International Journal of Research in Orthopaedics
Mahajan NP et al. Int J Res Orthop. 2022 May;8(3):411-414
http://www.ijoro.org

DOI: https://dx.doi.org/10.18203/issn.2455-4510.IntJResOrthop20221136

Case Report

Chronic non-bacterial osteomyelitis of distal ulna in a paediatric patient: a case report and review of literature

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Received: 01 March 2022
Revised: 04 April 2022
Accepted: 06 April 2022

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ABSTRACT

Chronic non-bacterial osteomyelitis (CNO) is a rare disorder characterized by focal aseptic inflammation with a self-limiting, relapsing course of disease with female predominance and usually observed in paediatric age groups diagnosed by clinical, radiological and histopathological findings after ruling out all other differentials. We reported this rare case found in a 7 year old female with a history of greenstick fracture of distal ulna 3 years ago with flaring up of pain and swelling since trivial trauma to left wrist 1 month ago. We emphasize on the relevant data and findings to reach the conclusive diagnosis and treatment of the disease. CNO can present as benign unifocal non-relapsing to more severe form of multifocal relapsing inflammatory lesions involving metaphysis of long bones, vertebrae, clavicle and mandible. Diagnosis is made after excluding infection, malignancy, auto-immune and metabolic disorders. Treatment is mainly empirical to reduce pain and inflammation through NSAIDs, corticosteroids, bisphosphonates and sometimes TNF alpha inhibitors as no proper guidelines are available.

Keywords: Chronic non-bacterial osteomyelitis, Chronic recurrent multifocal osteomyelitis, Metaphysis of long bones, Paediatric age group

INTRODUCTION

Chronic non bacterial osteomyelitis (CNO) is a rare non infectious inflammatory condition which usually affects metaphysis of long bones, vertebrae, clavicle and sometimes maxillofacial bones commonly observed in the first decade of life with female preponderance.[1]

CNO comprises a spectrum of inflammatory conditions that ranges from being unifocal non-relapsing to multifocal and relapsing. Most severe form is termed Chronic relapsing multifocal osteomyelitis (CRMO). CNO was first described in 1972 by Gideon which was followed by Bjorksten's report in 1978 who termed CRMO for multifocal form of CNO.[2,3]

Most common symptoms patients present include dull pain and swelling on the site of bone involvement. Local skin changes include tenderness and redness.[4] Commonly involved locations include long bone metaphysis and vertebral bodies.[5]

It is a diagnosis of exclusion based upon clinical, radiological, histopathological and laboratoristic findings with differential diagnosis consisting of bacterial osteomyelitis, malignancy, benign bone lesions, Langerhans cell histiocytosis.

Here we describe this rare case in a 7 year old female presenting with post traumatic pain and swelling of distal forearm.

CASE REPORT

A 7 years old female presented to the outpatient department of Orthopaedics, JJ hospital, Mumbai, India with chief complaints of pain and swelling over her left distal forearm that was persisting since last 1 month.

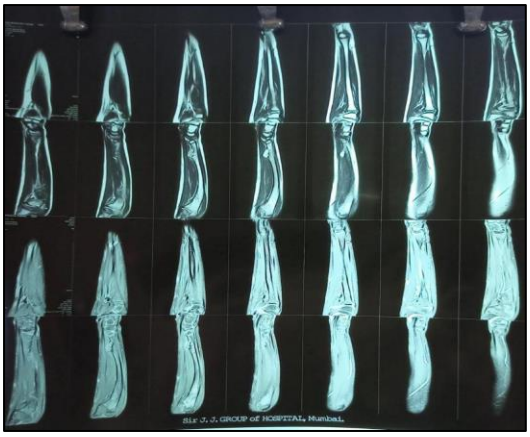

Figure 1: Distal ulna osteomyelitis MRI image sagittal view.

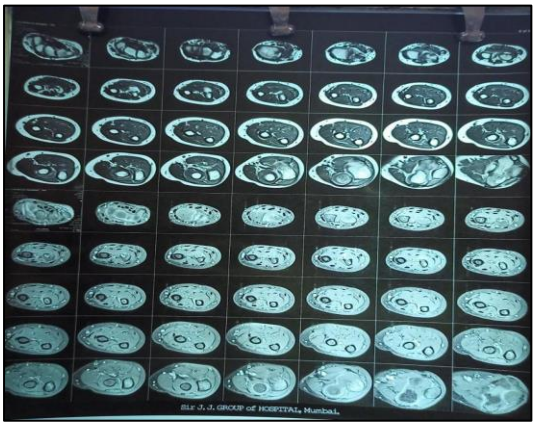

Figure 2: Distal ulna osteomyelitis MRI radiograph axial view.

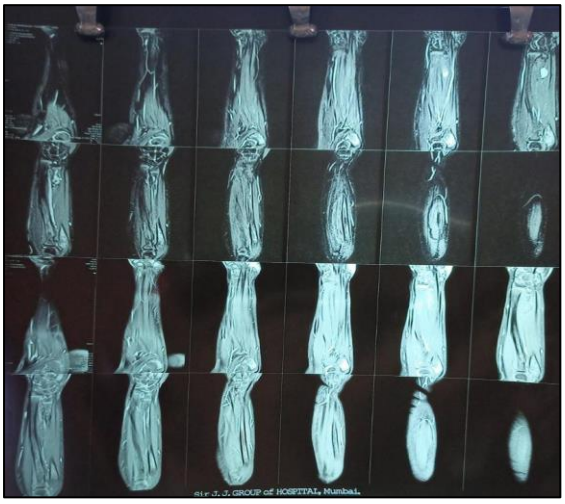

Figure 3: Distal ulna osteomyelitis MRI radiograph coronal view.

Pain started after a trivial trauma to left wrist, localized, diffuse, responding to NSAIDs with overlying skin erythematous and swollen. Patient had a history of left supracondylar non-displaced fracture 5 years ago for which the patient was treated conservatively with above elbow cast for 1 month.

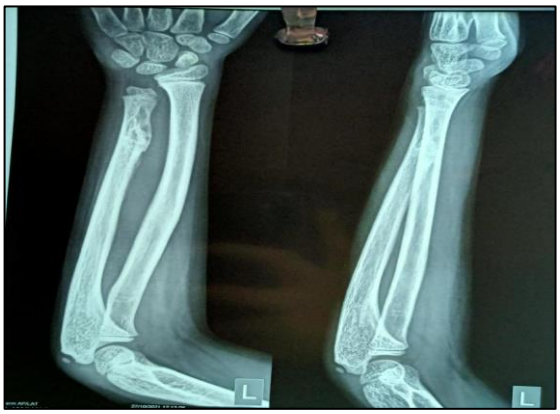

Figure 4: Pre-op X-ray radiograph of left side distal ulna osteomyelitis.

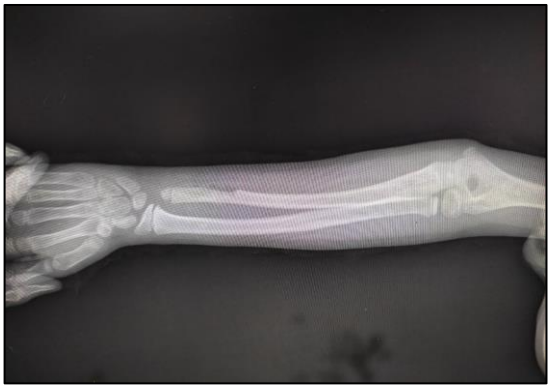

Figure 5: X-ray radiograph of left sided distal ulna osteomyelitis post curettage and biopsy.

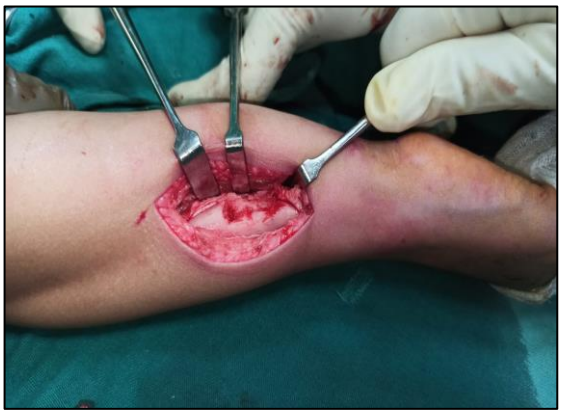

Figure 6: Intra-operative image of left distal ulna curettage and biopsy.

Again 2 years after the patient sustained injury to her left wrist and was diagnosed with greenstick fracture of distal ulna managed conservatively and since then the patient had largely been asymptomatic. Parents give a history of trivial trauma 1 month ago after which pain and swelling started. No significant personal or family history present

for the patient. Physical examination showed tenderness over the area and full range of motion at wrist joint with slight rise in local temperature. Constitutional symptoms of fever, weight loss, polyarthralgia and arthralgia were absent. Patient was having overall good general health with normal weight and paediatric milestones as per age.

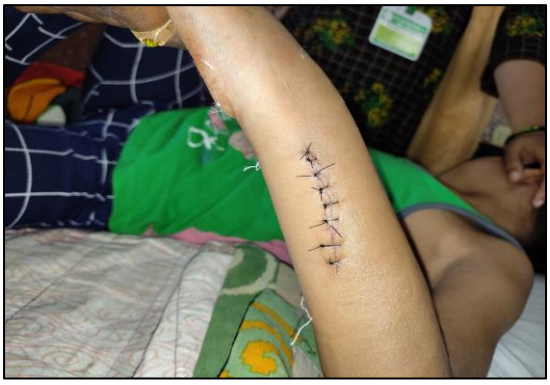

Figure 7: Post-operative check-dress of distal ulna osteomyelitis.

Laboratory results showed normal blood count with consistent negative CRP and normal ESR repeated every 3-5 days. RA factor and HLAB27 are negative as well. On the first presentation X-ray was performed which revealed an osteolytic lesion of distal ulna. (Figure 4 and 5) On suspicion of osteomyelitis, a contrast enhanced MRI was done which revealed conspicuous results. Altered marrow signal intensity involving distal metaphysis and diaphysis suggestive of marrow oedema. (Figure 1, 2 and 3) A 10×7 mm well defined peripherally enhancing collection along the anterior cortex of distal meta-diaphyseal region of ulna extending into medullary cavity through 3 mm cloaca. All these radiographic findings along with the clinical history strengthened the presupposition of chronic osteomyelitis.

For histopathological and microbiological evidence, the patient underwent saucerization and curettage under general anaesthesia. (Figure 6 and 7) An incision made over the ulnar aspect with blunt dissection of soft tissues and periosteum elevated to expose the lesion. Samples of bone taken and sent for biopsy and culture sensitivity. Biopsy showed unremarkable cartilaginous and collagenous tissues admixed with chronic lymphocytic inflammatory infiltration and no evidence of caseous necrosis, granuloma, atypia or malignancy. Tissue and pus c/s were negative for any organism. Genexpert for MTB was negative. All these findings validate the aseptic nature of lesions and signifies more towards inflammatory conditions.

DISCUSSION

CNO/CMRO is a diagnosis of exclusion and requires combined clinical, radiological and histopathological data to come to a final diagnosis. Lab tests are nonspecific and may have modest elevation in inflammatory markers like leukocytosis, ESR and CRP. Clinically symptoms may vary from unifocal to multifocal and persistent to relapsing course of the disease. CRMO is frequently associated with other autoimmune disorders like inflammatory bowel disease, peripheral arthritis, sacroiliitis, psoriasis, pyoderma gangrenosum, Takayatsu's arteritis. It is also considered as the paediatric equivalent of SAPHO syndrome.[6]

An initial investigation in such cases with bone pain is usually a plain X-ray which at an early stage may not show any significant changes but shows osteolytic or osteosclerotic lesions only in later stages of the disease.[7] On MRI, the investigation of choice, inflammatory lesions are hypointense in T1 and hyperintense in T2/STIR. Bone marrow oedema is also identified through altered marrow signal in contrast enhanced MRI. In culture, aseptic condition in the absence of any antibiotic coverage is necessary to rule out bacterial or fungal osteomyelitis. Genexpert done to rule out tubercular etiology of osteomyelitis. Histopathological findings in CNO are non-specific and usually show inflammatory infiltration. Bone biopsy is done primarily to rule out malignancy.

There's no existing guidelines to treat CNO at present and empirical treatment is provided to the patient. NSAIDs are the first line of treatment to relieve the pain as well as reduce the inflammatory process to minimize bone damage.[8] Oral corticosteroids may be used in cases of relapsing symptoms. Bisphosphonates are required in multifocal and spinal lesions. TNF Alpha blockers are used in patients not responding to aforementioned treatments.[9] MRI full body screening is required to visualise any new lesions that may occur.

In this case, the patient underwent empirical treatment with NSAIDs for a month followed by surgical intervention for saucerization and curettage for contour reduction and sampling of bone and pus for investigation. A long term follow up is required to evaluate for any relapsing symptoms.

Case of CNO is quite rare and usually diagnosis is difficult and takes a lot of investigations and long term follow up which are great limitations for research purposes due to migration of patients, loss of follow ups or economic constraints. Lack of enough previous literature on the topic has also led to less awareness among the fraternity.

CONCLUSION

CNO in children is an aseptic autoinflammatory condition of the musculoskeletal system with characteristic radiographic and pathological changes. Diagnosis can be made on detailed history, clinical, radiological and pathological features. Diagnosis is often delayed due to lack of awareness about the condition and the patient has to undergo numerous investigations and interventions. MRI is an excellent modality to delineate the extent of disease and its response to treatment. From our case report and review of literature we conclude that CNO is a

diagnosis of exclusion and responds well to NSAIDs and sometimes corticosteroids. CNO is often a benign disorder that may transform into more severe and persistent symptoms.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Gicchino MF, Diplomatico M, Granato C. Chronic recurrent multifocal osteomyelitis: a case report. Ital J Pediatr. 2018;44:26.
2. Giedion A, Holthusen W, Masel LF. Subacute and chronic "Symmetrical" osteomyelitis. Ann Radiol. 1972;15:329-42.
3. Björkstén B, Gustavson KH, Eriksson B, Lindholm A, Nordström S. Chronic recurrent multifocal osteomyelitis and pustulosis palmoplantaris. J Pediatr. 1978;93(2):227-31.
4. Oberc A, Sułko J. Chronic recurrent multifocal osteomyelitis (CRMO) in children - single-centre clinical observations and experience. Ortop Traumatol Rehabil. 2014;16:297-305.
5. Falip C, Alison M, Boutry N, Job-Deslandre C, Cotten A, Azoulay R et al. Chronic recurrent multifocal osteomyelitis (CRMO): a longitudinal case series review. Pediatr Radiol. 2013;43:355–375.
6. Jelušić M, Čekada N, Frković M, Potočki K, Skerlev M, Murat-Sušić S et al. Chronic Recurrent Multifocal Osteomyelitis (CRMO) and Synovitis Acne Pustulosis Hyperostosis Osteitis (SAPHO) Syndrome - Two Presentations of the Same Disease? Acta Dermatovenerol Croat. 2018;26(3):212-9.
7. Khanna G, Sato TS, Ferguson P. Imaging of chronic recurrent multifocal osteomyelitis. Radiographics. 2009;29:1159-77.
8. Schnabel A, Range U, Hahn G, Berner R, Hedrich CM. Treatment response and longterm outcomes in children with chronic nonbacterial osteomyelitis. J Rheumatol. 2017;44:1058-65.
9. Hedrich CM, Hahn G, Girschick HJ, Morbach H. A clinical and pathomechanistic profile of chronic nonbacterial osteomyelitis/chronic recurrent multifocal osteomyelitis and challenges facing the field. Expert Rev Clin Immunol. 2013;9:845-54.

Cite this article as: Mahajan NP, Sangma SM, Jarika AK, Mhatre JA. Chronic non-bacterial osteomyelitis of distal ulna in a paediatric patient: a case report and review of literature. Int J Res Orthop 2022;8:411-4.