Severe destruction of the temporomandibular joint with complete resorption of the condyle associated with synovitis, acne, pustulosis, hyperostosis, and osteitis syndrome

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The synovitis, acne, pustulosis, hyperostosis, and osteitis (SAPHO) syndrome consists of a combination of inflammatory bone disorders and dermatologic pathology. Bone lesions as a form of diffuse sclerosing osteomyelitis in the mandible occur in the posterior body and ramus. Bone lesions rarely spread to the temporomandibular joint (TMJ) where ankylosis may result. Herein we present an unusual case of SAPHO syndrome with TMJ involvement in which severe destruction of the TMJ occurred. We observed an extension of the invasive soft tissue lesion into the infratemporal fossa from the TMJ with complete resorption of the condyle. In contrast to other previously reported cases, in our case the condyle was strongly suspected as the primary site of the bone lesion with subsequent extension to the ramus and infratemporal fossa. The destructive nature and related symptoms resembled a malignant tumor. (Oral Surg Oral Med Oral Pathol Oral Radiol 2013;116:e128-e133)

The synovitis, acne, pustulosis, hyperostosis, and osteitis (SAPHO) syndrome belongs to the class of rheumatic diseases mediated by immunologic pathways. The term SAPHO refers to the spectrum of inflammatory bone disorders, which may or may not be associated with dermatologic pathology. Bone manifestations, such as hyperostosis intermixed with areas of osteolysis and arthritis, are the most typical findings. The most common locations of the lesions are the anterior chest wall (sternoclavicular, sternocostal, and manubrium regions), pelvic girdle, and spine.[1,2]

The mandible can be affected by inflammatory bone disorders in patients with the SAPHO syndrome. The frequency of mandibular involvement is approximately 10%,[2,3] and the most common site is the posterior body and ramus.[4] The SAPHO syndrome with TMJ involvement is rare. In 120 patients with SAPHO syndrome, 13 patients had mandibular osteomyelitis (10.8%), but TMJ involvement was reported for only 1 case.[2] Another report documented TMJ involvement in 3 of 62 patients with mandibular involvement.[3] There have been only 8 cases of TMJ involvement (including the current case) reported in the literature.[3,5-10]

Mandibular lesions are termed diffuse sclerosing osteomyelitis of the mandible (DSOM).[4] The characteristic radiologic finding in patients with DSOM is a periosteal reaction with diffuse sclerosis or intermingled sclerotic and lytic lesions, leading to a deformity of the mandible in the end stage.[11,12] When the TMJ is affected by DSOM, ankylosis develops. Ankylosis is thought to occur with mandibular osteitis of the adjacent joint. Previous reports of SAPHO syndrome with TMJ involvement, therefore, have focused on TMJ ankylosis, in particular in relation to reconstruction of the TMJ.[3,6]

Herein we present a case of SAPHO syndrome with TMJ involvement. In contrast to previous reports, severe destruction of the condyle occurred rather than TMJ ankylosis. Complete resorption of the right condyle and an extensive soft tissue mass around the right TMJ were detected with computed tomography (CT) and magnetic resonance imaging (MRI). There

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has been no other report involving severe destruction of the TMJ.

REPORT OF A CASE

A 50-year-old man was referred from the Department of Thoracic Surgery to our clinic for evaluation of uptake in the right TMJ with technetium-scintigraphy. Two years earlier abnormal chest radiographic findings were noted in a routine medical examination. As a result, whole-body scintigraphy was performed in the Department of Thoracic Surgery of our hospital, and pathologic uptake was detected in the left sternoclavicular joint, sternum, bilateral claviculars, and right TMJ (Figure 1). Based upon CT, the sternal and clavicular lesions, including the sternoclavicular joint, were first thought to have osteomyelitis of mixed types presenting with progressive bone sclerosis and scattered osteolysis. A mediastinal, soft tissue lesion was also suspected after MRI. A biopsy of the mediastinal, soft tissue mass revealed fibrotic, neoplastic tissue without a sign of malignancy.

On admission to our clinic, the patient complained of oppressive pain in his right TMJ with limited ability to open his mouth and slight spontaneous pain in his right cheek area. The patient also had tinnitus. Clinically, diffuse swelling of the right TMJ region was observed. The inter-incisal distance was 30 mm. There was a 3 mm mandibular mid-line shift to the left in occlusion (Figure 2). The C-reactive protein (CRP) level was elevated at 3.6 mg/dL. The blood count and other laboratory tests were within normal limits. The patient was able to eat but reported that 5 years before admission to our clinic he experienced (for the first time) pain while chewing involving the right TMJ and subsequently had diffuse swelling of the right side of his face and limited ability to open his mouth. At that time, arthritis of the right TMJ was diagnosed in a dental clinic, where he received antibiotics and non-steroidal anti-inflammatory drugs (NSAIDs), with symptom improvement.

A panoramic radiograph (Figure 3) and CT (Figure 4) obtained in our clinic showed complete resorption of the right condyle. A three-dimensional CT confirmed, complete destruction of the condyle (Figure 5). Osteomyelitis of the mixed type with progressive bone sclerosis and scattered osteolysis suggestive of DSOM was detected in the ramus and coronoid process. A fibrous, tissue-like, hyperplastic lesion around the right TMJ was also observed with MRI (Figure 6). Based on diagnostic criteria (Table I), the clinical and radiologic findings were consistent with the diagnosis of SAPHO syndrome.[5] No skin lesion was noted.

In this case, the treatment strategy focused on symptom management. NSAIDs (loxoprofen sodium, 120-240 mg/day) are effective for the relief of TMD

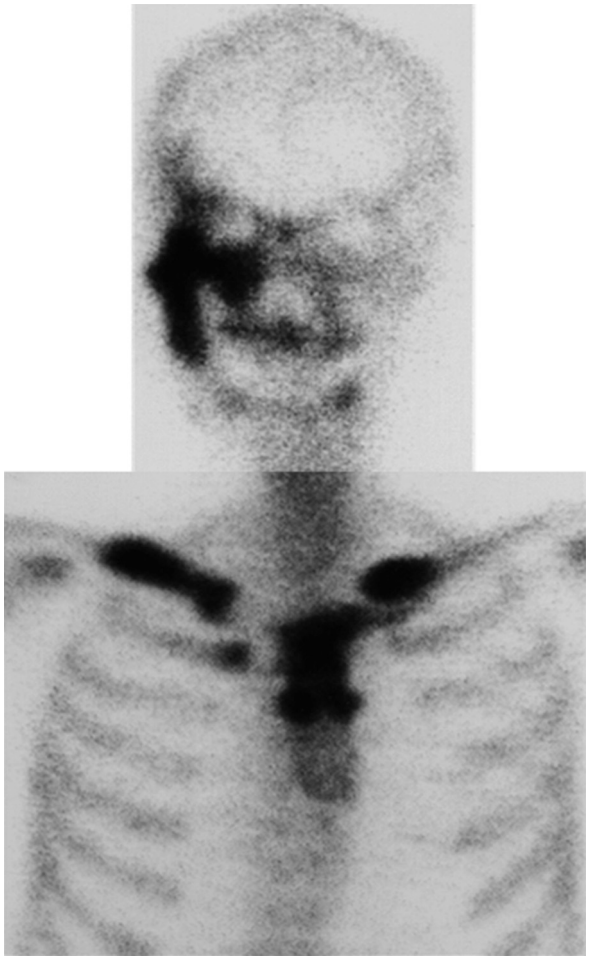

Fig. 1. Scintigraphy findings show intensive uptake of the radiopharmaceutical technetium-99m at the left sternoclavicular joint, sternum, bilateral clavicles, and right TMJ.

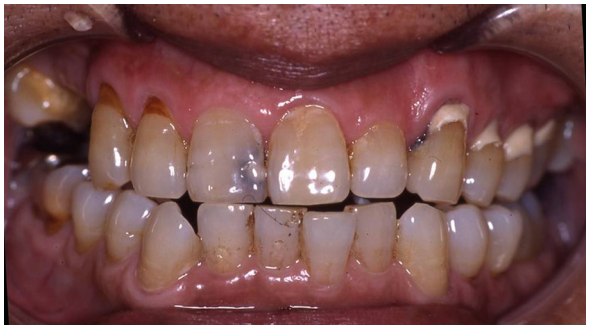

Fig. 2. Intraoral photograph on admission to our clinic shows a 3-mm, mandibular, mid-line shift to the left in occlusion.

symptoms. For the first 2 years of initial treatment, the mouth opening was maintained at approximately 25 mm with controllable pain. Thereafter, the patient spontaneously developed proptosis of the right eye and had difficulty keeping his eye closed; he also had double vision. At this point, the width of the mouth opening had