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Understanding Non-Ossifying Fibroma or Chondroid Myxoid Fibroma: Causes, Diagnosis, And Challenges

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Abstract: ***Non-ossifying fibromas (NOFs) are common lesions most frequently found in the growing bones of children and adolescents. Although NOFs are benign, their presence as incidental findings often triggers further investigation as they are sometimes mistaken for more sinister bone lesions such as aneurysmal bone cysts. NOFs can also pose an increased risk of pathological fractures. However, there are currently no guidelines on the follow-up of NOFs. We present a case series of five patients from Addenbrooke's Hospital with NOFs illustrating their morphological features on plain radiographs and highlighting specific characteristics to support clinicians in diagnosing and managing NOFs. Chondromyxoid fibroma (CMF) is a rare benign bony tumour. Our objectives are three-fold: first, comparing MRI, conventional radiography (CR) and CT characteristics of CMF; second, providing a literature review; and third, summarizing the role of imaging landmarks in the differential diagnosis with other bony lesions.***

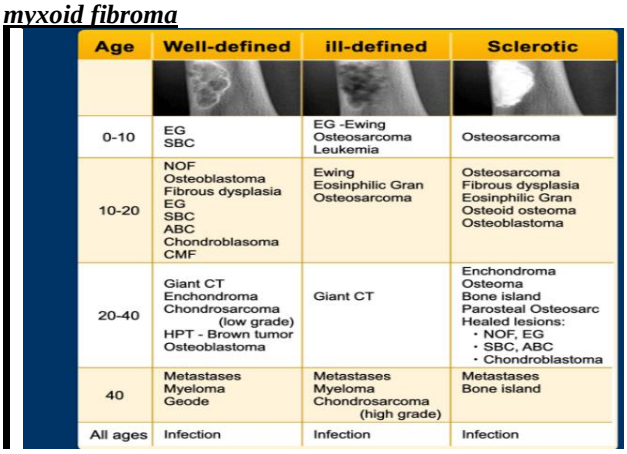
Keywords: non-ossifying fibroma, bone lesion, paediatric orthopedics

1. Introduction

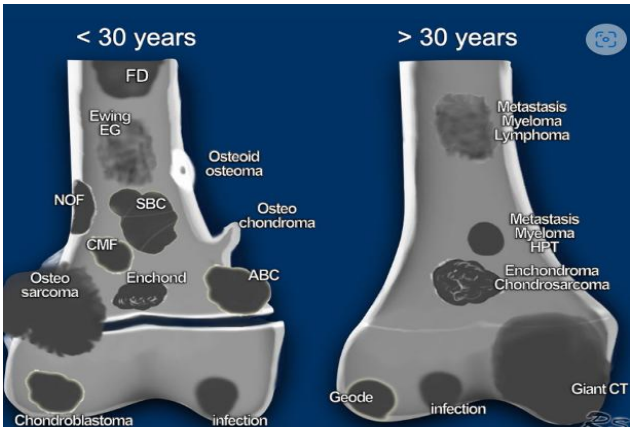
Non-ossifying fibromas (NOFs) are common bone lesions, estimated to be present in 20-30% of all four to ten-year-olds. They are characterized by benign growths of osseous and fibrous tissue and most frequently affect the metaphysis of the distal femur and the proximal tibia. Although NOFs are benign, their presence on X-ray imaging often leads to further, usually unnecessary investigations such as MRI, CT, and biopsies. It is thought that certain NOFs increase the risk of pathological fractures in the growing bones of children. However, little progress has been made in understanding which of these lesions are at higher risk. The Ritschl stage has been proposed to classify NOFs based on their natural radiological progression. These stages are based on the appearance of NOFs on plain radiographs. Stage A lesions are lucent with clear margins. Stage B lesions are lucent with a thin sclerotic border while stage C lesions show increasing sclerosis and stage D lesions are completely sclerosed. It has been reported that Ritschl stage B lesions are at higher risk of fractures and thus follow-up has been recommended until stage C is achieved.

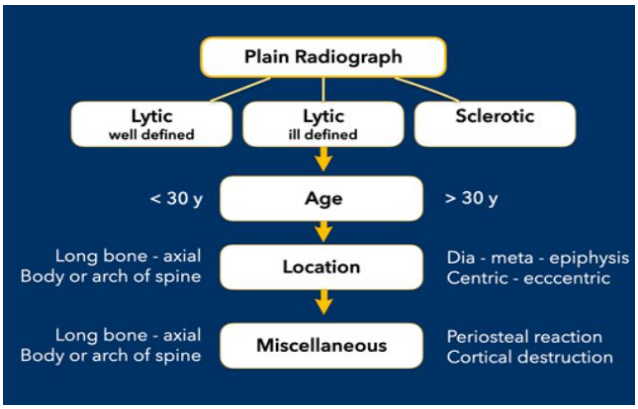
Chondromyxoid fibroma (CMF) is a rare entity, representing <1% of all primary bone neoplasms. It belongs to the group of benign cartilage tumours and has first been described in 1948 by Jaffe and Lichtenstein. It is frequently diagnosed in the second decade of life and is slightly more frequent in males, with a male: female ratio of 1.28: 1. The final diagnosis is not always easy because of its rarity but also due to the overlap of characteristics with other bone tumours. Additionally, CMF can occur in any part of the skeleton, although it appears most commonly in long bones. The radiographic features include a well-defined focal bone lesion with geographic bone destruction, a sclerotic rim, lobulated margins and internal trabeculations. Frequently, cortical ballooning and expansion is visible and even complete cortical destruction may be seen in almost one-third of the cases.4 The differential diagnosis consists of aneurysmal bone cysts, giant-cell tumours (GCTs), chondrosarcoma, chondroblastoma, enchondroma and non-ossifying fibroma.

ANATOMY: Non ossifying fibroma AND Chondroid myxoid fibroma



Age	Well-defined	ill-defined	Sclerotic
0-10	EG SBC	EG -Ewing Osteosarcoma Leukemia	Osteosarcoma
10-20	NOF Osteoblastoma Fibrous dysplasia EG SBC ABC Chondroblasoma CMF	Ewing Eosinphilic Gran Osteosarcoma	Osteosarcoma Fibrous dysplasia Eosinphilic Gran Osteoid osteoma Osteoblastoma
20-40	Giant CT Enchondroma Chondrosarcoma (low grade) HPT - Brown tumor Osteoblastoma	Giant CT	Enchondroma Osteoma Bone island Parosteal Osteosarc Healed lesions: · NOF, EG · SBC, ABC · Chondroblastoma
40	Metastases Myeloma Geode	Metastases Myeloma Chondrosarcoma (high grade)	Metastases Bone island
All ages	Infection	Infection	Infection





2. Case Report

A 11 year old female presented to Orthopedic OPD with complains of pain in right leg, swelling on upper 1/3 rd of tibia since 2 years

On examination: CNS study was unremarkable.
CVS study was unremarkable
RS study was unremarkable
PA: soft and non tender

MRI RIGHT LEG with CT CUT was performed.
MRI RIGHT LEG

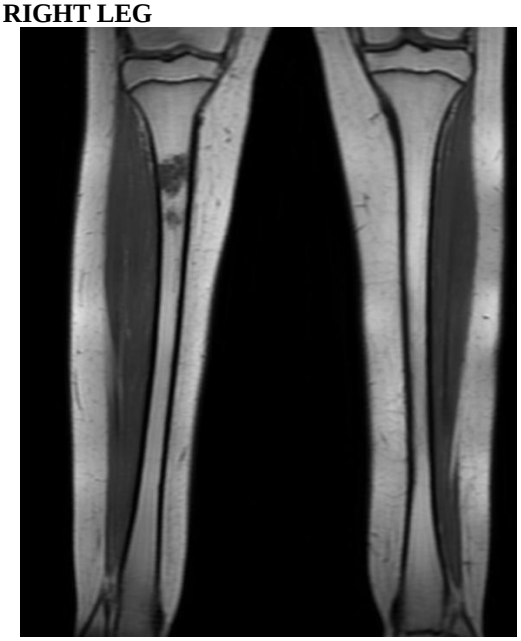


Figure 1: COR-T1

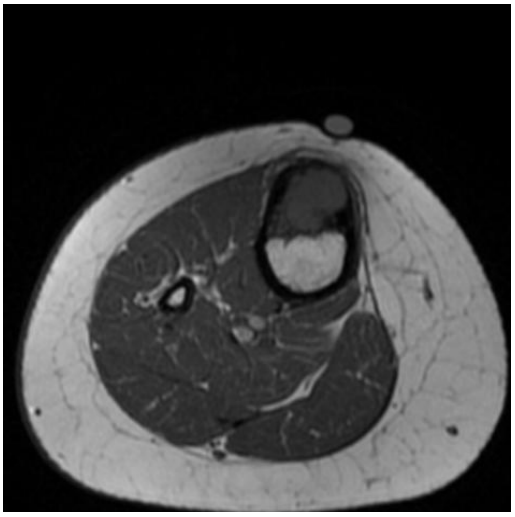


Figure 2: AXIAL T1W

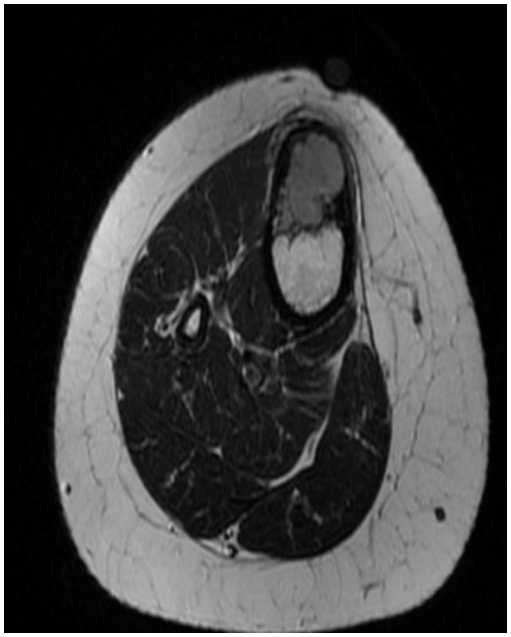


Figure 3: AXIAL T2W

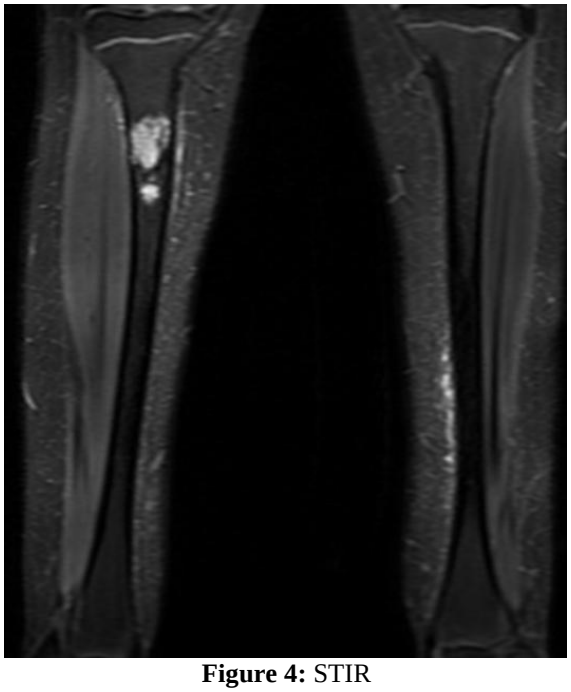
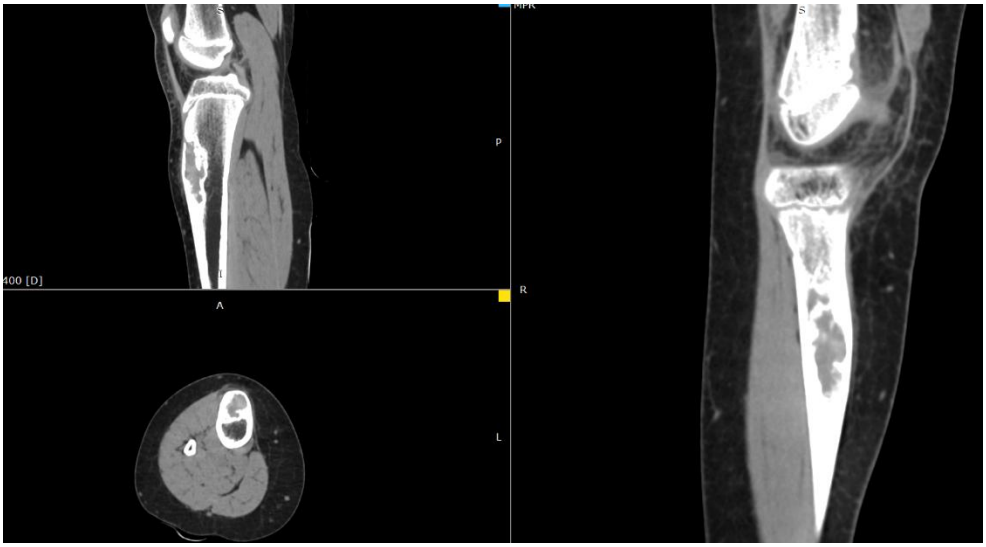


Figure 4: STIR



Limited CT Cut Images

3. Case Report

A 5.7 x 1.9 cm sized well defined intra-cortical area of altered signal intensity is noted involving the metaphysis of proximal tibia anteriorly with narrow zone of transition. It appears hypointense on T1W, intermediate sequence on T2W and hyperintense on STIR images. Few tiny areas of cortical breech are also noted involving it's anterior and lateral aspect. On Diffusion weighted images study, the lesion appears hyperintense with no drop on ADC images. T1W and T2W hypointense perilesional sclerotic rim is noted (confirmed on CT cuts).

On limited available CT cuts, there is defect measuring 2.3 mm in the outer cortical margin of the which is suspicious for pathological fracture. An eccentric expansile lytic lesion with few specks of calcification within and adjacent sclerosis is noted involving the proximal metaphysis of tibia. There is resultant cortical thinning in the anterior aspect of the lesion.

Intra-cortical area of altered signal intensity involving the metaphysis of proximal tibia with narrow zone of transition, extension and morphology as described above. In view of location and age demographics, Possible radiological differentials are:

- Non ossifying fibroma.
- Chondroid myxoid fibroma.

4. Conclusion

Although NOFs are very common in the pediatric population, there is a lack of awareness for identifying these lesions as well as guidelines regarding their management. Our case series has shown an overuse of MRI involved in NOF diagnosis. There needs to be greater confidence in identifying NOFs by using X-ray imaging in combination with the patient's clinical presentation. In this way, increasing awareness of NOFs as benign lesions and highlighting their characteristic findings on plain radiographs can reduce the burden on more expensive imaging modalities and result in fewer unnecessary investigations for the patient. Unlike X-ray imaging, MRIs and CT scans require the patient to lay immobile in a closed space for a fixed period of time, which can be difficult for some patients. Furthermore, CT scans result in an unnecessary greater radiation exposure than X-ray imaging and MRIs produce harsh sounds which may not be tolerated well by some patients, especially children. It is important to be aware that NOFs do not require invasive interventions such as biopsy. By correctly recognizing NOFs on radiographs, physicians can ease the anxiety of patients by reassuring them of the benign nature of these lesions and by avoiding less tolerable investigations.

The diagnosis of CMF is difficult because of overlap of characteristics with other bony lesions. Our comparative study puts forward advantages and limitations of different imaging modalities in the diagnosis of CMF.

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