Spectrum of imaging features of SAPHO/ CRMO syndrome

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Authors: A. Kalpaxi¹, V. Bizimi¹, E. Skoufa¹, I. Misichroni¹, N. S. Siden², D. K. Filippiadis³, A. D. Keleakis¹, C. Kontopoulou¹, N. L. Keleakis¹, O. Papakonstantinou¹; ¹ATHENS/GR, ²Chaidari/GR, ³MAROUSI - ATHENS/GR
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Learning objectives

To present and discuss a spectrum of imaging findings in four patients (three adults and one child) diagnosed with SAPHO / CRMO syndrome.

Background

SAPHO is an acronym for the following conditions: synovitis, acne, pustulosis, hyperostosis and osteitis. It represents a spectrum of inflammatory bone lesions which may or may not be associated with dermatological lesions, with negative bacterial cultures and a clinical course characterized by relapses and remissions. The main target area of skeletal involvement is the anterior chest wall, followed by spine, pelvis and long bones.

Many authors believe that chronic recurrent multifocal osteomyelitis (CRMO) is the pediatric presentation for the SAPHO syndrome. The most common sites of skeletal involvement include the long tubular bones and clavicle, but lesions have been described throughout the skeleton.

SAPHO/CRMO syndromes are thought to be uncommon, but may be underdiagnosed because of their rarity and the lack of specific clinical, laboratory test or radiological findings.

Imaging findings OR Procedure Details

We reviewed the radiographic, CT and MR imaging findings of four patients, three adults and a child, with a final diagnosis of CRMO or SAPHO.

Patients presented with non specific complaints, such as pain, tenderness and swelling at the involved site or sites. All had a protracted clinical course characterized by cronicity.

The diagnosis was confirmed by means of bone biopsy and negative bone cultures in 3 of the 4 patients. Biopsy was not performed in patient 3, but the diagnosis was based on clinical observation over many years and pain relief from treatment by steroids.

Table 1 presents location of the lesions in each patient.
<table>
<thead>
<tr>
<th>Sex</th>
<th>Age at presentation</th>
<th>Skin lesions</th>
<th>Skeletal lesions</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient 1</td>
<td>Male</td>
<td>41</td>
<td>Erythematous papules and plaques at the site of previous bone biopsy (left clavicle)</td>
<td>Anterior chest wall Scapula Spine</td>
</tr>
<tr>
<td>Patient 2</td>
<td>Female</td>
<td>47</td>
<td>Generalized acne</td>
<td>Anterior chest wall</td>
</tr>
<tr>
<td>Patient 3</td>
<td>Female</td>
<td>50</td>
<td>No</td>
<td>Spine Pelvis</td>
</tr>
<tr>
<td>Patient 4</td>
<td>Male</td>
<td>13</td>
<td>No</td>
<td>Long bones</td>
</tr>
</tbody>
</table>

Table 1

Anterior chest involvement:

Anterior chest involvement was observed in two patients.

The first patient (Patient 1) had a long disease course with progressive appearance of new lesions over time. At presentation he had hyperostosis and osteitis affecting the medial and middle third of the left clavicle and the first rib on the left side, with involvement of the left sternoclavicular and first sternocostal junctions on both sides (Fig. 1 on page 6).

Nine years later a scapular radiograph showed additional sclerotic lesions in the glenoid cavity, the coracoid process and the spine of the left scapula. In the mean time the patient had an unspecified surgical resection of part of the left clavicle (Fig. 2 on page 6).

Eight years later, new lesions appeared on the left side (second rib and sternocostal joint) and on the right side (clavicle, first rib, sternoclavicular and first sternocostal joint) (Fig. 3 on page 7).

A bone scan obtained showed increased uptake in the right clavicle, both sternoclavicular joints, manubrium of the sternum, left 1st-3rd ribs and left scapula. The presence of increased activity at the sternoclavicular region, the so called "bull's head", is a specific manifestation of the SAPHO syndrome (Fig. 4 on page 8).
An #R imaging study of the thorax additionally revealed involvement of adjacent soft tissues, especially the muscles of the left scapula (supraspinatus, infraspinatus, subscapularis muscles) (Fig. 5). It also showed involvement of the first and second thoracic vertebra and corresponding rib vertebrae articulations, that was not clearly demonstrated in the radiographs.

The second patient (patient 2) had a more limited extension of the disease. A CT exam at presentation showed osteosclerotic lesions at the left side of the manubrium of the sternum and involvement of the left sternoclavicular junction. An MR and a bone scan confirmed the findings (Fig. 6 on page 10). The patient experienced chronic pain characterized by relapses and remissions but she did not develop additional sites of inflammation over time, during prolonged follow up.

**Comment:** Anterior chest involvement is the most common manifestation of the disease in adult patients (65-90%). It is characterized by osteitis and hyperostosis affecting the medial ends of the clavicles, the sternum and the upper ribs. Osteitis is imaged as osteosclerosis. Hyperostosis is the result of chronic periosteal reaction and cortical thickening, that leads to bone hypertrophy. Joint erosions of the adjacent articulations (sternoclavicular, upper costosternal, costochondral and manubriosternal) is a frequent finding, due to a primary arthritis or extension of adjacent osteitis. #ssification of the costoclavicular ligament is an important early finding.

Although all the abnormalities can be seen on plain radiographs, CT is the modality of choice to determine the extent of the lesions. MRI can reveal subclinical foci and identify active lesions by the presence of bone marrow edema. The bony changes may extend to involve adjacent soft tissues. Whole body scintigraphy can identify subclinical foci.

**Spine and ileosacral joint involvement:**

Two of our patients showed spine involvement, one had also involvement of the sacroiliac joints.

Patient 3 had extensive spine involvement. She had diffuse hyperostosis of several thoracic and lumbar vertebral bodies and costovertebral articulations (Fig. 7 on page 10).

She also showed involvement of the sacroiliac joints that was associated with more pronounced osteosclerosis of the iliac bone (Fig. 8 on page 12).

A bone scan showed increased activity in thoracic and lumbar vertebra, especially T11, and in sacroiliac joints.
As already mentioned, patient 1 showed involvement of the bodies of the first and second thoracic vertebra and costovertebral articulations, but anterior chest was the principal site of the manifestation of the disease.

**Comment:** In adults the spine (usually the thoracic spine, followed by the lumbar and cervical spine) and the sacroiliac joint are affected by the disease in 33% and 13-52% respectively. There are different radiographic manifestations: non specific spondylodiscitis, osteosclerosis of one or more vertebral bodies and paravertebral ossifications. Sacroiliitis is usually unilateral. Extensive osteosclerosis of the iliac bone is highly suggestive of SAPHO.

**Long bones involvement**

One of our patients was diagnosed with CRMO and long bones involvement.

Patient 4 was a 13-year-old boy that presented with pain and swelling at the ulnar side of the right wrist. A frontal radiography showed an expansile metaphyseal lesion with solid periosteal reaction (Fig. 9 on page 12).

MR imaging of the right forearm showed broadening of the ulna with cortical thickening along with endosteal and ulilamelar periosteal reaction, without cortical breach. Bone marrow was hypointense on T1 weighted image. On contrast enhanced T1 image with fat suppression, both bone marrow and adjacent soft tissues present diffuse uptake of contrast (Fig. 10 on page 14).

After five months, the swelling of the right wrist regressed, but a new lesion at the third metacarpal appeared.

An x-ray of the right hand showed expansion and cortical thickening of the distal metaphysis and diaphysis of the third metacarpal bone by a mixed sclerotic lesion with subtle lytic foci within. Solid periosteal reaction was apparent. The new lesion had similar clinical presentation and imaging features with the older self treated lesion of the ipsilateral ulna (Fig. 11 on page 14).

**Comment:** The metaphysis of the long bone, usually in the lower extremities, is the most common site of CRMO at initial presentation. The lesions are often lytic at the beginning of the disease. With time, progressive sclerosis is developed around the lytic lesion, resulting in progressive sclerosis and hyperostosis. Radiographs most frequently demonstrate association of lytic and sclerotic lesions. Frequently, soft tissue swelling is present, but there is no abscess formation.

In the acute phase MR imaging shows findings or marrow edema, which appears hypointense on T1 weighted images, hyperintense on T2 weighted images and presents
contrast enhancement. The sclerotic bone appears hypointense on T1 and T2 weighted images.

Long bone is a less common site of disease in SAPHO.

**Images for this section:**

**Fig. 1:** Chest X-ray: Bone hypertrophy and sclerosis of the left clavicle and first rib, with involvement of the left sternoclavicular and first sternocostal junctions on both sides.
Fig. 2: Left Scapula X ray 9 years later: New sclerotic lesions in the left scapula.
Fig. 3: X-ray 8 years later (17 years after presentation): Additional lesions in the left 2nd rib, right clavicle, right first rib and their respective joints (sternoclavicular and sternocostal)
Fig. 4: Bone scan: Increased activity in manubrium, both sternoclavicular joints, right clavicle, left 1st-3rd ribs and left scapula.
Fig. 5: A T1-weighted (right) and a T2-weighted with fat suppression (left) axial image at the level of the first sternocostal joint. There is thickening of the cortex of first ribs and sternum with bone hypertrophy (right) and bone marrow edema (right and left). The left scapula exhibits also bone marrow edema which extends to the adjacent soft tissues. Bone marrow and soft tissue edema are evident in left costovertebral joint (left).

Fig. 6: Bone scan: Increased radiotracer activity in the inferior aspect of the sternal manubrium.
**Fig. 7:** CT sagittal reformatted image of the spine: sclerotic lesions in many vertebral bodies. Disc spaces are preserved. Bridging osteophytes are shown at the L1-L2 level.

**Fig. 8:** CT pelvis: bilateral sacroiliitis, with sclerosis mainly involving the iliac side of the joint.
Fig. 9: #ray right wrist: lytic lesion with a sclerotic rim of the distal ulna, adjacent to the epiphysis.
**Fig. 10:** Axial precontrast T1 weighted MR image (left) and postcontrast fat saturated T1 weighted MR image (right) of the right forearm: Contrast enhancement of the ulnar lesion and the adjacent soft tissues.
Fig. 11: Xray of the right hand: Mixed sclerotic and lytic lesion with periosteal reaction of the distal metaphysis and diaphysis of the third metacarpal bone.
Conclusion

SAPHO and CRMO are diagnoses of exclusion. It is important that radiologists are familiar with the imaging findings of these diseases because they may be the first to suggest this diagnosis. The diagnosis SAPHO/CRMO should be suspected when the typical osteoarticular lesions are located in characteristic target sites and skin disease is present.

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Personal Information

This work comes from the Radiology Department of University General Hospital Attikon, Athens, Greece. First author: Radiology Resident M.Sc Angeliki Kalpaxi

aghiko@gmail.com.