

CASE REPORT

Osteonecrosis and ankylosis of temporomandibular joints (TMJ) in juvenile onset systemic lupus erythematosus (JSLE).

Cuttica RJ, Marcantoni MB, Laham M.
Hospital de Pediatría Pedro de Elizalde
Rheumatology Unit and Maxilofacial Surgery Unit
Buenos Aires - Argentina

Contact:

Rubén J. Cuttica, MD Head Rheumatology Unit
Hospital de Pediatría Pedro de Elizalde Av. Montes de Oca 40 1270
Buenos Aires Argentina
Tel/Fax: (+5411)4361-0900
e-mail: elizalde_reumato@buenosaires.gov.ar

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Abstract:

Osteonecrosis is a well known complication in JSLE but involvement of TMJ has been rarely reported. We report an 11 year old girl diagnosed as JSLE with chronic steroid treatment that developed osteonecrosis of both mandibular condyles with ankylosis of TMJ with only a 3 mm interdental distance. The patient underwent surgery with coronoidectomy and resection of the ankylosed bone, creating a new glenoid fossa without adding any material in the space. An extraoral retractor was placed with the purpose of keeping separated the new surfaces and lengthening soft tissue preventing reankylosis. The child had an excellent outcome with a post-operative interdental distance of 30 mm.

Introduction:

Osteonecrosis is a well known complication in systemic lupus erythematosus. It is reported in about 10 to 15% of patients with the femoral head being the most frequently involved bone. [1-3] The main risk factor for osteonecrosis is corticosteroid treatment at doses greater than 10 mg/day for 6 or more months. Other risk factors include leucopenia, Raynaud's phenomenon, vasculitis, and antiphospholipid syndrome. [2] In our knowledge, there are only a few case reports in the English literature of osteonecrosis of the TMJ, particularly in children. [4-6] The objective of this paper is to describe a patient with juvenile onset SLE who developed osteonecrosis and ankylosis of bilateral mandibular condyles with severe functional impairment that required surgical treatment.

Case report:

An 8 year 10 month girl was diagnosed to have JSLE in 2002. She presented with prolonged fever, a malar rash, diffuse alopecia, polyarthralgias, leucopenia, pleuritis, pericarditis, a positive ANA with a homogeneous pattern, a positive DNA and a low C3 and C4. Since diagnosis, she was treated with prednisone at doses greater than 1mg/Kg/day for two years. She also received cyclophosphamide, first oral and then pulse therapy due to renal involvement.

When this child was seen in our center for the first time in 2004, she was noted to have severe cushinoid features, generalized muscle hypotrophy, a malar rash, diffuse alopecia, arthralgias and dyspnea even at rest. She also had bilateral cataracts and right ear deafness. No arthritis, organomegaly, lymphadenopathy, Raynaud's phenomenon, CNS problems, or vasculitic rash were seen. She had a positive ANA at a titer of 1/1280 (homogeneous pattern), an anti-double-stranded DNA titer of 1/80, a negative rheumatoid factor, and normal complements and normal renal function. A chest radiograph showed an elevated and poorly mobile diaphragm. Pulmonary functional tests demonstrated a severe restrictive pattern and a high resolution lung CT scan showed bilateral basal fibrosis. A muscle strength evaluation detected an impairment of the diaphragm and accessory muscles. Cardiac evaluation revealed an electrocardiogram-impressive ST segment changes compatible with a myocardial involvement. A two dimensional and Doppler echocardiogram revealed a hypokinetic interventricular septum without pericardial effusion. After this overall assessment, her SLEDAI score was noted to be 12 with a CHAQ score of 1.

The child was continued on the monthly cyclophosphamide pulse therapy, hydroxychloroquine was added, and tapering of steroid dose was started. Physical therapy was begun. She was then lost to follow-up for the next eight months. She returned in February 2005 complaining of 6 months of pain upon chewing that was referred to ears as well as clicking on mandibular movements. At examination, she had severe limitation of mouth opening with 3 mm interdental distance (Fig. 1). Examination revealed mild polyarthritis. Laboratory testing demonstrated improvement of lung functional tests with a normal CO diffusion. Her Sm was 20 U, and the RNP, Ro, and La antibodies were negative. Densitometry showed -2 SD from the Z score (Osteopenia). The SLEDAI score had decreased to 4 and the CHAQ to 0.15. She had begun to grow again.



Fig. 1. The child's maximal mouth opening in February 2005

Fig. 2 a, b, c. Three views of the child's temporomandibular joints showing flattening of both mandibular condyles, loss of meniscal discs, and absence of joint space.



Fig. 2 a Orthopantomography (Panorex)



Fig. 2b 3D CT scan of the TMJ



Fig 2 c MRI of the affected TMJ's

The patient next underwent surgery on both temporomandibular joints. After nasotracheal intubation for general anaesthesia, preauricular boarding was performed. After the exposure of the mass of the ankylosed joint, the mass was nearly completely removed creating a new glenoid fossa, without inserting any material in the new space. Next a coronoidectomy was performed with temporalis muscle dissection. Finally, after obtaining an oral opening of 30 mm, a Molina extraoral distractor was fixed to the zygomatic area and the lateral mandibular ramus. The purpose of this retractor was to prevent reankylosis by keeping the new surfaces well-separated as well as lengthening the soft tissue (Fig. 3 a-b).



Fig. 3 a



Fig. 3 b

Figure 3a Surgical procedure with removal of the ankylosed joint

Figure 3b Extraoral retractors

The child's mother was trained in the management of the retractors and a rehabilitation program was started. The external distractors were removed 45 days post-operatively. Figure 4 demonstrates the excellent post-operative result with an interdental distance of 30 mm (Fig. 4).



Fig. 4. Mouth opening after removal of distractors and rehabilitation with a new interdental distance of 30

mm.

Discussion

In about 5 to 10% of SLE adult patients, clinically symptomatic osteonecrosis with radiographic findings has been reported. When asymptomatic patients studied with magnetic resonance imaging were included, this prevalence increased to 35%. The femoral head was the most frequently involved bone. Involvement of the TMJ has been well recognized in juvenile idiopathic arthritis (JIA). In this case, it was our judgement that the TMJ damage was much more likely due to osteonecrosis than JIA-like disease due to: 1) the short period of time between TMJ involvement symptoms and the radiological findings; 2) the fact that the child did not have chronic polyarthritis or the radiographic changes in the TMJ or other joints typical of overlap syndromes with a JIA-like course. This osteonecrosis of the TMJ is much less frequently noted in JSLE patients than adult SLE patients and osteonecrosis of the mandibular condyles has been rarely reported in these children.

As growth of lower maxillary bone depends on the growth plate located on the condyle, the involvement of TMJ in children has a risk of micrognathia, dental malocclusion, eating difficulties [7], and problems with intubation before general anesthesia. [8] In our patient, the symptoms of TMJ involvement were pain on chewing that was referred to her ears as well as clicking on movement of her jaw. These clinical findings in a child with JSLE who has a history of a chronic high dose corticosteroid treatment for more than two years should suggest to a clinician the possibility of condyle osteonecrosis, particularly as steroid treatment is considered the most important risk factor for bone mass loss. [9-10] Other risk factors for osteonecrosis must be kept in mind, including leucopenia, Raynaud's phenomenon, vasculitis, and antiphospholipid syndrome. [2]

In this patient, if the child had had a dental and orthodontic consult at the time the symptoms started, some steps to protect the joint such as an oral myorelaxing splint and orthodontic therapy could have been used to enhance condyle remodeling and mandibular reposition. [5-8, 11] Imaging studies that allow serial reevaluation of the bones, meniscal discs, soft tissue structures and the functional relationship between them are very valuable. The most useful method to evaluate TMJ is the MRI followed closely by 3D CT scan. [12] In our patient, the severity of the lesions on imaging studies and severe impairment of mouth opening led to the decision for immediate surgical treatment with excellent results.

It is concerning that even when our patient was severely handicapped with opening her mouth and eating, her CHAQ score did not reflect this decreased functional ability. In reviewing other scales to measure functional ability and quality of life in children with chronic illness, none of them evaluates TMJ and a fundamental aspect of daily living activities such as eating. This omission should be addressed. In the meantime, it is important to ask about TMJ problems in JSLE children even if the patient has had no prior signs or symptoms. With greater awareness of this potential problem, early detection may increase and help minimize TMJ damage in JSLE children.

In severe cases, the options for surgical treatment might include: a) prosthetic joint replacement, b) replacement with homologous bone with costochondral ossification nucleus and c) transient bone retractor [13-21]. The problem with the first option is that small size prostheses are not often available as well as

the risk of prosthesis failure. Regarding the second option, there is a risk of further osteonecrosis that requires a new surgical procedure. The third option was chosen because it is easy to perform and the bone separation produced by the retractors permits reestablishment of a joint space between the zygomatic arch and the lateral mandibular ramus. Both parts of the joint are kept separated by the distractors in order that after some time the bone structures remodel and soft tissue elongate resulting in a new joint space without an Intraarticular disc. This procedure must be followed by an early and intensive rehabilitation program for the TMJ in order to avoid reankylosis.

Summary

Osteonecrosis of the TMJ is an unusual complication in juvenile SLE and appears to be associated with chronic corticosteroid use so common in the treatment of JSLE. As the functional scales often utilized miss TMJ issues in these JSLE patients, pediatric rheumatologists should consider routinely asking about TMJ problems and checking the oral opening just as in JIA patients. Early detection of osteonecrosis may lead to less TMJ damage and need for surgery. In cases of ankylosis of the TMJ, arthroplasty with the use of TMJ distraction and rehabilitation may be the best alternative.

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