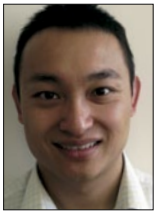
Chronic recurrent multifocal osteomyelitis



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Chronic recurrent multifocal osteomyelitis (CRMO) in children is an increasingly recognised non-infective, auto-inflammatory condition of bone that responds well to anti-inflammatory therapy but is difficult to diagnose, always by exclusion of other common pathology. The mean age of onset is reported to be pre-teen years (9-12 years), however CRMO can affect children of any age. While delays in diagnosis are understandable, delayed treatment can lead to long-term joint damage, pain, bone deformities, and growth impairment.

Presentation

Patients with CRMO commonly present with bone pain and radiologic bone abnormalities.

Patients can either have a single focus of disease, or multifocal areas (often asymptomatic and picked up on nucleotide bone scan). The common sites affected include ends of long bones, mandible, clavicle, and vertebrae.

CRMO may be associated with skin changes such as pustulosis and psoriatic skin/nail changes – the constellation of clinical findings is known as SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis, osteitis). CRMO is also associated with autoimmune conditions such as inflammatory bowel disease.

The reported course of disease is post-pubertal remission.

Diagnosis

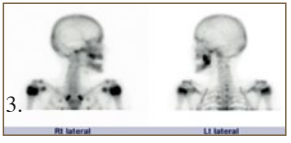
CRMO can be extremely difficult to diagnose, reached only after exclusion of infective and malignant causes. It is understandable that there is often a delay from onset of initial symptoms to diagnosis and treatment, as multiple investigations are required to exclude other common diagnoses including infective or malignant causes. After extensive investigations, patients are usually considered to have "atypical" chronic infective osteomyelitis.

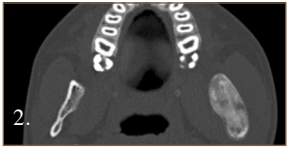
Nucleotide imaging usually reveals one or more focal vascular areas suggestive of infection or inflammation. Serology and biopsies of bone for infective causes (bacteria, tuberculosis, fungal infection) and malignant causes (e.g. osteosarcoma) are commonly performed and are always negative.

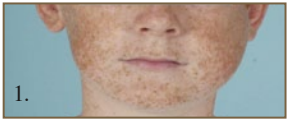
Patients are frequently started on long-term antibiotic treatment for chronic osteomyelitis, but fail to respond significantly.

Treatment

Due to the inherent chronic inflammatory nature of pathology, patients usually respond dramatically to anti-inflammatory therapy. First line therapy comprises of either NSAID or corticosteroid therapy. Generally, a high proportion of patients (85%) respond to NSAIDs alone. In the minority who do not have adequate response to first-line therapy, second-line agents such as methotrexate and bisphosphonates are considered. ■







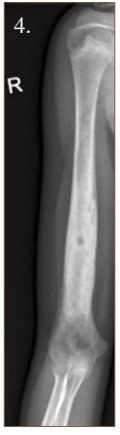


Figure 1. A 14-yo boy with CRMO. Picture shows a large hard bony swelling of the left cheek, due to firm bony enlargement of the left mandible.

Figure 2. CT of mandible of 14yo boy, showing expansion of left mandibular rami, with thickened cortex, and extensive new bone formation.

Figure 3. Nucleotide Bone Scan of 14yo boy, showing increased vascularity in left mandible. No other focus was identified.

Figure 4. X-ray of a 10yo girl with CRMO – large bony swelling of the right mid-distal humerus, with associated synovitis and fixed flexion deformity of her right elbow. Bone biopsy site (central circular area of lucency) is seen.