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The SAPHO Syndrome

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We report two cases of the SAPHO syndrome (synovitis, acne conglobata, pustulosis palmoplantaris, hyperostosis and osteitis). This syndrome has been published in the pediatric and rheumatological literature in recent years. ***Key words: Acne conglobata; Pustulosis palmoplantaris; Seronegative osteoarthropathies; Hyperostosis; Osteitis.***

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In 1985 Chamot et al. (3) reported an entity that has been called the SAPHO syndrome (synovitis, acne conglobata, pustulosis palmoplantaris, hyperostosis, osteitis). This syndrome can be found from 1961 in the literature under different names, as hyperostosis sternoclavicularis, osteomyelitis multifocalis chronica, ossificatio sternoclavicularis, pustulotic arthroosteitis or osteoarthrite pustuleuse. We report here 2 new cases.

CASE REPORTS

Case 1

A 14-year-old boy with a negative family history had suffered from pain in the back for half a year, and later on, in large and small joints. His first diagnosis at the pediatric department was juvenile chronic arthritis. At the same time, acne conglobata was observed on the trunk (Fig. 1).

Clinical investigation included x-ray, tomography and scintigraphy. Fig. 2 shows the x-ray findings.

In tomography, destructive inflammatory changes were noted in the spine and also on the sternal part of the clavicle with periosteal apposition. Furthermore, swelling of the periarticular tissue around the talocrural joint was present. The interpretation of this finding was multifocal osteomyelitis, hyperostosis and enthesopathy.

Through scintigraphy, a pathological increase of phosphates in sternoclavicular joints was noted.

There was no focal infection. *Propionibacterium acnes* and *Staphylococcus epidermidis* were found in skin lesions.

The laboratory tests were ESR 65 mm/h, WBC 16 800 mm^3. In electrophoresis a decrease of alpha-1-globulins, an increase of alpha-2-globulins, and a mild decrease of gamma globulins were found. ANF was mild-positive; other tests were negative. Furthermore, a decrease of T-lymphocytes and an increase of C_3 were found.

In the treatment Ibuprofen® (ibuprofenum) and Oxacilin® were successful. At relapse, a combination of the cephalosporine antibiotic Duracef® and the NSAID Naprosyn® (naproxenum) gave the best response. For acne manifestations Airol lotion was used.

Case 2

A 16-year-old boy had suffered from a congenital cataract and spine scoliosis from 10 years of age. Cystic acne appeared at 15 years on the trunk and face. Later, intense pain of the lumbosacral spine was noted. The sternoclavicular joint of the left side showed inflammatory swelling.

X-ray investigation confirmed scoliosis of the thoracic spine and osteolytic changes of the sternoclavicular joint.

Through scintigraphy, a high level of phosphates was found. No changes were found through thermography.

Ophthalmological findings included palpebral hordeolosis, congenital cataract and an artificial loss of lens.

In bacteriological examinations typical acne flora was found.

The laboratory tests were ESR 40 mm/h, antistreptolysin titer 380, latex test and ANF negative. Immunoglobulins, C-reactive protein and circulating immune complexes were normal, C_3 0.92, C_4 0.31 g/l, HLA B 27 negative.

As in the previous case, Oxacilin® and Ibuprofen® treatment for several weeks gave the best result. The patient was also given external treatment and long-term rehabilitation.

DISCUSSION

The first clinical signs of the SAPHO syndrome are usually a migratory pain of the joints or spine. Later, a true rheumatoid osteoarthritis with hyperostosis is found. Painful swelling of the sterno-costo-clavicular junction appears to be the most typical symptom. Sacroiliitis, spondylitis or involvement of other various juxtaarticular areas may also be found. Ostitic lesions dominate, whereas joint cartilages and muscles are spared.

Insertion of tendons or ligaments can also be affected (enthesopathy) (1–3, 5). In the early stages of the SAPHO syndrome, aseptic chronic recurrent multifocal osteomyelitis can be found (1, 3, 11).

Skin involvement includes severe forms of acne vulgaris, occasionally suppurative hidradenitis (3, 9) or pustulosis pal-

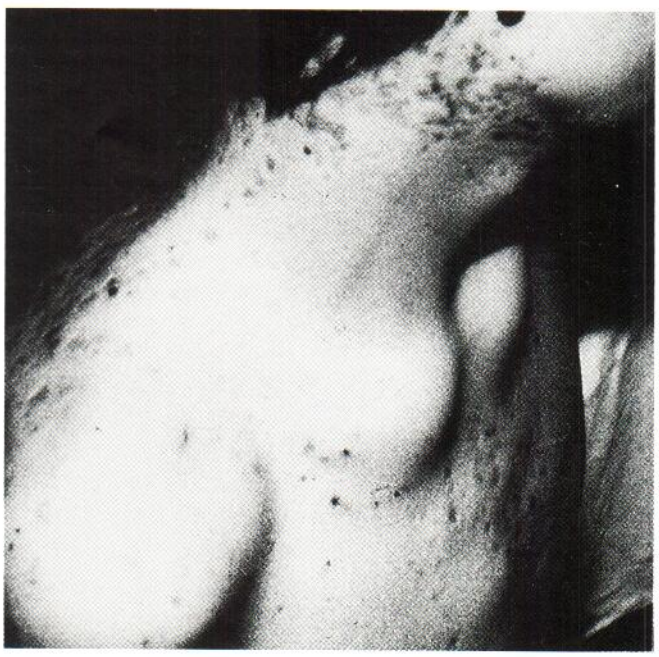

Fig. 1. Acne lesions and swelling of sternal parts of clavicles.

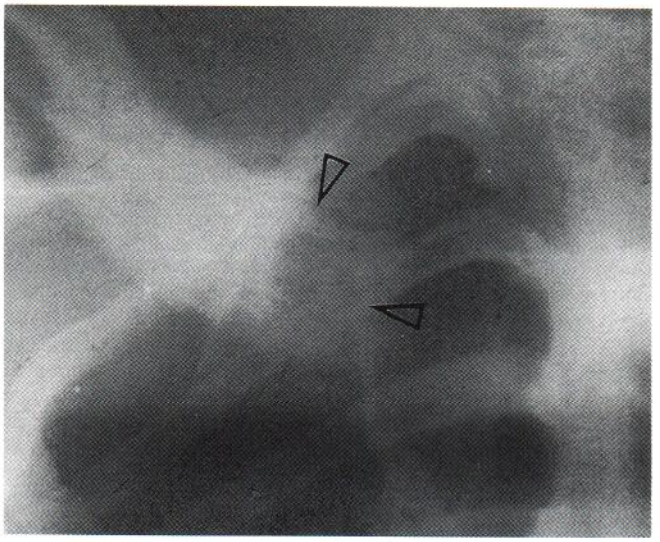

Fig. 2. Roentgen assessment: The clavicular bone is stronger on the sternal parts (right: 35 mm, left: 27 mm). The contours on both sternal ends are not very clear. The external half of the bone structure is normal in comparison with the medial, where pathological changes are seen, including the existence of cystoid formations.

moplantaris. A correlation between skin lesions and rheumatoid osteitis exacerbations was observed in our 2 patients. Cases of the SAPHO syndrome without skin manifestations are also described in the literature (3, 4, 12).

The pathogenesis of the SAPHO syndrome is not yet clear. It seems to be a seronegative osteoarthropathy and therefore it is close to psoriatic arthritis. HLA antigen B 27 was found in only 9–19.2% of the cases (2, 3). Frequently, increased circulating immunocomplexes are found (6–9). The incidence of aseptic bone and skin abscesses might be caused by a dysfunction of polymorphonuclear leucocytes (1, 2), whereas osteoarthritis seems to be a consequence of a focal infection (2, 3, 5, 10, 12). Immunological changes are supposed to be the result of hypersensitivity to bacterial antigens in the skin.

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