Radiology-Rheumatology Rounds

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By:

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Case 1

• 60 y woman with weight loss and chronic back pain

• Past history:
  • long-standing postular lesions
  • 8 years ago → radiography and CT evidence of:
    » End-plate erosions
    » Diffuse sclerosis of some vertebral bodies
    » Anterior para-vertebral ossification
    » Soft tissue swelling
    » More prominent in the thoracic than the lumbar spine
• Open bone biopsy of T7 and T8:
  • Bone trabeculae moderately thickened
  • Sclerotic
  • Marrow fibrotic, chronic lymphocytic inflammation
  • No acute inflammation or necrosis
• Bone scan:
  • Increased uptake at:
    » Several vertebrae (dorsal and lumbar)
    » Manubero-sternal junction
    » Anterior upper ribs

• Lateral chest x-ray:
  – Severe erosive and hyperostotic changes at:
    » Sternoclavicular junction
    » Manubriosternal junction
    » Diffuse sclerosis of the manubrium
• Repeat radiographs:
  – Collapse of several vertebral bodies
  – Severe kyphosis
  – End-plate erosions and adjacent erosions
  – Narrowing of the dorsal disc spaces
  – Diffuse hypersostosis of vertebrae bodies
  – Osteophyte formation
  – Anterior para-vertebral ossifications
  – Superior wedging of L1
  – Normal SI joints
• MRI:
  • sagital and axial
  • Slice thickness of 3 mm and space of 0.5 mm

• Result:
  » Wedging of several vertebral bodies
  » End-plate irregularities
  » Papar-vertebral ossifications
  » Low signal intensity in lower thorasic and lumbar
  » High signal intensity at mid-thorasic area
  » Hypointensity signal of intervertebral discs
• MRI with gadolinium:
  » Focal or diffuse abnormal marrow
  » Mid-thoracic vertebrae → increased signal intensity
  » Diffuse paravertebral soft tissue swelling following gadolinium injection
SAPHO

- A conditions associated with:
  - Synovitis
  - Acne
  - Postulosis
  - Hyperostosis
  - Osteitis
• The term SAPHO first proposed in 1987
• Older name:
  » Chronic multifocal recurrent osteomyelitis
- **SAPHO syndrome:**
  - Inflammatory osteitis
  - Negative bacterial culture
  - Rarely *propionibacterium acne* isolation from bone
  - Main target: anterior chest wall
    » Osteosclerosis of the internal portion of the clavicles
    » Sclerosis of the axial skeleton
  - Palmoplantar pustolosis (PPP)
  - Acne (*conglobata, ulcerans, hydroadenitis suppurative*)
  - Various patterns of psoriasis
• Natural history not well understood
• No large follow up studies
• Possible association with:
  » HLA B27
  » IBD
  » Spondyloarthropathies
Fig 6. Technetium bone scan showing osteitis of the sternum and the proximal portion of both clavicles.
Fig 1. Radiography of the left clavicle showing SAPHO osteitis. The classical combination of osteosclerosis and hypertrophy are seen.
Fig. 1 Palmoplantar pustulosis (PPP). Sterile pustules on the soles of the feet surrounded by a mild inflammatory reaction.
Fig. 2 Acne conglobata. Extensive involvement of the back with multiple papules, pustules and haemorrhagic crusts
Fig 2. Palmoplantar pustulosis of the feet.
Fig 3. Computed tomography of sacroiliac joints showing bone sclerosis of the sacrum adjacent to the articular space, with apparent integrity of the ilium.
Fig 4. Unusual peripheral osteitis affecting the first metacarpal bone.
Fig 5. Osteitis of part of the left pubic symphysis.
Fig. 3 Oblique radiograph of the left clavicle showing increased density related to the medial end and the left first rib associated with an enthesopathy of the costoclavicular ligament
Fig. 4 Oblique radiographs of the right sternoclavicular joint (SCJ) showing A irregularity of the articular surfaces with early sclerosis in the juxta-articular bone due to SCJ arthropathy and B established sclerosis involving the medial end of the clavicle.
Fig. 11 Axial contrast-enhanced CT scan showing bilateral sternocostal ankylosis with extensive retrosternal soft tissue infiltration displacing the innominate vein.
Fig. 12 $^{99m}$Tc isotope scan. Delayed image of the anterior chest wall showing the classic "bull's head" sign due to increased uptake in the manubrium and both sternoclavicular joints.
Fig. 14  A Sagittal reformatted CT image showing irregularity of the superior vertebral end plate of T8 with loss of the disc space anteriorly. B Sagittal T2-weighted MR image shows uniform increase in signal intensity in the bodies of T9 and T10 with low signal intensity in the intervening disc.
Fig. 17 Sacroileitis with extensive osteosclerosis of the left ilium. Erosions and osteosclerosis of the end plates of L5/S1 with adjacent paravertebral ossifications are also seen.
Fig. 19 AP radiograph of the lower leg shows irregular chronic periostitis along the insertions of the interosseous membrane into both the lower tibial and fibular shafts.
Fig. 21 Oblique radiograph of the right ankle of a 21-year-old male showing A central erosions in the distal tibia with B progressive changes 5 years later with joint space narrowing, sclerosis and marginal spurring of both malleoli.
Fig. 22 AP radiograph of the right wrist showing erosions, extensive cartilage loss and subchondral sclerosis of the radiocarpal and carpal joints
Fig. 23 AP radiograph of the left shoulder showing a larger central subchondral erosion in the left humeral head together with cartilage loss and marginal osteophytes.
Fig. 24 A AP radiograph of the left hip of a 21-year-old male shows significant cartilage loss, central erosions of the acetabulum, and subchondral sclerosis. B Follow-up radiograph 5 years later shows progression with almost complete ankylosis.
Fig. 25  A Axial CT scan of a 4-year-old male showing irregularity of the iliac side of the right sacroiliac joint (SIJ) with a periosseous reaction in the adjacent ilium. B Axial T2-weighted MR image shows increased signal intensity related to the right SIJ, particularly in the ilium.
Fig. 26A, B Radiographs of a 5-year-old male who presented with multiple lesions. A AP film of the left wrist shows lytic lesions in the scaphoid and radial epiphysis. B AP film of the left elbow shows a large lytic lesion in the humeral metaphysis.
Fig. 27 Lateral radiographs of the proximal left tibia (A) and the distal left tibia (B) in a 13-year-old male showing multiple lytic lesions in the metaphysis with minor periosteal reactions.
Fig. 28  A AP film of the right ankle of a 2-year-old male shows a lytic lesion in the distal tibial metaphysis with an extension to the physis and no periosteal reaction. B T2-weighted MR image shows corresponding areas of high signal intensity but no marrow oedema, consistent with a chronic lesion.
Fig. 31A, B Sagittal MR images of the thoracic spine in a 19-year-old female. T1-weighted image (A) shows low signal intensity in the vertebral body adjacent to the superior end plate with corresponding high signal intensity in the T2-weighted image (B)
Fig 7. Radiograph of lower vertebrae showing sclerosing spondylitis, discitis, and syndesmophyte in a patient with the SAPHO syndrome.
Fig 8. Histology of late-stage SAPHO osteitis. Osteosclerosis predominates. Inflammatory cell infiltrate is absent (original magnification ×40).
Follow up of 120 SAPHO cases in France

- Patients diagnosed between 1974-1997
- Prospective F/U from 1992-1997
- 120 cases
- Data before 1997 analyzed retrospectively

- Objective:
  » To assess the long-term outcome of the SAPHO syn.
• Efficacy of drugs assesses
  – Efficacy Index:
    » EI= 0 → less than 30% improvement
    » EI=0.5 → partial efficacy
    » EI= 1 → more than 60% improvement
Table 1: Inclusion and Exclusion Features of the SAPHO Syndrome

<table>
<thead>
<tr>
<th>Category</th>
<th>Features</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>A. Inclusion features</strong></td>
<td></td>
</tr>
<tr>
<td>1. Osteoarticular manifestations of severe acne</td>
<td></td>
</tr>
<tr>
<td>2. Osteoarticular manifestations of palmoplantar pustulosis</td>
<td></td>
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<tr>
<td>3. Hyperostosis with or without dermatosis</td>
<td></td>
</tr>
<tr>
<td>4. Chronic recurrent multifocal osteomyelitis involving axial or peripheral skeleton, with or without dermatosis</td>
<td></td>
</tr>
<tr>
<td>One of these 4 items is sufficient to diagnose the syndrome, in the absence of an exclusion feature.</td>
<td></td>
</tr>
<tr>
<td><strong>B. Sometimes reported</strong></td>
<td></td>
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<tr>
<td>Possible association with psoriasis vulgaris</td>
<td></td>
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<tr>
<td>Possible association with inflammatory enterocolopathy</td>
<td></td>
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<tr>
<td>Features of ankylosing spondylitis</td>
<td></td>
</tr>
<tr>
<td>Presence of low virulence bacterial infections</td>
<td></td>
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<tr>
<td><strong>C. Exclusion features</strong></td>
<td></td>
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<tr>
<td>Septic osteomyelitis</td>
<td></td>
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<tr>
<td>Infectious chest wall arthritis</td>
<td></td>
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<tr>
<td>Infectious palmoplantar pustulosis</td>
<td></td>
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<tr>
<td>Palmoplantar keratodermia</td>
<td></td>
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<tr>
<td>Diffuse idiopathic skeletal hyperostosis</td>
<td></td>
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<tr>
<td>Osteoarticular manifestations of retinoid therapy</td>
<td></td>
</tr>
<tr>
<td>Gender</td>
<td>N</td>
</tr>
<tr>
<td>------------</td>
<td>----</td>
</tr>
<tr>
<td>Men</td>
<td>50</td>
</tr>
<tr>
<td>Women</td>
<td>70</td>
</tr>
<tr>
<td>Osteitis</td>
<td></td>
</tr>
<tr>
<td>Axial</td>
<td>92</td>
</tr>
<tr>
<td>Peripheral</td>
<td>7</td>
</tr>
<tr>
<td>Axial + peripheral</td>
<td>10</td>
</tr>
<tr>
<td>Absent</td>
<td>11</td>
</tr>
<tr>
<td>Arthritis</td>
<td></td>
</tr>
<tr>
<td>Axial</td>
<td>71</td>
</tr>
<tr>
<td>Peripheral</td>
<td>9</td>
</tr>
<tr>
<td>Axial + peripheral</td>
<td>31</td>
</tr>
<tr>
<td>Absent</td>
<td>9</td>
</tr>
<tr>
<td>Total</td>
<td>120</td>
</tr>
</tbody>
</table>

**Abbreviations:** PPP, palmoplantar pustulosis; PV, psoriasis vulgaris; SA, severe acne.
**Table 2: Main Clinical Features of 120 Patients With the SAPHO Syndrome**

<table>
<thead>
<tr>
<th>Feature</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender</td>
<td>70 F/50 M</td>
</tr>
<tr>
<td>Age at 1st symptom (years)</td>
<td>28.6 ± 13.7 (4-63)*</td>
</tr>
<tr>
<td>Age at SAPHO diagnosis (years)</td>
<td>37.7 ± 12.9 (5-67)*</td>
</tr>
<tr>
<td>Skin diseases (# of patients)</td>
<td>101 (84)</td>
</tr>
<tr>
<td>PPP, isolated</td>
<td>38 (32)</td>
</tr>
<tr>
<td>PV, isolated</td>
<td>12 (10)</td>
</tr>
<tr>
<td>SA, isolated</td>
<td>22 (18)</td>
</tr>
<tr>
<td>PPP + PV, isolated</td>
<td>21 (18)</td>
</tr>
<tr>
<td>SA, associated with PPP and/or PV</td>
<td>8 (7)</td>
</tr>
<tr>
<td>No skin lesions (nb of patients)</td>
<td>19 (16)</td>
</tr>
<tr>
<td>Age at first skin lesion</td>
<td>29.5 ± 13.6 (4-63)*</td>
</tr>
<tr>
<td>Osteitis (# of patients)</td>
<td>109 (91)</td>
</tr>
<tr>
<td>Age at first osteitic lesion (years)</td>
<td>33.0 ± 13.4 (6-67)*</td>
</tr>
<tr>
<td>Peripheral arthritis (# of patients)</td>
<td>41 (34)</td>
</tr>
<tr>
<td>Age at first joint lesion (years)</td>
<td>33.9 ± 13.4 (4-67)*</td>
</tr>
</tbody>
</table>

**NOTE.** Numbers in parenthesis expressed as percentages.

**Abbreviations:** PPP, palmoplantar pustulosis; PV, psoriasis vulgaris; SA, severe acne.

*Mean ± SD (range).*
Results

• Significant association between palmoplantar pustulosis (psoriatic vulgaris) and axial osteitis (P=0.007)
• No association between the dermatologic presentation and the severity of rheumatic symptoms
• No significant association between HLA B27 and the distribution of arthritis
• No severe or disabling complications
Results …

• Efficacy Index (EI):
  • NSAIDS → EI= 67%
  • Steroids → EI= 67%
  • Colchicine → EI= 36%
  • Sulfasalzine → EI= 16%
  • MTX → EI= 64%
  • Doxycycline → EI= 26%
  • Intra-articular injection of steroid → EI= 77%
Conclusion

• SAPHO syndrome is:
  • A stable disease
  • Good long-term prognosis
  • NSAIDS and intra-articular steroid more effective
  • Physicians to ask about “acne” or “pustolusis” history in any arthritis case
References

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• Skeletal Radiology (2003), 32

• Skeletal Radiology (1999), 28

• Rheumatology 2003, 42, pp 1398

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