Pediatric Treatment-Resistant Nonbacterial Osteomyelitis of the Mandible Associated with SAPHO Syndrome

Yu Kamata, Tomohiro Yamada*, Tomoki Sumida, Hiroyuki Nakano, Goro Sugiyama, Azusa Nakashima and Yoshihide Mori
Section of Oral and Maxillofacial Surgery, Division of Maxillofacial Diagnostic and Surgical Sciences, Faculty of Dental Science, Kyushu University, Japan

*Corresponding Author: Tomohiro Yamada, Section of Oral and Maxillofacial Surgery, Division of Maxillofacial Diagnostic and Surgical Sciences, Faculty of Dental Science, Kyushu University, 3-1-1 Maidashi, Higashi-ku, Fukuoka, 812-8582, Japan, Tel: +81-92-642-6452, Fax: +81-92-642-6392, Email: tyamada@dent.kyushu-u.ac.jp

Abstract

SAPHO syndrome is a chronic disease of unknown etiology, which is characterized by synovitis, acne, pustulosis, hyperostosis, and osteitis, however, pediatric osteomyelitis of the jawbone is rarely reported. We present a report of pediatric nonbacterial osteomyelitis of the mandible associated with SAPHO syndrome.

A seven-year-old girl presented complaining of pain in, and swelling of, the right retromolar region. Antibiotics had no effect and her past history included linea alba, strabismus, Palmoplantar Pustulosis (PPP) and old fractures of the Thoracic Vertebrae (Th7-9). Together with histological findings, she was diagnosed as suffering from SAPHO syndrome with mandibular osteomyelitis.

NSAIDs (Naproxen), Corticosteroids (Dexamethasone), and Methotrexate (MTX) were effective for several months but the effects were transient. As the third line therapy, an anti-TNFα agent (infliximab) was administered in addition to MTX. The mandibular symptoms have subsequently been under control for over a year.

For treatment of mandible osteomyelitis with SAPHO syndrome, systemic immunosuppressive therapy should be considered, beside continuous oral management.

Keywords: SAPHO syndrome, Osteomyelitis, Mandible, TNF-α, Pediatric

Introduction

SAPHO syndrome is a chronic disease of unknown etiology, which is characterized by synovitis, acne, pustulosis, hyperostosis, and osteitis [1]. SAPHO syndrome slightly predominates in females, 70:50, and the mean age at symptom onset is 28.6 years (SD ± 13.7 years; range 4-63) [2,3]. Bone lesions are manifested in more than 90% of cases [2], mostly Diffuse Sclerosing Osteomyelitis (DSO). Pediatric osteomyelitis of the jawbone is rarely reported, and there are no evidence-based treatment protocols [4,5].

We present a report of pediatric nonbacterial osteolytic osteomyelitis of the mandible associated with SAPHO syndrome.

Case Report

Patient history

A seven-year-old girl was referred to the Department of Oral and Maxillofacial Surgery with the complaint of pain and swelling of the right retromolar region. She had noticed the right cheek swelling and pain one month earlier, and had seen a dentist. Antibiotics were prescribed, but no effects were evident. Her past history revealed that she had suffered a linea alba hernia as an infant and conservative treatment had been successful. At 6 years-old, she had an operation on the left strabismus, and was treated Palmoplantar Pustulosis (PPP) with ointment by a dermatologist. At seven years-old, she was pointed out old fractures of the Thoracic Vertebrae (Th7-9).

Physical examination and diagnosis

Her general growth was normal but she had diffuse swelling of the right cheek and retromolar region (Figure 1). Trismus was not obvious. A blood examination showed no inflammatory signs, including elevated value of C-reactive protein and white blood cells count. An orthopantomogram and CT scan (Aquillion 64, Toshiba; reconstruction matrix=0.35mmx0.35mmx0.5mm) revealed a diffuse osteolytic lesion and periosteal

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subsequently flared up. Corticosteroids had only limited effect and methotrexate (MTX) was administered as the second line therapy. The perimandibular symptoms were initially alleviated, but flared up again after two months. As the third line therapy, an anti-TNFα agent, infliximab was administered in addition to MTX. The symptoms have subsequently been under control for over a year (Figure 5).

Discussion

SAPHO (synovitis, acne, pustulosis, hyperostosis, and osteitis) syndrome was first reported by Chamot in 1987 [1]. It occurs in less than 1 in 10,000 of the population, however, the mandible may be involved (Table 1) so it is important for dentists to be aware also. The jawbone is occasionally involved (11%) [2], and affected with osteomyelitis as a part of Chronic Recurrent Multifocal Osteomyelitis (CRMO) [6]. SAPHO syndrome is mostly diagnosed at a younger age (mean; 28.6 years) [2], however, osteomyelitis of the jawbone usually occurs at a later age [7-12] (mean; 44.3 years, Table 1). This may be because of late onset in the jawbone, or difficulty of diagnosis in the early stage, where it might be misdiagnosed as a simple dental infection.

Manifestation of SAPHO syndrome in the mandible is mostly Diffuse Sclerosing Osteomyelitis (DSO), while 42% of DSO in the mandible is reportedly SAPHO syndrome. Suei, et al. [13] reported that osteolytic and periosteum reactions mainly occurred at an early stage, and changed to DSO after repeated reactions [12]. So, it is desirable to detect and treat in earlier stage. Our case also showed an osteolytic lesion with pain and swelling, and the differentiated diagnosis was dental infectious osteomyelitis, neoplastic lesion (LCH), or aseptic osteomyelitis. The biopsy revealed no neoplastic lesion but osteomyelitis confirmed. Bacterial examination was negative and antibiotics had no effect.
Together with dermatological and thoracic vertebrae lesions, she was diagnosed with aseptic osteomyelitis of the mandible with SAPHO syndrome.

The etiology of SAPHO syndrome is thought to have infectious, genetic, and immunological causes. It may relate to with HLA-B27 [14], however, there are few positive cases, and HLA is not yet in the diagnostic criteria [15]. Wannfors reported that identification of infectious loci and their removal are important, because the initial phase of the lesion is a bacterial infection [9].

Reported treatments for mandibular osteomyelitis of SAPHO syndrome are as surgery, NSAIDs, corticosteroids, Methotrexate (MTX), and anti-TNF agents [10,11]. Conservative surgery such as decortication, alone tends to cause recurrent inflammation [8]. So, radical surgeries with microsurgical reconstruction [3] or

Table 1: Mandible involvement in SAPHO syndrome.
TMJ replacement [7] are sometimes unavoidable. However, some authors caution that radical surgery should be avoided because of the natural course of this systemic disease [3]. In our case, despite NSAIDs, corticosteroids, and MTX showing limited effects, an anti-TNFα agent (infliximab) eventually produced a favorable response. The symptoms of SAPHO syndrome may sometimes be self-limiting [16], but is usually a treatment-resistant disease. Early detection of this disease and inflammation control in childhood would be expected to avoid the development of DSO and unnecessary radical surgery.

Conclusions

For treatment of pediatric mandible osteomyelitis with SAPHO syndrome, it is important that systemic immunosuppressive therapy under the care of a pediatrician is conducted. Furthermore, oral management should be maintained to minimize or remove any dental infectious loci.

References


