MI-COD

MSS INDIA- Case Of the Day

17/10/2024

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Musculoskeletal Society of India *(MSS)* &

Indian Journal of Musculoskeletal Radiology

(IJMSR), the official publication of MSS India

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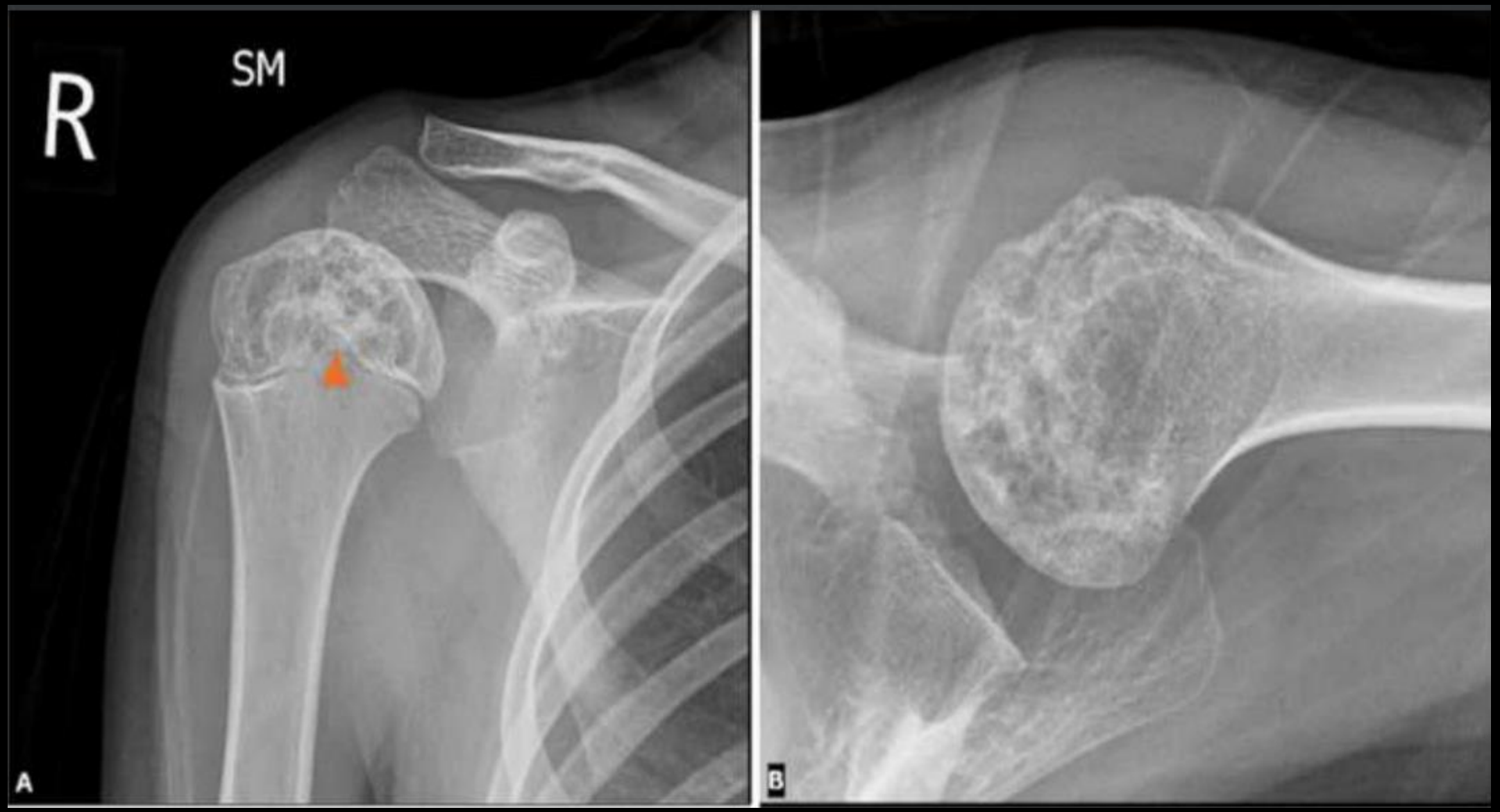
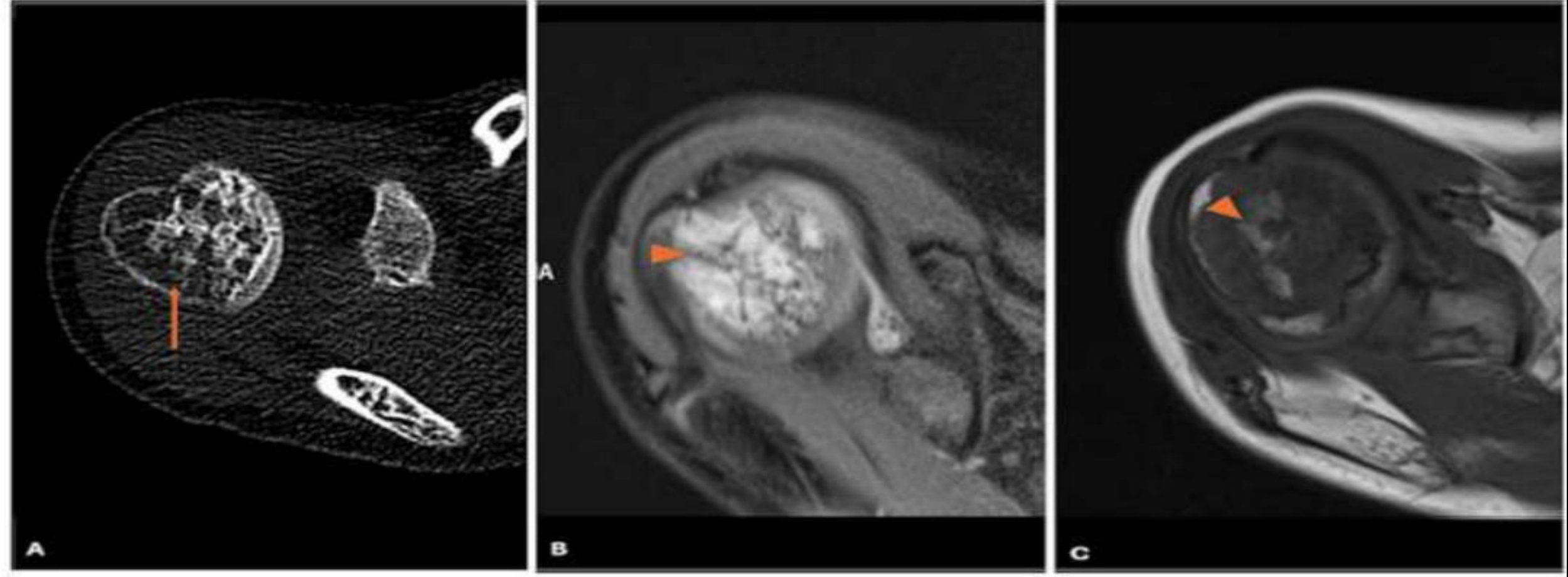
History :

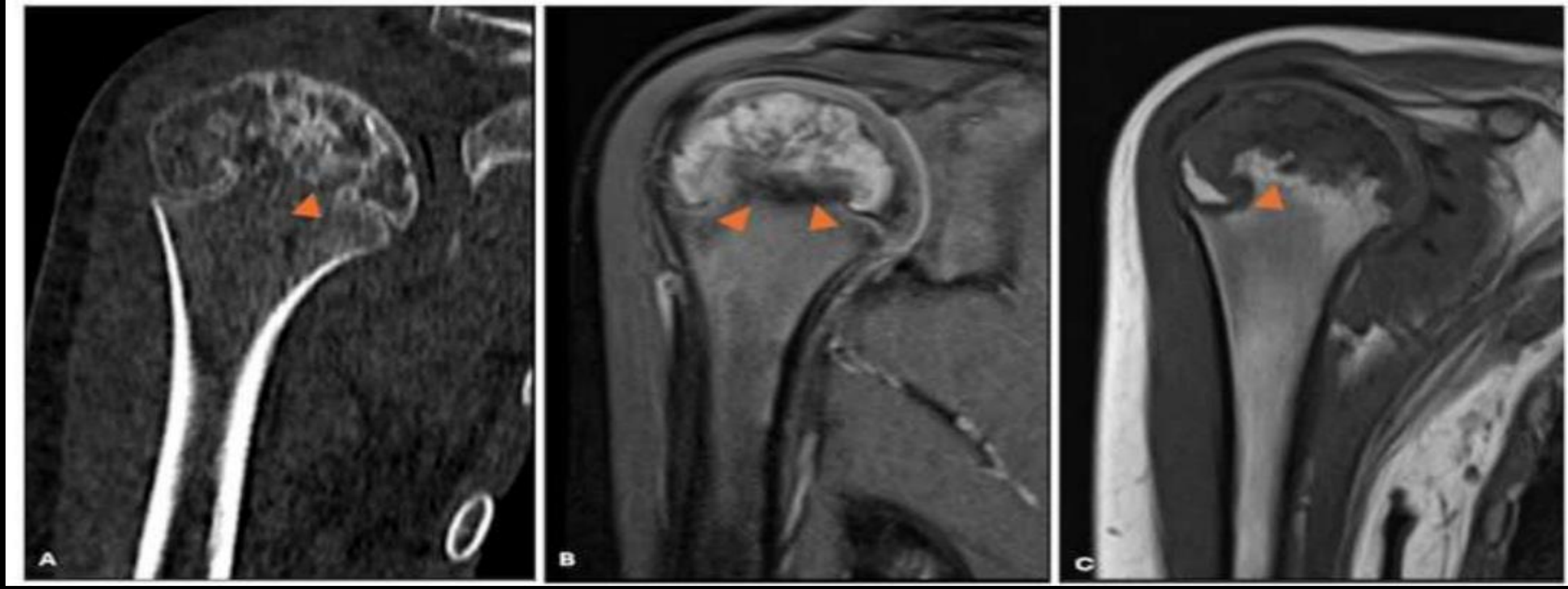
A 11-year-old male presented with a 2-month history of right sided shoulder pain. There was no history of trauma, and the patient is systemically well.



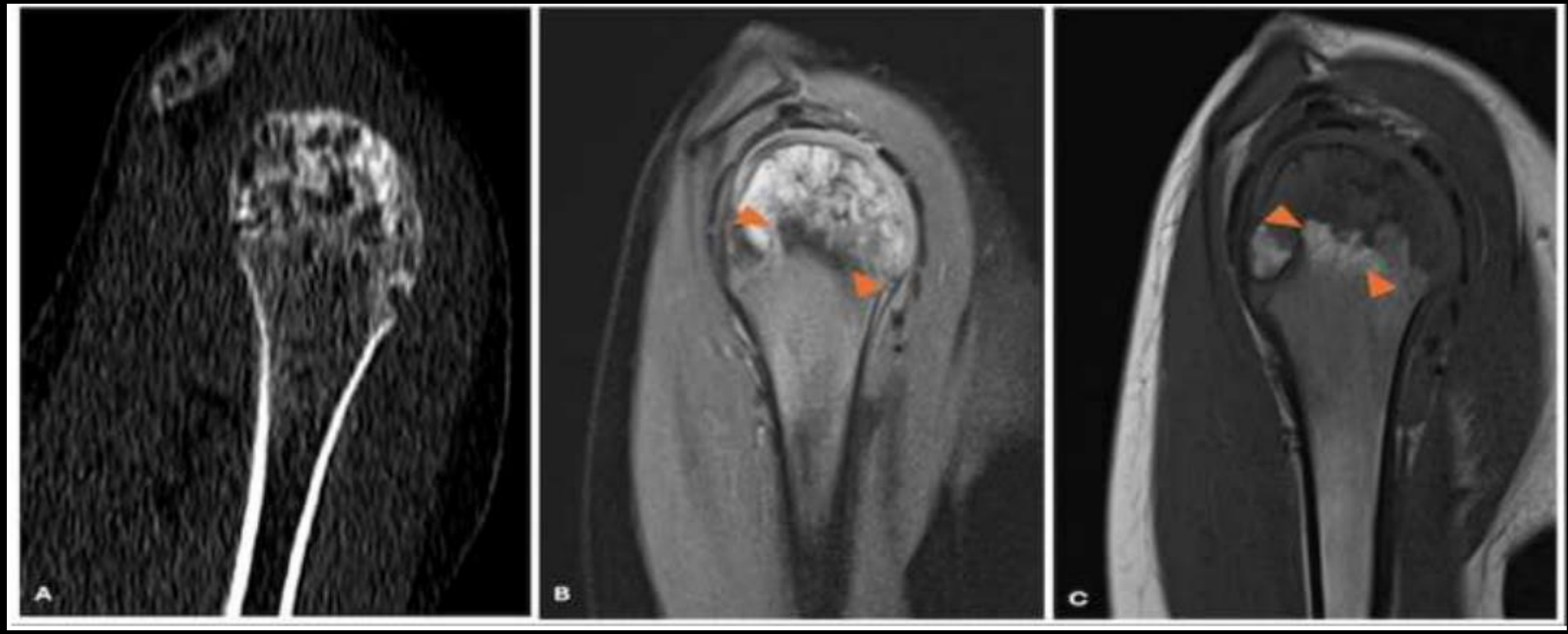
AP (1A) and Axial (1B) Shoulder Radiographs show a mixed sclerotic and lucent lesion in the right proximal humeral epiphysis with ring-and-arc like calcification. Central closure of the physis noted (arrowhead). No cortical destruction or periosteal reaction.



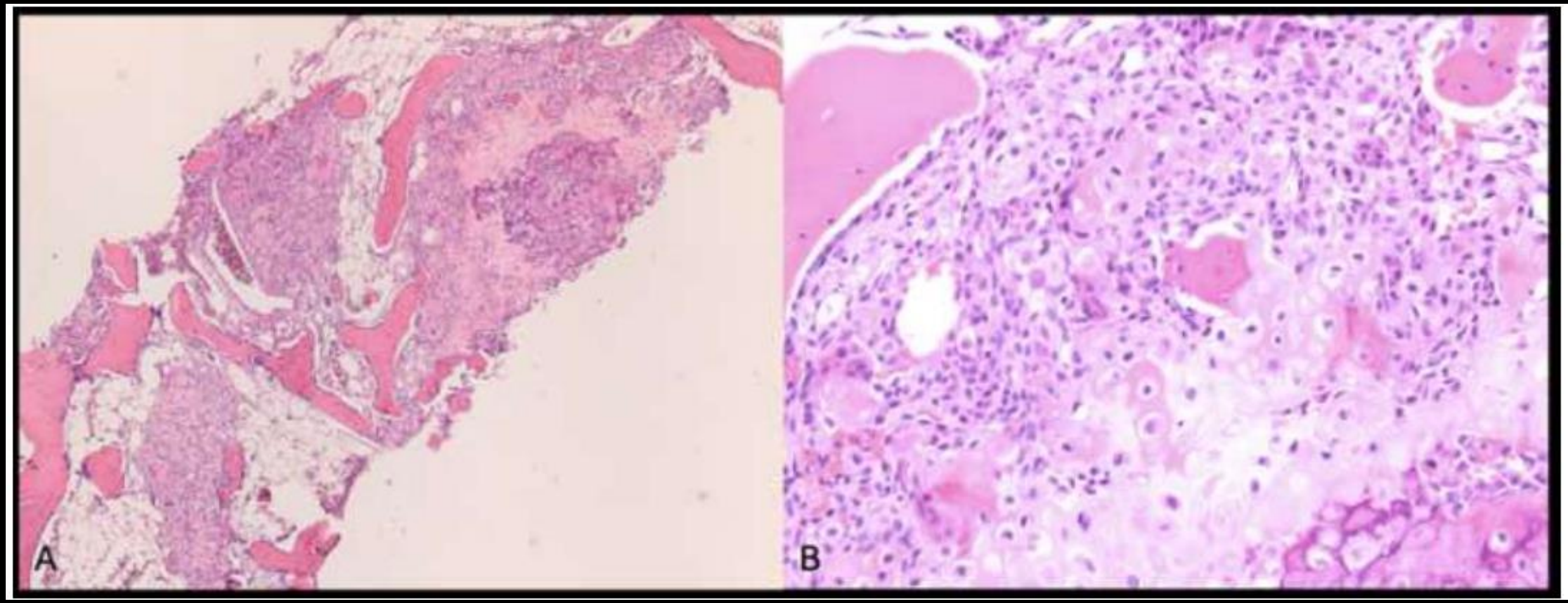
Axillary CT (A), Proton Density (B) and T1-W sequences (C) confirm the ring-and-arc like calcification, with central low T1, high T2 signal of hyaline cartilage/cystic areas (asterisk) and low T1/T2 septae of enchondral calcification (arrowheads). Thinning of the cortical bone and multiple areas of endosteal scalloping (arrow) but no cortical erosion, bony expansion or soft tissue extension.



Coronal CT (A), Proton Density (B) and T1-W sequences (B) – chondral pattern calcification and MRI characteristics as per previous figure. The premature closure of the central epiphysis is more clearly demonstrated suggesting the lesion has been present for some time, while this is not a definite feature of clear cell chondrosarcoma, it does correlate with the low-grade, slowly progressive nature of the lesion.



Sagittal CT (A), Proton Density (B) and T1-W sequences (C) – again demonstrating the ring-and-arc chondral pattern. Note the minimal adjacent oedema in the metaphysis adjacent to the physeal closure (arrowheads) and no periosteal reaction – favouring clear cell chondrosarcoma over chondroblastoma.


A
B

DIAGNOSIS

- **Clear cell chondrosarcoma**

Clear cell chondrosarcoma is a low-grade malignant lesion of cartilage origin, typically having an indolent slow growing course, and with pain being the most common presenting complaint.

In many cases there is delayed diagnosis because of the reassuringly benign imaging findings on initial radiographic films and MRI. Nevertheless, there is potential for bony and lung metastasis with an overall 5-year mortality of 15%.

Typically, the age of onset is from the 3 rd to 5th decades of life.
Lesions are centred on the epiphyses with the femoral and humeral heads accounting for around two thirds of all cases.

Radiographic and CT features show a well-defined, expansile lytic lesion with a variable zone of transition and matrix calcification.

Often there is a narrow zone of transition with sclerotic rim simulating a benign lesion.

The classical ‘ring-and-arc- calcification is only seen if hyaline cartilage/enchondral calcification is present and is better appreciated on CT than on plain radiograph or MRI.

MRI features are generally of a low T1, high T2 lesion – T2 heterogeneity is common owing to variable matrix calcification or haemorrhage.

There is minimal surrounding oedema, no periosteal reaction and no soft tissue extension (unless pathological fracture is present).

Differentials include chondroblastoma, osteoblastoma and if matrix mineralisation is minimal/absent then even giant cell tumours or aneurysmal bone cysts.

Of particular importance is the differentiation between clear cell chondrosarcoma and chondroblastoma – the most reliable features to help distinguish these two are the greater degree of oedema and periosteal reaction with chondroblastoma compared to clear cell chondrosarcoma.
Additionally, chondroblastoma is generally smaller and with less matrix calcification than clear cell chondrosarcoma, although these are somewhat less reliable features.

Early intervention and wide-margin excision the preferred treatment.
Long term follow-up more than 10-years is likely required given high rates of local and metastatic recurrence.