Skeletal lesions in palmar-plantar pustulosis

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To link to this article: https://doi.org/10.3109/17453678408992376

Published online: 08 Jul 2009.

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In four women low-grade spondylitis-arthritis-osteitis coincided with palmar-plantar pustulosis. Both conditions are believed to be aseptic and part of a common immuno-defect reaction.

Palmar-plantar pustulosis (PPP) is a chronic skin disease with intra-epidermal abscesses on the soles and palms – predominantly the mid-palms, thenar regions, heels and insteps. Cultures from the pustules are negative for bacteria, virus and fungi (Everall 1957), and there is no effective treatment. The condition is related to but not identical with psoriatic pustulosis. The prevalence of PPP in Sweden is about 0.05%. The majority of the patients are women (75–82%) and the age at onset is usually between 40 and 60 years (Enfors & Molin 1971, Hellgren & Mobacken 1971).

Chronic recurrent multifocal osteomyelitis (CRMO), another aseptic condition, was first described in four children by Giedion et al. (1972). Another 16 cases were presented by Solheim et al. (1980). In this disorder the clavicles are commonly affected. Cultures from the bone lesions are nearly always negative. The patients are usually children or adolescents. In a Swedish study six of 13 patients had concomitant palmar-plantar pustulosis (Björkstén et al. 1978). Sonozaki et al. (1979) presented 22 patients with “inter-sterno-costoclavicular ossification”, many of whom also had signs of spondylitis, and found that not less than 13 also had a history or symptoms of palmar-plantar pustulosis.

In this paper we present case reports of four adult women with palmar-plantar pustulosis in conjunction with arthritis and/or osteomyelitis.

Case reports

Case 1. A 24-year-old woman presented with pain in the upper end of her sternum. ESR was 36 mm, leucocytes at most 13 400 with a slight eosinophilic shift and leucocyte antigen HLA B 27 negative. The physical examination revealed a swelling corresponding to the junction at the manubrium and the corpus sterni. The radiographic findings were most obvious on a lateral tomography which demonstrated widening and irregularity of the junction (Figure 1).

From the age of 13, this patient had had pustulosis, starting on her left foot and spreading to both hands and feet. She had been seen regularly by a dermatologist and had been treated with cortisone (perorally and local application) with doubtful effect.

Figure 1. Case 1. Tomography of junction manubrium-corpus sterni.
Skeletal lesions in palmar-plantar pustulosis

After a few months, the sternal pain subsided, and since then neither the skin nor the skeletal lesions have caused any substantial problems.

Case 2. A 40-year-old woman presented with back pain and fatigue. After a few weeks there was localized pain to the left in her chest close to the upper end of the sternum, and swelling and tenderness of her sterno-costal junctions. ESR was 50 mm, white blood count and distribution normal and leucocyte antigen HLA B\textsuperscript{27} negative.

There was intense pain for a few weeks and slow improvement with exacerbations up to 1 year, and during this time there was also low back pain. Radiographic examination of the spine was normal but tomography of the chest demonstrated changes similar to arthritis in the junction between the rib cartilage and the sternum (Figure 2).

A few weeks after the onset of pain, the patient developed skin lesions typical of palmar-plantar pustulosis on her left hand and foot and also a few skin lesions of para-psoriasis type on the trunk. After 1 year, both the para-sternal pain and the pustulosis lesions had disappeared. Ten years later, there was a mild exacerbation with para-sternal pain and a few pustulosis lesions.

Case 3. A 50-year-old woman presented with low back pain. Radiographic examination was negative but the symptoms persisted. Two years later, she also had an episode of pain in her left clavicle lasting several months. After one remission and one exacerbation, this pain subsided. Because of persistent symptoms from the back, another radiographic examination was carried out 3 years after the onset of back pain and discitis-type changes on the L III - L IV levels were observed (Figure 3). There were also small changes of a similar appearance in D X - D XI. \textsuperscript{99m}Technetium-MDP scintigraphy demonstrated increased uptakes at D X - L IV and in the left sterno-clavicular joint; radiographic examination demonstrated destruction of this joint.

This woman had had the diagnosis of palmar-plantar pustulosis for 12 years and had been treated with cortisone and antibiotics. She had had a remission and had then been free from symptoms until she developed low back pain and an exacerbation of the skin eruptions. ESR was 29, blood count was normal and the HLA B\textsuperscript{27} test negative.

Case 4. A 50-year-old woman presented with pain in her left clavicle with a normal radiographic examination. Three years later, increasing pain from both clavicles, back pain, abdominal pain and irradiating pain in her left leg were present. ESR was 65–80 mm, plasma electrophoresis indicated inflammatory reaction and leucocyte antigen HLA B\textsuperscript{27} positive.

Four months later, radiographic examination revealed spondylitis-like changes in the L IV - L V intervertebral disc (Figure 4). When the orthopaedic consultant prompted the patient to take her socks off for the neurological investigation, typical palmar-plantar pustulosis was found on her left heel and instep. A dermatologist verified the diagnosis; the lesion never spread outside this location. \textsuperscript{99m}Technetium-MDP scintigraphy demonstrated an increased uptake in the left clavicle and first and second ribs, and possibly in the second rib on the contra-lateral side plus increased uptake in the spine. A few months later, the patient also developed pain in her mandible with osteitis-like radiographic findings (Figure 5). This lesion was exposed by a dental surgeon and the bone tissue removed showed evidence of non-specific inflammation; cultures were negative. The CT-scan revealed sclerosis in the sternum and adjacent cartilage.

This patient still has back pain after more than 1 year, but the skin lesions are unchanged or even improved.
Figure 3. Case 3. Changes in lumbar spine 3 years after onset of pain.
A. AP-view.
B. Lateral view.

Figure 4. Case 4. Lumbar spine with changes in the L IV–V intervertebral space.
A. AP-view.
B. Lateral view.
Discussion

Chronic recurrent multifocal osteomyelitis (CRMO) usually occurs in childhood and adolescence (Giedion et al. 1972, Björkstén et al. 1978, Solheim et al. 1980). It is considered to be an aseptic condition since cultures are nearly always negative. There is usually an insidious onset with local swelling and pain in the affected bones. Sometimes there is a low grade fever and an elevated ESR (Björkstén et al. 1978). Radiographically, the lesions look like osteomyelitis with osteolytic areas surrounded by sclerosis. The lesions are often found in the metaphyses close to the epiphyseal cartilage, the clavicle being a favourite site for this disease. In a series of 101 lesions, 15 were located in the clavicle (Solheim et al. 1980). Our four cases are similar to CRMO since they also had an insidious onset with local pain and swelling and a moderately elevated ESR, but differed in some other aspects. The patients were all adults at the time of onset with a mean age of 41 (24–50) years and in addition to osteitis they had arthritis or spondylitis. The six cases of PPP and CRMO described by Björkstén et al. (1978) all had osteitis lesions in the clavicle and the sternum was also affected; there seems to be a strong association of PPP and the sterno-clavicular region. In a screening for clinical symptoms or roentgen tomography pathology in patients with PPP, Bergdahl et al. (1979) found sternum lesions in three patients and clavicular lesions in none. Two of our cases had spondylitis and so had two of the six in the series of Björkstén et al. (1978). Apparently, in all our cases there were pathological changes in the border-line between cartilage and bone, as in the metaphyseal lesions of children with CRMO.

The female dominance in our series is in accordance with the dominance of women among PPP patients (Enfors & Molin 1971, Hellgren & Mobacken 1971).

The combination of PPP and osteomyelitis in the clavicle has been described by Björkstén et al. (1978) and Sasaki (1967), who found a clinical appearance similar to chronic osteomyelitis. Our cases had arthritis and spondylitis, and in this respect they were more similar to psoriasis (Moll 1974). Psoriatic arthritis has also been observed in the sternoclavicular joint (Spar 1978). It is generally agreed that the two conditions, PPP and pustular psoriasis, are not identical but closely related (Rajka 1979).

The HLA B 27-antigen is frequent in patients with ankylosing spondylitis. In patients with psoriasis and arthritis, it is present in 21 per cent and in those with psoriasis and sacroiliitis in 33 per cent (Barraclough et al. 1977). The occurrence of HLA B 27 in one of our four patients does not permit any conclusions to be drawn.

The onset of the skeletal symptoms preceded the skin eruptions in two of our cases. One part of the symptoms does not necessarily cause the other – both may be due to interference with immunological factors and altered leucocyte activity (Molin & Rajka 1971, Rajka 1979). The arthritis in two of our patients became inactive – in one case despite active persistent pustulos. From our four cases – in addition to the data in the literature – it may be concluded that low-grade, osteitis-arthritis-like skeletal changes may be associated with palmar-plantar pustulosis. Inspection of palms and soles should not be neglected.
Acknowledgements

Financial support was obtained from the Swedish Medical Research Council (project No. B 83-17X-02737-15) and the Alfred Österlund, and Greta and Johan Kock Foundations.

References


