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Case Report

SAPHO Syndrome of the Temporomandibular Joint Associated with Sudden Deafness

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Summary: We report a case of arthritis of the temporomandibular joint (TMJ) associated with sclerosing osteomyelitis of the mandible and temporal bone, causing deafness. The presence of a palmoplantar pustulosis established the diagnosis of SAPHO syndrome. SAPHO (an acronym referring to synovitis, acne, palmoplantar pustulosis, hyperostosis, and osteitis) syndrome is defined by the association of characteristic osteoarticular and dermatologic manifestations, with diffuse sclerosing osteomyelitis of the mandible being a part of this entity. We review the literature of SAPHO syndrome with mandibular manifestations and discuss the mechanisms of inflammatory spread from the TMJ to the cochlea. To our knowledge, this is the first description of skull base involvement in a patient with SAPHO syndrome leading to sudden deafness.

The term SAPHO syndrome was coined in 1987 (1, 2) to describe an association of characteristic bone, joint, and skin lesions. The acronym SAPHO refers to synovitis, acne, palmoplantar pustulosis, hyperostosis, and osteitis. Not all these clinical manifestations need to be present to establish the diagnosis (1, 3) (see Table). The main characteristic features of skeletal manifestations are hyperostosis intermixed with areas of osteolysis and arthritis, most frequently located at the anterior chest wall (sternoclavicular, sternocostal, and manubrium regions), the pelvic girdle, and the spine (1, 4). Various patterns of psoriasis and severe acne can be associated with these bone lesions. Articular manifestations have been reported in the sternoclavicular, sacroiliac, wrist, and hip joints. An aseptic skeletal inflammatory process is the common denominator of these manifestations. The radiologic pattern of the skeletal lesions may be helpful in the diagnosis, especially if the sternoclavicular joint and the sternum are involved. However, SAPHO syndrome may be misdiagnosed if the location is atypical or if the clinical presentation is unusual (4). In this article, we describe a patient with SAPHO syndrome of the temporomandibular joint (TMJ) region in whom deafness was the major sign.

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Case Report

A 57-year-old man was referred for sudden left-sided deafness after a 4-year history of progressive hearing loss and tinnitus. He also reported occasional brief attacks of vertigo for 1 year, left-sided TMJ pain at the mouth opening for 4 years, which had been treated as internal derangement, and psoriasis and palmoplantar pustulosis for 10 years. He did not have weakness, nocturnal sweats, or fever. His medical history was otherwise unremarkable, except for multiple dental extractions. At physical examination, a soft-tissue swelling was noted in the region of the left TMJ without cervical adenopathy. Erythrocyte sedimentation rate was elevated, and C-reactive protein rate was increased (6.4 mg/L). Blood counts and other laboratory tests were normal. Lumbar puncture revealed no growth on culture, no abnormal cells (less than one cell per mm^3), normal glucose protein, and chloride. Lyme and syphilis tests of the CSF and serum were negative.

Contrast-enhanced MR imaging of the brain showed focal enhancement of the dura covering the endocranial part of the left temporal bone, extending into the internal auditory meatus to the modiolus and cochlea (Fig 1A). Associated enhancement of the epitympanic part of the tympanic cavity was shown. The T2-weighted images showed normal high signal of the labyrinthine fluid and normal vestibulocochlear nerve. T1-weighted images showed a large soft-tissue mass involving the region of the left TMJ (Fig 1B). The capsule of the TMJ was thickened and enhanced intensely. No TMJ effusion was noticed. The bone marrow of both the mandibular ramus and the condyle displayed abnormal low signal intensity on T1- and T2-weighted images, and did not enhance after contrast administration.

High-resolution CT (performed the same day as the MR study) showed cortical bone erosions of both the temporal squama and the condyle and diffuse endosteal sclerosis involving the mandibular condyle and ramus (Fig 1C and D). The temporal squama was partially resorbed with areas of bone remodeling. The lesion of the temporal squama extended posteriorly to the petrotympanic suture, sparing the tympanic and petrous bones, anteriorly on the greater wing of the sphenoid, and inferomedially on the sphenoid bone. The glenoid fossa was enlarged relative to the normal contralateral one, and no intra- or periarticular calcifications were evident. The normally aerated middle ear was partially filled by a soft-tissue mass, engulfing the lateral part of the incudomalleal joint. Neither the soft-tissue mass nor bony erosions were present in the external auditory canal.

Results of high-resolution CT of the chest and bronchoscopy were normal. A transbronchial biopsy specimen showed no evidence of tuberculosis, sarcoidosis, or malignant neoplasm. The examination of CSF was normal. A biopsy specimen of

Current criteria for SAPHO syndrome*

Chronic recurrent multifocal osteomyelitis
Usually sterile†
Spine may be involved
With or without skin lesions
Acute, subacute, or chronic arthritis with any of following:
Palmoplantar pustulosis
Pustular psoriasis
Severe acne
Any severe osteitis‡ with any of following:
Palmoplantar pustulosis
Pustular psoriasis
Severe acne

* Any of the three presentations is sufficient for the diagnosis (from Kahn [13]).

† Or with the presence of *Propionibacterium* acnes.

‡ Involvement of a single site, including spondylodiscitis, is sufficient.

the left TMJ capsule showed a dense fibroadipose vascularized tissue without granulomas. Immunohistochemistry tests and acid-fast bacilli and fungi cultures were negative.

One year later, the cutaneous lesions improved under PUVA therapy. The attacks of vertigo disappeared, while the left-sided deafness and tinnitus persisted. A follow-up MR study showed complete regression of the meningeal and labyrinthine enhancement and a decrease of the infratemporal soft-tissue mass. The signal of medullary bone of the mandible remained unchanged.

The diagnosis of SAPHO syndrome of the TMJ region involving the skull base was based on the following criteria: psoriasis with palmoplantar pustulosis, sclerotic mandibular bone, and articular lesions.

Discussion

SAPHO syndrome is a rare disease of unknown origin (4). Many different terms have been applied to this syndrome, which consists of associated bone and skin abnormalities (Fig 2), including subacute chronic bilateral osteomyelitis, acute pseudoseptic arthritis and palmoplantar pustulosis, arthroosteitis with pustulosis palmoplantaris, acquired hyperostosis syndrome, and diffuse sclerosing osteomyelitis (3). SAPHO syndrome is distinct from psoriatic arthritis, but both diseases share common features: 2% of patients with psoriatic arthritis have features of SAPHO syndrome (5), and some sternoclavicular and manubrial inflammation occurs in association with psoriasis vulgaris (1). On the other hand, psoriatic arthritis most frequently involves the joints of the hands and feet and on rare occasions it may affect the TMJ, usually unilaterally (6). Typical radiologic findings of TMJ involvement are marginal erosions, flattening, and cortical sclerosis of the condyle (7–9).

The fundamental component of SAPHO syndrome is an inflammatory, pseudoinfectious, sterile osteitis (1). Its clinical course is rather indolent, chronic, and self-limited. The most characteristic hallmarks of the bone lesions observed in SAPHO

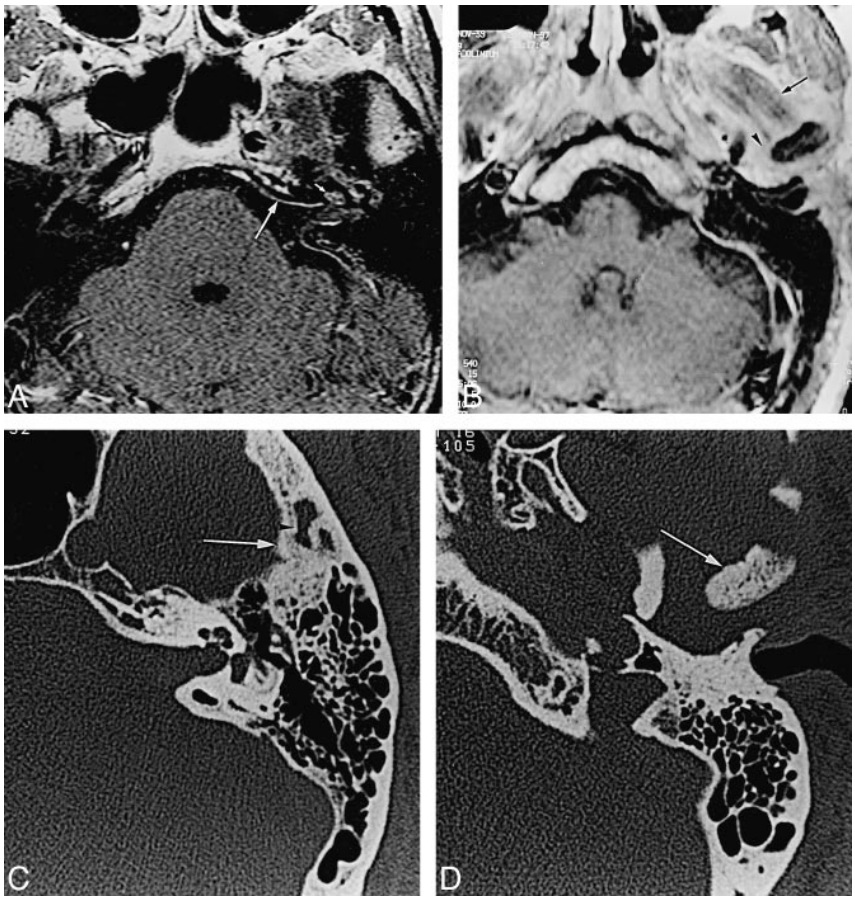

FIG 1. 57-year-old man with SAPHO syndrome.

A, Axial contrast-enhanced T1-weighted MR image (500/15) reveals focal enhancement of the dura (*large arrow*) of the temporal bone, spreading into the internal auditory meatus with enhancement of the cochlea (*small arrow*). An enhancing soft-tissue mass of the middle ear, outlining the bony ossicles, is also present.

B, Axial contrast-enhanced T1-weighted MR image of the infratemporal fossa (540/15) (1 cm below *A*) shows a large soft-tissue mass around the TMJ (*arrowhead*), involving the lateral pterygoid muscle (*arrow*). Note the low intensity signal of the bone marrow of the condyle.

C, Axial CT scan of the left temporal bone shows abnormal endosteal sclerosis of the temporal squama (*arrow*) with subcortical erosions (*arrowhead*). A soft-tissue mass (*arrowhead*) engulfs the incudomalleal joint.

D, Axial CT scan (2 cm below *C*) shows osteosclerosis of the mandibular ramus (*arrow*).

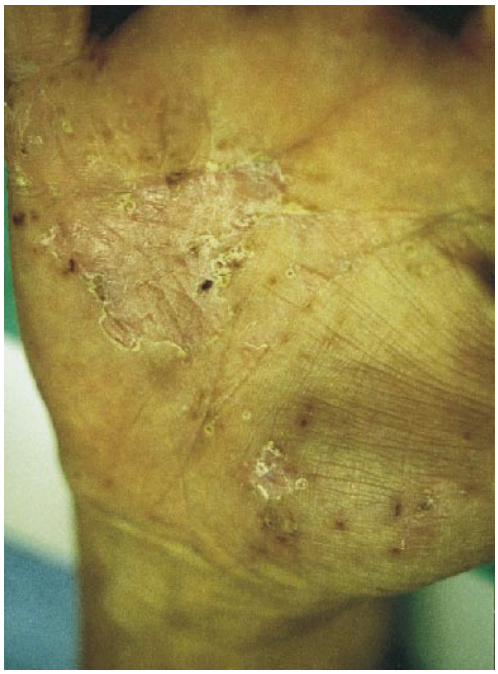

FIG 2. Palmoplantar pustulosis. Characteristic aseptic pustular and hyperkeratotic lesions involve palmar surface of the hand. Similar lesions were present on the soles of the feet of this patient (reproduced by permission of Dr. F. Hayem, Paris).

syndrome consist of areas of osteosclerosis with hyperostosis and periosteal reaction intermixed with areas of osteolysis, associated with arthritis of the adjacent joint, occurring most frequently in the sternoclavicular and anterior chest wall regions (70% to 90% of patients) (1, 2, 10). Spine, pelvic girdle, and peripheral bones are also affected (1, 2, 10), with the peripheral and axial locations being involved equally in about 30% of patients. Other causes of osteosclerosis, including osteomyelitis, sarcoma, osteoblastic metastases, sclerotic myeloma, myelofibrosis, and monostotic lesions of fibrous dysplasia, must be excluded. Bone biopsy may be helpful for excluding osteomyelitis or bone malignancy if no skin lesions are present (3, 11). However, histology is nonspecific, showing the occasional presence of lymphocytes, histiocytes, and polymorphonuclear cells. Bone scintigraphy may be suggestive, showing intense tracer uptake with a typical pattern of distribution and may display further asymptomatic bone lesions (3).

As described previously (see Table), the association of osteitis and cutaneous lesions is sufficient for the diagnosis of SAPHO syndrome, but skin lesions may be absent or appear after some delay or may be so subtle as to escape attention (11).

The origin and pathogenesis of SAPHO syndrome are unknown. No organism has been isolated in most of the cases of synovial and osseous inflammation. The theory of a genetic predisposition or an autoimmune response to a microorganism in the skin mimicking a normal bone or joint molecular structure has been proposed (11).

A mandibular manifestation of SAPHO syndrome has been described in 10% of cases (1). The bone lesions are similar to those reported by stomatologists for years as diffuse sclerosing osteomyelitis of the mandible (DSOM) (12). Moreover, in a series of 85 patients with SAPHO syndrome (13), seven had mandibular lesions similar to those reported in DSOM. The striking radiologic features are diffuse sclerosis or intermingled sclerotic and lytic lesions, leading to a deformity of the mandible in the end stage. A history of repeated dental extractions has frequently been noticed.

The findings in our patient are unusual because of the temporal bone and dural involvement. To our knowledge, extension to the bones of the skull base and the dura has not been described in association with a mandibular location of SAPHO syndrome (12, 13). This may be partly explained by less extensive investigation of the temporal bone in other cases.

The association of TMJ arthritis and hearing loss has only been reported in skull base osteomyelitis (malignant external otitis) (15). The extensive inflammatory lesion involving the TMJ and the periarticular soft tissues depicted by MR imaging is quite similar to the associated changes encountered in some thoracic manifestations (eg, thoracic outlet syndrome, subclavian vein obstruction) reported in severe manubriosternal joint disorders (1, 11). One explanation for the deafness experienced by our patient is that the inflammatory process of the temporal squama osteitis induced a focal aseptic inflammatory reaction of the dura that involved the internal auditory meatus, passing through the modiolus with a concomitant inflammation of the cochlea, similar to that encountered in meningogenic labyrinthitis (16). Other possibilities could be that abnormal meningeal thickening around the aperture of the cochlear aqueduct, which connects to the subarachnoid space, resulted in increased pressure in the perilymphatic spaces of the cochlear canal (17). However, in this patient, only the caudal part of the aqueduct enhanced, and the labyrinthine enhancement did not prominently involve the basal turn. Meningeal thickening along the posterior temporal bone could also have involved the endolymphatic duct and sac, resulting in hearing loss.

The nature of the enhancement of the cochlea on contrast-enhanced T1-weighted images is presumed to be reactive, as the endo- and perilymphatic fluid displayed a normal high signal on T2-weighted images. This hypothesis was confirmed in a 1-year follow-up study, in which the cochlea showed neither enhancement on MR images nor reactive ossification on CT scans. Spontaneous regression is frequently encountered in SAPHO syndrome (11). The mechanism of labyrinthine involvement is quite different from that reported in granulomatous or autoimmune labyrinthitis (15, 17), in which the cochlea is involved by a granuloma and may ossify secondarily.

This example of cochlear involvement underscores the belief that inner ear MR screening should combine T2-weighted imaging with pre- and postcontrast T1-weighted imaging to exclude labyrinthitis or pachymeningitis (19–21). Moreover, in patients with labyrinthitis and focal pachymeningitis, an inflammatory process of the skull base, including the TMJ region, should be excluded.

Conclusion

Radiologists may play a role in the diagnosis of SAPHO syndrome, as it may be virtually asymptomatic. Characteristic osteoarticular and dermatologic manifestations define this disorder. Accurate identification of SAPHO syndrome may obviate misdiagnosis, unnecessary biopsy, or antibiotic therapy.

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