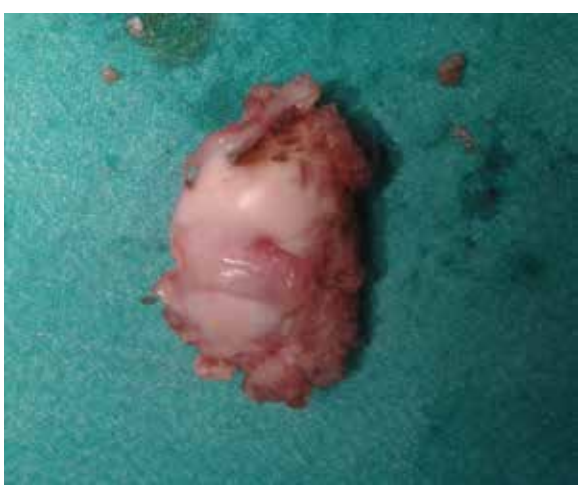




- cyst seen with the lesion have no obvious associated solid component & are showing heterogeneously hyper-intense signal on T2W and STIR images. Cysts are showing varying signals intensities suggestive of blood degradation products with fluid –fluid level. The entire area is exhibiting typical honey comb appearance.

Fig 4a: exposed a bulging bony mass from distal part of Scaphoid with surrounding fibrosis , making hard for smooth removal of Scaphoid.

Fig4b,c: Gross examination of Scaphoid showed replacement of entire medullary cavity with spongy, gelatinous material. There was breech of cortex at capitate articular surface without involvement of capitate. Lunate was firmly adhere to Scaphoid .



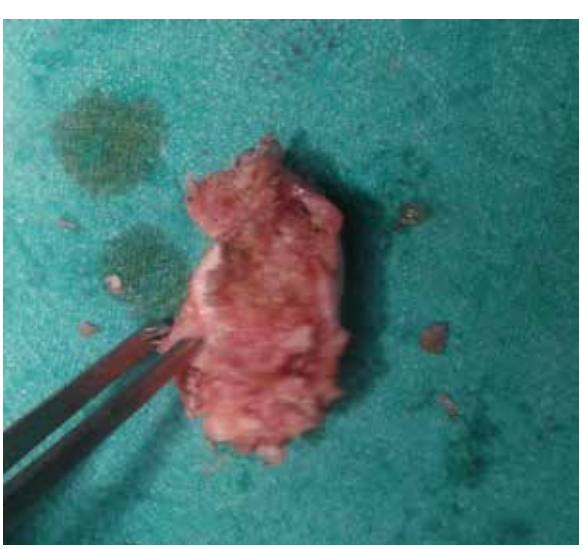


Fig5a & 5b: AP and Lateral post operative radiographs of total wrist arthodesis

Fig 6a: HPE (low power) Band-like peripheral cellular condensation surrounding and dividing the pale-staining matrix-rich central parts of tumor

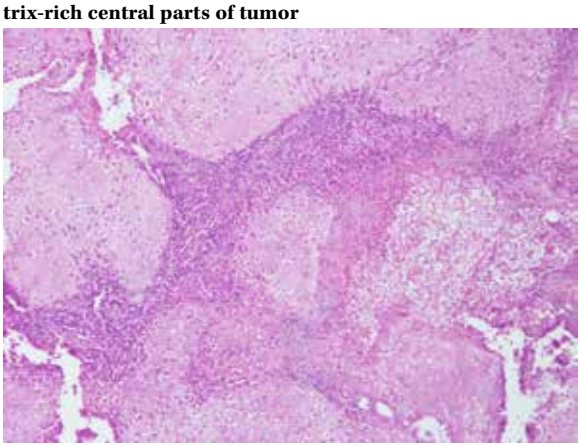
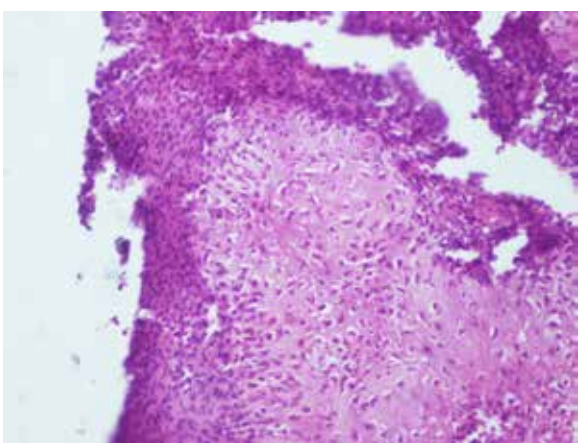


Fig 6b: HPE (high power) Large lobulated areas of spindle-shaped or stellate cells distributed within abundant myxoid or chondroid intercellular material. A characteristic finding of increased cellularity of the tissue near the septa. Findings suggestive of Chondromyxoid Fibroma.



Discussion

This is the first report of Chondromyxoid fibroma in Scaphoid. The most common clinical presentation is of pain and swelling, and the characteristic radiographic features are those of a radiolucent lesion, often in an eccentric metaphyseal location. The cortex is thinned and shows internal scalloping. A thin sclerotic rim separates the tumor from the underlying normal trabecular bone [4, 6, 7, 9, 10, 12]. In small tubular bones such as the metacarpal or metatarsal, the lesion usually has fusiform expanded structure with thinning of all cortices [2, 3, 4, 5, 11]. Treatment options include en-bloc resection, curettage, or curettage with bone grafting. We performed scapholunatecomy and total wrist arthodesis with recorn plate. As tumor had breached the Scaphoid cortex with local spread, excision of Scaphoid and lunate had to be done. The risk of local recurrence is reported to range from 4% to as high as 80% depending on the type of surgical treatment [2, 5, 7, 9]. The incidence of recurrence is also higher in younger patients who have undergone simple curettage of the lesion [1, 5, 8], but drops considerably when bone graft is introduced to fill the defect. Most authors advise that the lesion be excised en-bloc whenever possible, or treated with thorough curettage and bone grafting when the site of the lesion makes more aggressive surgery unreasonable [2, 5, 8, 9]. Although there have been sporadic reports of malignant degeneration of chondromyxoid fibrom most of these are believed to have been in circumstances where a chondrosarcoma was initially misdiagnosed as a chondromyxoid fibroma[1, 2, 8, 14].

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