

Case 14212

Chronic Recurrent Multifocal Osteomyelitis (CRMO) of the clavicle

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Section: Musculoskeletal System

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Patient: 11 year(s), female

Clinical History

An 11-year-old girl presented with a progressive, painless swelling of the medial side of the right clavicle (Fig. 1). There are no systemic complaints and her previous medical history is unremarkable.

Imaging Findings

Standard radiography shows bone expansion at the medial side of the right clavicle. There is extensive bone sclerosis with a solid periosteal reaction (Fig. 2).

Subsequent Computed Tomography (CT) confirms the sclerotic expansile lesion in the right medial clavicle (Fig. 3). Magnetic Resonance Imaging (MRI) shows hypo-intensity of the lesion compared to normal bone marrow on T1-weighted image (Fig. 4) and hypo-intense signal compared to fluid on fat suppressed (FS) T2-weighted image (Fig. 5). Slightly heterogeneous enhancement is seen on T1-weighted image after gadolinium contrast administration (Fig. 6). Biopsy of the lesion reveals no arguments in favor of malignancy on histopathological examination. Culture of the biopsy specimen is negative with absence of growth of micro-organisms. Upon careful retro-anamnesis, the patient remembers having felt vague pain episodes at the right knee and ankle. Complementary conventional radiography show inhomogenous sclerosis at the distal femoral metadiaphysis (Fig. 7) and distal tibial metadiaphysis (Fig. 8).

Discussion

CRMO is an auto-immune disease with recurrent inflammatory lesions of the bone and can be

classified in the spectrum of spondyloarthropathy. The first case was reported by Giedion in 1972[1]. Since then 250 cases have been reported but the true prevalence is estimated to be higher. CRMO typically affects children while the adult counterpart of the disease is known as SAPHO syndrome (Synovitis, Acne, Pustulosis, Hyperostosis and Osteitis).

Patients with CRMO typically present with complaints of progressive swelling and pain. The proximal or distal tibial metaphysis, distal femoral metaphysis or medial clavicle are sites of predilection. If the clavicle is affected this may lead to reduced shoulder mobility and thoracic outlet symptoms. CRMO is the most frequent non-neoplastic disease of the clavicle in children less than 20 years old [2]. In most patients, multiple bones are synchronously or metachronously affected with both active and 'silent' lesions. Search for multifocality and recurrence is of utmost importance for a correct diagnosis of the disease.

No infectious etiology has been proven. Positive microbiology tests result from contamination of skin organisms. Histology shows inflammatory changes and granulocytic infiltration with polymorphonuclear leukocytes and osteoclastic bone resorption in the active phase of CRMO [3].

Radiographic findings depend on the stage of the lesions. Initially, an osteolytic lesion is found sometimes associated with periosteal reaction having a so-called "onion ring" appearance. Later on, there is progressive sclerosis of the bone. CRMO of the clavicle typically affects the medial part, whereas the lateral portion is spared. The sternoclavicular joint is normal in children whereas adults may show arthritic changes or ligamentous ossification as part of the SAPHO syndrome. Active lesions show bone marrow edema which is hypo-intense on T1-weighted images (WI) and hyperintense on FS T2-WI MRI images. Transphyseal invasion may be visualized. Progressive sclerosis results in hypointense signal of the lesion on both pulse sequences as seen in subacute and chronic lesions. There is no strict correlation of activity on imaging with clinical symptoms. Clinically silent lesion can be active on imaging. Whole body MRI or ^{99m}Tc bone scintigraphy may be useful tools for demonstrating multifocality [4].

Nonsteroidal anti-inflammatory drugs will relieve pain and swelling in 80% of the patients. Bisphosphonates can be added in case of failure of NSAIDs, but caution is warranted for possible jaw necrosis. TNF blockers are a third option but are avoided due to associated risk of infections and malignancy [5].

Final Diagnosis

Chronic Recurrent Multifocal Osteomyelitis

Differential Diagnosis List

Ewing's sarcoma, Osteosarcoma, Infectious osteomyelitis, Langerhans cell histiocytosis

Figures

Figure 1 Clinical image



Clinical image of the patient with a marked swelling of the medial side of the clavicle (white arrow)

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Area of Interest: Musculoskeletal bone;
Imaging Technique: Conventional radiography;
Procedure: Localisation;
Special Focus: Inflammation;

Figure 2 Standard radiography

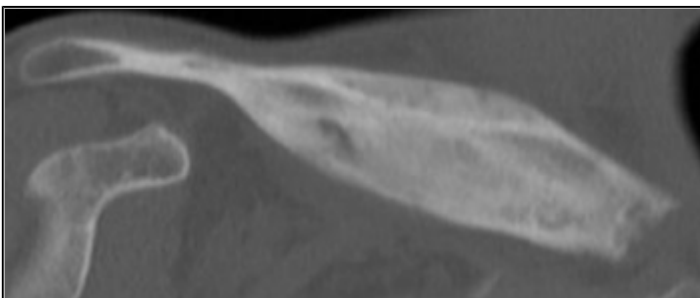


Standard radiograph shows an expansion of the bone at the medial side of the right clavicle. There is osteosclerosis of the bone with a solid periosteal reaction (arrow).

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Area of Interest: Musculoskeletal bone;
Imaging Technique: Conventional radiography;
Procedure: Diagnostic procedure;
Special Focus: Inflammation;

Figure 3 Computed Tomography

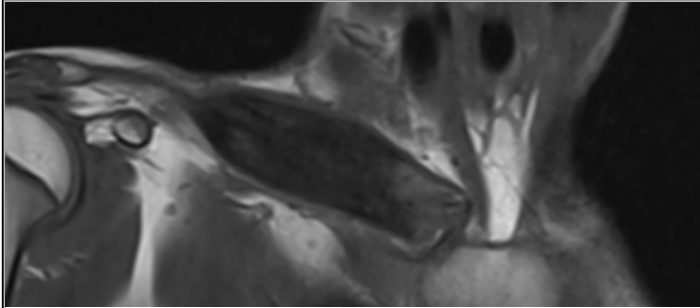


Computed tomography (coronal reformatted image). Inhomogeneous and sclerotic aspect with a solid periosteal reaction.

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Area of Interest: Musculoskeletal bone;
Imaging Technique: CT;
Procedure: Diagnostic procedure;
Special Focus: Inflammation;

Figure 4 Coronal T1-weighted MRI Image

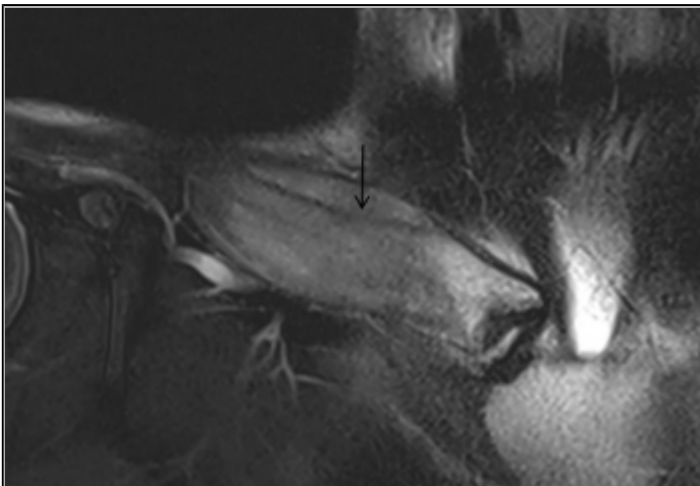


Coronal MRI image. The lesion is hypo-intense compared to bone marrow on T1-weighted images.

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Area of Interest: Musculoskeletal bone;
Imaging Technique: MR;
Procedure: Diagnostic procedure;
Special Focus: Inflammation;

Figure 5 Coronal T2 - weighted MRI image



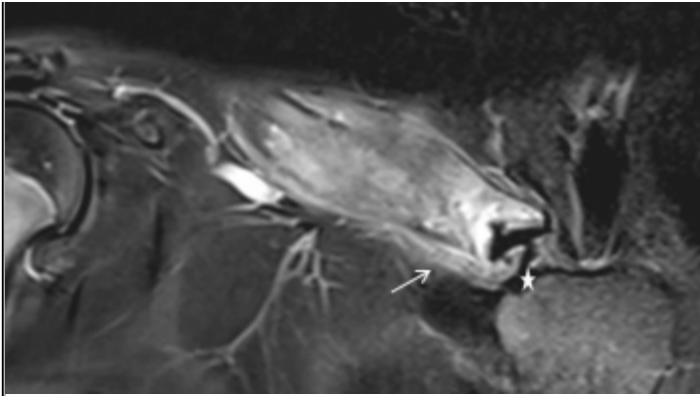
Coronal MRI image. The lesion remains relatively hypointense compared to fluid within the sternoclavicular joint on fatsuppressed T2-weighted images. Note osseous expansion of the lesion beyond the original cortex of the clavicle (black arrow).

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Area of Interest: Musculoskeletal bone;
Imaging Technique: MR;
Procedure: Diagnostic procedure;
Special Focus: Inflammation;

Figure 6 Coronal Fatsuppressed T1-weighted image after intravenous administration of gadoliniumcontrast





Note slightly heterogeneously contrast enhancement within the affected part of the clavicle, which is most prominent adjacent to the sternoclavicular joint. Note also subtle enhancement of the soft tissues adjacent to clavicle (arrow).

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Area of Interest: Musculoskeletal bone;
Imaging Technique: MR-Diffusion/Perfusion;
Procedure: Diagnostic procedure;
Special Focus: Inflammation;

Figure 7 Standard radiography femur



Standard radiography shows inhomogeneous sclerosis of the distal metaphysis and diaphysis of the right femur (predominantly of the dorsal portion) (arrow).

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Area of Interest: Musculoskeletal bone;
Imaging Technique: Conventional radiography;
Procedure: Diagnostic procedure;
Special Focus: Inflammation;

Figure 8 Standard radiography tibia



Standard radiography shows inhomogeneous sclerosis of the distal metaphysis and diaphysis of the right tibia (predominantly of the anterior portion) (arrow).

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Area of Interest: Musculoskeletal bone;
Imaging Technique: Conventional radiography;
Procedure: Diagnostic procedure;
Special Focus: Inflammation;

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Citation

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