

CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS MIMICKING OSTEIOD OSTEOMA

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In this article we illustrate an unusual case of chronic recurrent multifocal osteomyelitis (CRMO) in a 9 year old girl who presented with right thigh pain. The initial plain radiographs were normal. The white cell count was normal and there was a mild rise in C-reactive protein. Cross sectional imaging of the right femur showed a lesion with features suggestive of osteoid osteoma. However, when the lesion was excised, the appearances on histopathological examination were non-specific showing chronic inflammatory cells and the microbiological testing revealed no organisms. Six months later, this child presented with right shoulder pain and the subsequent imaging demonstrated bilateral clavicular lesions. At this stage, the diagnosis of CRMO was made based on the combination of the clinical, radiological, histopathological and microbiological features. This case demonstrates that the cross sectional imaging features of the bony lesion in CRMO can mimic osteoid osteoma.

Key-word: Bones, infection.

CRMO is an idiopathic inflammation of the bone that was first described by Giedion et al in 1972 as "sub-acute and chronic symmetrical osteomyelitis" (1). It has non-specific clinical, radiological and histopathological features (2). Therefore, it is a diagnosis of exclusion. The cause of CRMO remains unclear. The typical imaging findings of CRMO include lytic and sclerotic lesions in the metaphyses of long bones and the medial clavicles (3). In this article, we illustrate a case of CRMO in which the presenting bony lesion had imaging features which are characteristic of an osteoid osteoma.

Case report

A 9-year-old girl presented to her family physician with right thigh pain. The clinical examination was unremarkable. The plain radiographs of the hip and pelvis were normal. As the symptoms persisted, the patient was referred to the Orthopaedic team when laboratory investigations, MRI and CT of the right femur were performed. The blood tests showed normal white cell count and a mild rise in C-reactive protein.

MRI of the right femur demonstrated a focus of high signal intensity related to the posterior cortex of the proximal femoral diaphysis on the STIR sequences which was surrounded by bone marrow oedema

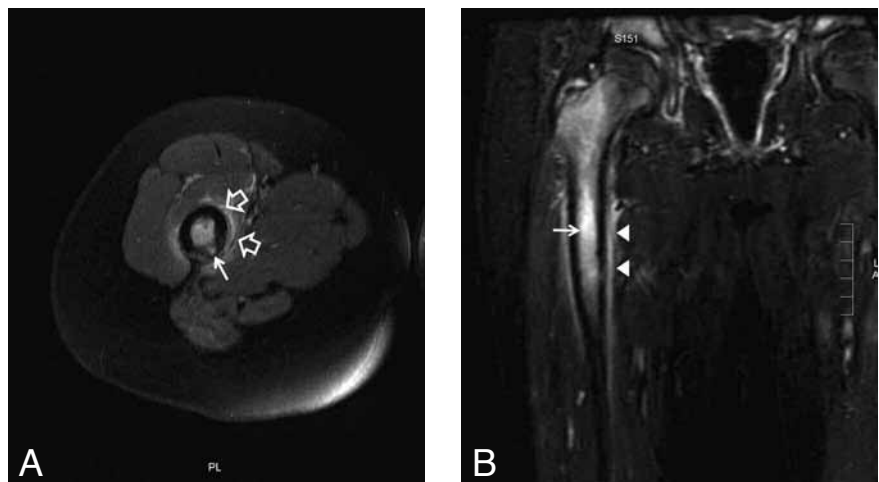


Fig. 1. — A. Axial STIR- MR image of the right thigh demonstrating a focus of high signal intensity within the posterior cortex of the femur (arrow) with bone marrow oedema and also high signal intensity (oedema) in the adjacent soft tissues (open arrows). B. Coronal STIR- MRI of the right thigh illustrating the extensive bone marrow oedema involving the proximal femur (arrow) and also the oedema in the surrounding soft tissues (arrowheads).

(Fig. 1). There was also a slight high signal change in the adjacent soft tissue of the thigh on the fluid sensitive sequences. CT was also performed for further characterisation. This showed a 10 mm radiolucent focus associated with cortical thickening in the same area of the proximal femur (Fig. 2). The appearances were reported as suggestive of osteoid osteoma.

Surgical excision of the right proximal femoral lesion was undertaken. However, the appearances on the histopathological examination were non-specific showing chronic inflammatory cells with no tumour cells identified. The microbiological testing revealed no organisms.

Six months later, the patient presented with right shoulder pain with swelling over the right clavicle. The plain radiograph demonstrated expansion of the medial end of the right clavicle which appeared sclerotic with small focal areas of radiolucency. The medial end of the left clavicle had a similar but less extensive appearance (Fig. 3). MRI scan was performed and demonstrated bone marrow oedema with

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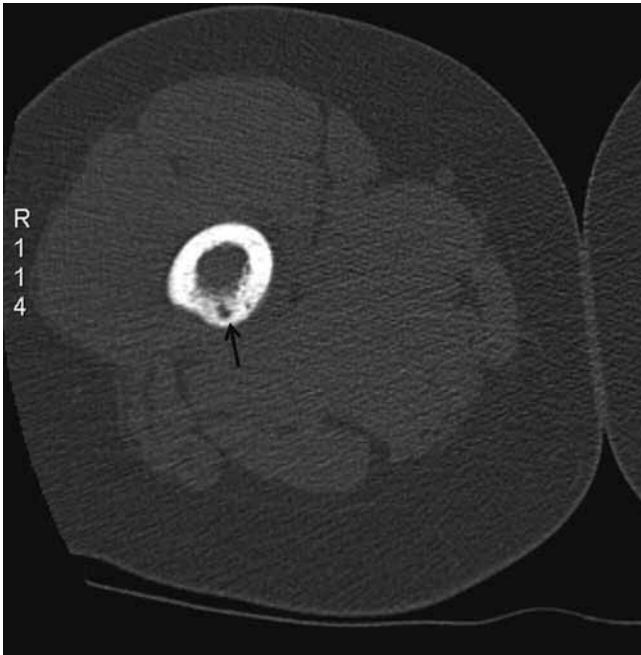


Fig. 2. — Axial CT image of the right thigh demonstrating a small lucent focus related to the posterior cortex of the proximal femoral diaphysis mimicking osteoid osteoma (arrow).

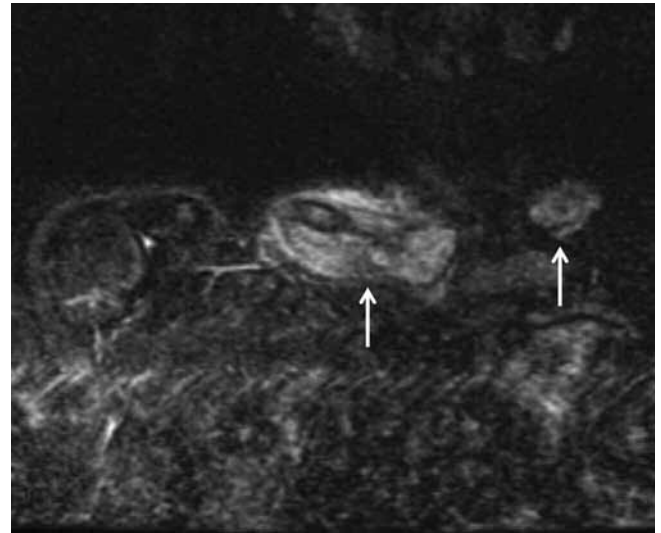


Fig. 4. — Coronal STIR- MRI sequences of the clavicles demonstrating bone marrow oedema and periosteal reaction with expansion of the medial end of the right clavicle and to a lesser extent the left clavicle which is associated with high signal within the surrounding soft tissues (arrows).



Fig. 3. — Plain radiograph of the right clavicle which also includes the medial end of the left clavicle. This shows expansion of the medial ends of both clavicles (arrows), more so on the right side, which appear sclerotic with foci of radiolucency. These appearances are typical of CRMO.

periosteal reaction and expansion of the medial end of the right clavicle and to a lesser extent the left clavicle which was associated with high signal within the surrounding soft tissues on the STIR sequences (Fig. 4).

A whole body radionuclide bone scan was obtained to search for other bony lesions. In addition to showing areas of high tracer uptake at the medial ends of both clavicles, the bone scan has revealed a focus of high uptake in the upper thoracic spine (Fig. 5). MRI scan of the thoracic spine was performed at a later stage and it demonstrated increased

signal intensity at T4 and T6 vertebral bodies on the STIR images with anterior wedging and collapse of T6 vertebral body resulting in kyphosis (Fig. 6).

At the stage when the right clavicular lesion was imaged, the diagnosis of chronic recurrent multifocal osteomyelitis was made. This was based on the multifocality of the disease process which involved the medial ends of both clavicles, the absence of abscess formation, the lack of a causative organism on microbiological testing and the non-specific histopathological findings

on the excised femoral lesion. The presenting right femoral lesion was in fact part of the CRMO disease entity and not an osteoid osteoma.

Discussion

CRMO is characterised by multifocal non-pyogenic inflammatory bone lesions and a course of exacerbations and remissions (3). The disease predominantly affects children and adolescents. It usually occurs in the latter half of the first decade and the first half of the second decade of life (mean age of 10.5 year) (4). It affects females more than males (5). The aetiopathogenesis of CRMO remains unclear and it is currently regarded as an auto-inflammatory syndrome (6). Reaching the diagnosis is important in avoiding unnecessary diagnostic procedures and to initiate appropriate therapy (7). The clinical presentation is non-specific with insidious onset of fever, local swelling and pain in affected bones. The laboratory findings are also non-specific often demonstrating raised ESR and CRP with a normal white cell count (5, 8). It has been reported that CRMO can be associated with skin lesions including palmoplantar pustulosis, psoriasis vulgaris, Sweet syndrome, and pyoderma gangrenosum (3, 9).

CRMO predominantly affects the metaphysis of long bones and the clavicles. The lesions can occur in

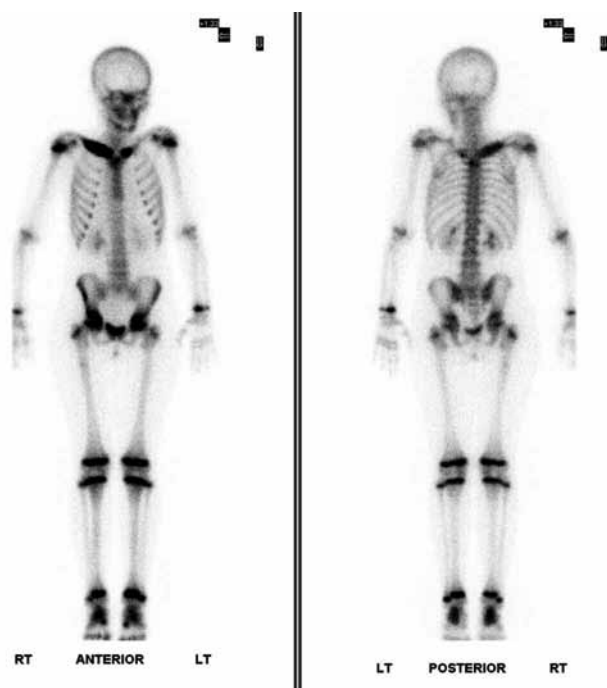


Fig. 5. — Whole body radionuclide bone scan showing high tracer uptake at the medial ends of both clavicles and at the 4th thoracic vertebra. There is also a subtle tracer uptake at the proximal part of the right femur, at the site of the surgically excised lesion.

other areas throughout the skeleton including the spine, pelvis, sacroiliac joints, ribs, sternum, scapulae, metatarsals, metacarpals, tarsal bones, phalanges and mandible (2).

Radiologists can be the first to suggest this diagnosis. Plain radiography typically demonstrates lytic lesions usually at the metaphysis of the tubular bone at the early stages of the disease. With healing, progressive sclerosis is usually seen around the lytic lesion with associated hyperostosis (3). During relapses, new lytic lesions appear with periosteal reaction resulting in bone thickening (2, 3). Spinal lesions are characterised by erosion of the vertebral end plate with adjacent sclerosis. CRMO lesion can lead also to vertebral collapse and subsequent kyphosis (2, 10).

The MRI appearances of the affected areas are non-specific showing abnormal hyperintensity on STIR images and contrast enhancement on T1-weighted images, which are features of oedema like lesions (11). MRI can also demonstrate periosteal reaction and soft tissue oedema (7, 12). Therefore, MRI is useful in monitoring disease activity (5). With healing, these MR features gradually disappear with

regeneration of normal marrow signal (5). MRI can also help in differentiating CRMO from infective osteomyelitis. This is particularly helpful in characterising the spinal CRMO lesion. This appears as irregular vertebral endplate with adjacent bone marrow oedema. The adjacent disc may become involved showing altered signal intensity but, unlike infectious osteomyelitis, the lesion does not cross the intervertebral disc and there is no abscess formation (3, 5). Given the multifocality of the disease and the lack of ionizing radiation with MRI, whole body MRI can have a role in establishing the diagnosis of this disease entity which predominantly affects the paediatric population (11). CRMO lesions can also be detected using radionuclide bone scan which can help in making the diagnosis and in showing the extent of the disease by identifying the multiple osseous involvement (4).

The histopathological findings in CRMO usually include non-specific subacute or chronic inflammation with intertrabecular spaces filled with fibrovascular material and inflammatory cells mainly lymphocytes (5, 8). Because the histopathologic features are non-specific, the



Fig. 6. — STIR sagittal MRI of the thoracic and cervical spine showing high signal intensity and collapse at T6 vertebral body (arrow) and also increased signal intensity at the superior end plate of T4 vertebral body (open arrow).

definite diagnosis should be made by combining the clinical picture, imaging studies, bacterial culture, and histopathologic analysis in a multidisciplinary approach (13).

The differential diagnosis includes multifocal bacterial osteomyelitis, multifocal trauma, stress fracture, juvenile idiopathic arthritis, histiocytosis, Scheuermann disease (the spinal lesion), leukemia, lymphoma, primary neoplasms (Ewing's sarcoma or osteosarcoma) and metastatic secondary malignancies (sarcoma, neuroblastoma) (2, 4, 5). This case has illustrated that osteoid osteoma is also part of the differential diagnosis.

In this case of CRMO, the right femoral lesion had a radiolucent cortically based focus that was associated with cortical thickening on the CT images. This focus had high signal intensity on the corresponding STIR-MR images. These features are uncharacteristic of CRMO. At the same time these appearances, and also the location of the lesion, are more typical of osteoid osteoma (14). Infective osteomyelitis in the form of intracortical abscess can mimic osteoid osteoma but it is not typical of CRMO to produce these features (14).

In conclusion, CRMO can create a diagnostic challenge due its non-specific manifestations and it is a diagnosis of exclusion. This case

illustrates that the cross sectional imaging features of the bony lesion in CRMO can mimic osteoid osteoma.

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