Case Report

Isolated Temporomandibular Synovitis as Unique Presentation of Juvenile Idiopathic Arthritis

GIORGIA MARTINI, UGO BACCILIERO, ALBERTO TREGNAGHI, MARIA CRISTINA MONTESCO, and FRANCESCO ZULIAN

ABSTRACT. Temporomandibular joint (TMJ) involvement is quite frequent in juvenile idiopathic arthritis (JIA). We describe a 15-year-old girl with isolated TMJ arthritis presenting as a unique manifestation of JIA, and its successful treatment. She underwent arthroscopic synovectomy followed by intraarticular steroid injection. Early use of synovectomy and intraarticular steroids in TMJ arthritis may reduce pain, improve jaw function, and prevent irreversible deformities. (J Rheumatol 2001; 28:1689–92)

Key Indexing Terms:
TEMPOROMANDIBULAR JOINT JUVENILE IDIOPATHIC ARTHRITIS

Temporomandibular joint (TMJ) involvement is quite frequent in juvenile idiopathic arthritis (JIA), particularly in polyarticular and systemic forms, with prevalence varying from 22% to 72%[1-5], but has never been reported as a unique manifestation of JIA.

We describe a case of isolated TMJ arthritis that was the unique manifestation of JIA and outline a successful treatment approach.

CASE REPORT

A 15-year-old girl was referred to our Pediatric Rheumatology Unit for further evaluation of orofacial pain. One year prior to our evaluation she had experienced orofacial pain during mastication and mandibular movement. At first these symptoms had been interpreted as myofascial pain secondary to malocclusion, and a nightly occlusal appliance was then utilized without any improvement. Two months later, an orthopantomograph showed no evidence of dental abnormalities, but flattening and reduced range of motion of the mandibular condyles.

Over the next months, the girl experienced increased pain intensity with difficulty in eating and mastication. Eight months later, computed tomography (CT) scan of the TMJ confirmed the bilateral erosions of the mandibular condyles with flattening of their surface. Bone scintigraphy (^{99}Tc) revealed an isolated increased tracer uptake on both mandibular condyles that was confirmed by SPECT scan; this allowed us to rule out possible artifacts (Figure 1A).

On admission, she complained of severe facial pain on mastication with difficulty eating and weight loss (5 kg in 6 mo). Her history was unremarkable. In her family history, a paternal uncle had rheumatoid arthritis.

On examination she was a very thin girl (weight 39.1 kg, less than the 3rd centile for her age), with poor subcutaneous fat distribution. She had decreased mandibular range of motion with maximal mouth opening capacity of 30 mm, bilateral crepitations, malocclusion with decreased mandible protrusion, and severe weakness of the mastication muscles. The remaining physical examination was unremarkable.

Complete blood count, erythrocyte sedimentation rate, C-reactive protein, serum electrolytes, thyroid, liver and kidney function tests were all normal. Tests for antinuclear antibody, dsDNA, rheumatoid factor were all negative, as well as testing for HLA-B27, her haplotype being A2, A33, B39, B65, DR1, DR8, DQ1, DQ4. Ophthalmologic examination revealed no sign of inflammation in the anterior chamber.

Gadolinium enhanced magnetic resonance imaging (Gd-DTPA MRI) confirmed severe deformities and decreased range of motion of the mandibular condyles, synovial fluid, and disc flattening and fragmentation. After Gd-DTPA injection, a clear enhancement of the synovial membrane suggested extensive bilateral synovial hyperplasia (Figure 2). She underwent bilateral arthroscopic synovectomy, and the pathology showed multilayered synovial lining composed of hyperplastic and hypertrophic synoviocytes diffusely infiltrated by lymphocytes and plasma cells.

The history, the clinical pattern, the radiological and histological findings were strongly suggestive of JIA diagnosis, oligoarticular type. She was given naproxen and, one month later, had intraarticular injection of triamcinolone hexacetonide 10 mg in both temporomandibular joints.

Within 6 months after treatment, a dramatic, clinical improvement was observed: she no longer experienced pain on mastication, and had started eating properly with a subsequent weight gain of 4.7 kg. On examination the mandibular range of motion had greatly improved, with opening distance of 42 mm, no crepitations, and normal strength of the mastication muscles. The examination of the other joints and of the anterior eye chamber was still normal.

At 12 month followup, 24 months after onset, no other joint involvement was evident. Bone scintiscan (^{99}Tc) was repeated and showed complete resolution of the active lesions previously highlighted in the TM

From the Department of Pediatrics, Radiology Institute, and Pathology Institute, University of Padua, Padua; and the Maxillofacial Surgery Division, Hospital of Vicenza, Vincenza Italy.

G. Martini, MD, Clinical Fellow, Rheumatology Unit, Department of Pediatrics, University of Padua; U. Bacciliero, MD, Assistant Professor, Maxillofacial Surgery Division, Hospital of Vicenza;

A. Tregnaghi, MD, Assistant Professor, Radiology Unit; M.C. Montesco, MD, Assistant Professor, Pathology Institute; F. Zulian, MD, Assistant Professor, Rheumatology Unit, Deparment of Pediatrics, University of Padua.

Address reprint requests to Dr. F. Zulian, Dipartimento di Pediatria, Università di Padova, Via Giustiniani 3, 35128 Padova, Italy.
E-mail: zulian@child.pedi.unipd.it

Submitted September 26, 2000; revision accepted January 29, 2001.

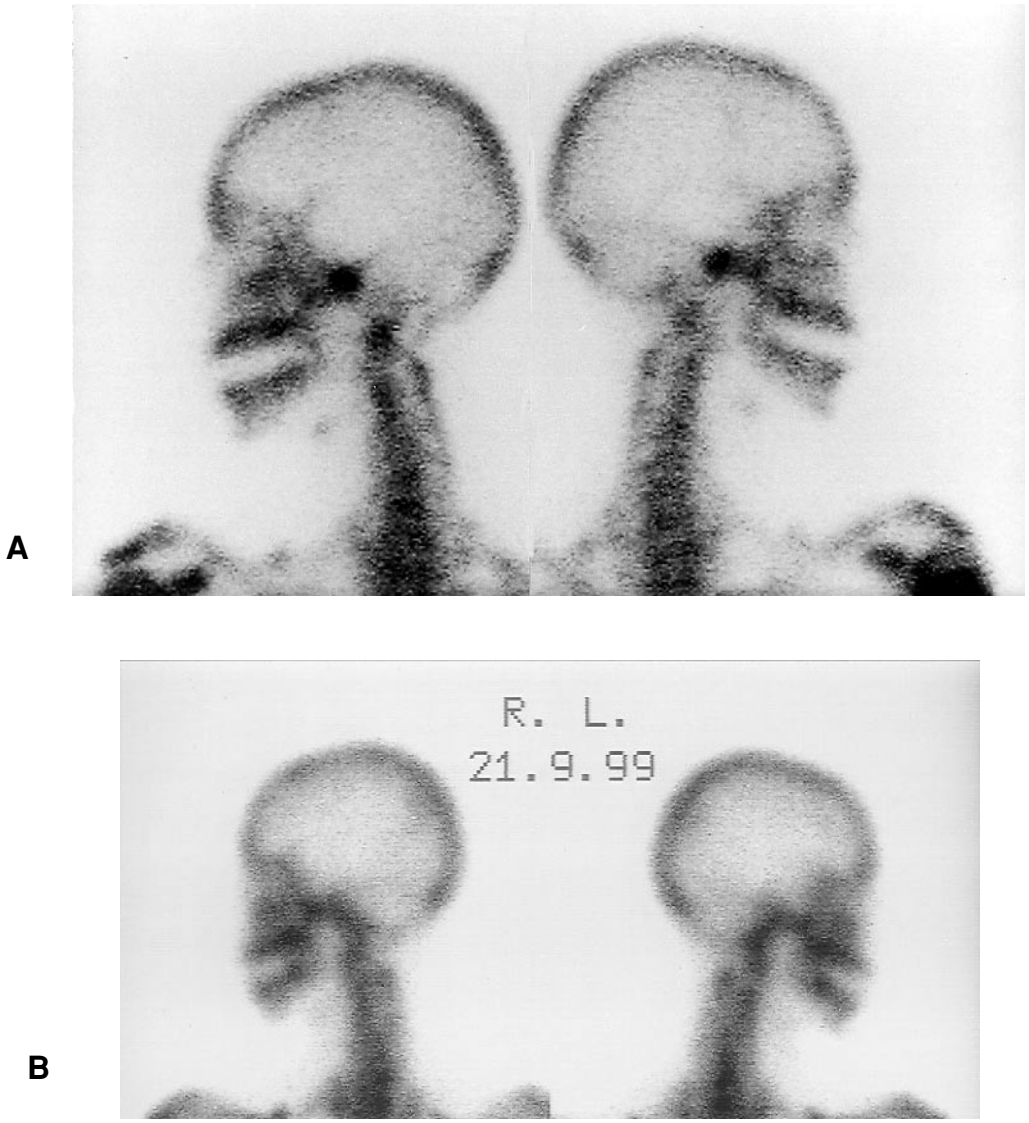

Figure 1. A. Radiophosphate (Tc-99m MDP) images show intense isolated tracer uptake of both TMJ. B. Radiophosphate (Tc-99m MDP) images 6 months after treatment showing complete normalization of TMJ.

joints (Figure 1B). One year later we repeated Gd-DTPA MRI, which revealed the persistence of unchanged condyle deformities and limited condyle range of motion on mouth opening, but disappearance of intraarticular effusion. After gadolinium injection the synovial thickening observed in the previous MRI was no longer present.

DISCUSSION

Radiographic TMJ abnormalities have been reported in 22–72% of patients with JIA[1-5]. TMJ involvement occurs more frequently in polyarticular onset than in either oligoarticular or systemic onset JIA[2-5].

No marked sex difference has been found either in prevalence or in severity of TMJ arthritis[1,2]. While some clinical variables, such as duration and early onset of the disease, are significantly associated with TMJ involvement, immunological tests (rheumatoid factor, antinuclear antibody, HLA-B27) are not[2,4,6].

Our patient's TMJ arthropathy represents an isolated manifestation of JIA; in fact, 24 months after onset of symptoms, the TMJ arthritis was still the only localization of the disease. The recently revised criteria for JIA, oligoarticular type, are fulfilled by exclusion of infective, hematological, and malignant conditions[7].

The typical symptoms of TMJ synovitis such as pain during jaw movement, crepitus, and restricted mouth opening capacity[2,8] were all present in our patient, as well as the radiological findings such as bilateral condylar flattening and erosions on CT scan, which have been found to be strongly related to TMJ involvement in JIA[9]. MRI, showing inflammation and thickening of the synovial membrane, confirmed this diagnosis. MRI has been shown to be a helpful diagnostic tool in the detection of early inflammatory changes in TMJ, with a strong correlation

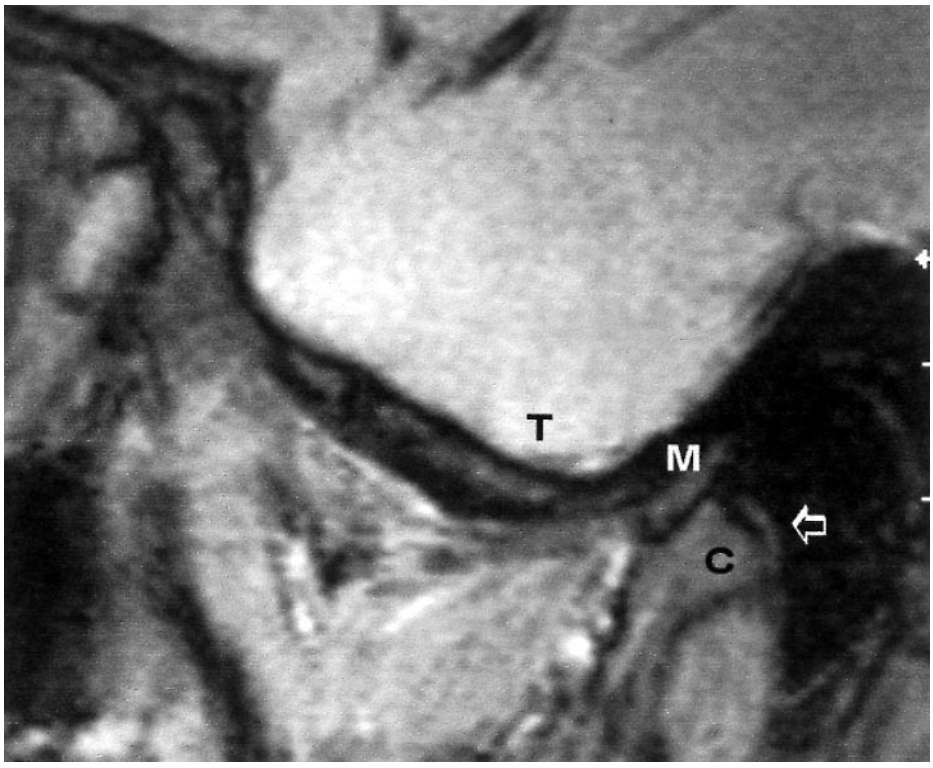

Figure 2. Gradient echo sequence MRI of the right TMJ (mouth open). Note thickening of synovial membrane (arrow), irregular profile of the flattened condyle, and narrowed articular space (M: fragmented meniscus, T: articular tubercle, C: mandibular condyle).

between degree of enhancement and degree of pathological findings in the synovial membrane[10,11]. The histology report, showing villous hyperplasia of the synovial membrane with inflammatory infiltrate of lymphocytes and plasma cells, gave final confirmation of the diagnosis.

Chronic synovitis of the TMJ can have early and late consequences: the earliest is the functional impairment due to synovial membrane thickening, masticator muscle weakness, and facial pain. Moreover, the limited mandibular movement makes airway maintenance very difficult in case of major procedures such as surgical operations, and this problem, in some cases, has led to lifesaving tracheotomy[12,13]. The most important late consequence of TMJ involvement in JIA is the mandible growth disturbance that leads to either micrognathia or to facial asymmetry if bilateral or unilateral, respectively. In our patient the reduced mouth opening capacity was severe and limited eating with a consequently important weight loss.

Considering the severity of the clinical picture and the unusual isolated localization we elected to perform bilateral arthroscopic synovectomy followed by intraarticular injection of steroids to improve symptoms quickly and to obtain histological confirmation of clinical and radiological suspicion of TMJ chronic synovitis. In a recent report the combination of arthroscopic synovectomy followed shortly by intraarticular steroid injection was found to be more effective than the same procedures alone in gaining long lasting reduction of local inflammation in unresponsive active knee synovitis[14].

Triamcinolone hexacetonide is one of the most widely used steroids for intraarticular injection, with a satisfactory rate of response (81% remission after 6 months)[15,16]. The dose of 1 mg/kg has been shown to be appropriate and safe for the articular cartilage.

The TM joint's delicate structure, with the mandibular growth plate just under the fibrocartilage of the condylar head, and therefore easily damaged in active synovitis, justifies an aggressive therapeutic approach to reduce the risk of severe growth impairment.

The dramatic response in our patient is very encouraging and indicates that intraarticular steroid injection in combination with arthroscopic synovectomy in the management of early TMJ involvement alleviates pain, improves jaw function, and prevents further joint damage that can lead to irreversible deformities.

REFERENCES

1. Pearson MH. Lesions of the mandibular condyle in juvenile chronic arthritis. Br J Orthodont 1996;23:49-56.
2. Larheim TA, Hoyeraal HM, Stabrun AE, Haanes HR. The temporomandibular joint in juvenile rheumatoid arthritis. Scand

J Rheumatol 1982;11:5-12.
3. Barriga B, Lewis TM, Law DB. An investigation of the dental occlusion in children with juvenile rheumatoid arthritis. Angle Orthod 1974;44:329-75.
4. Kurhulahti T, Ylijoki H, Ronning O. Mandibular condyle lesions related to age at onset and subtypes of juvenile rheumatoid arthritis in 15-year-old children. Scand J Dent Res 1993;101:332-8.
5. Mericle PM, Wilson VK, Moore TL, et al. Effects of polyarticular and oligoarticular onset juvenile rheumatoid arthritis on facial and mandibular growth. J Rheumatol 1996;23:159-65.
6. Stabrun AE, Larheim TA, Hoyeraal HM, Rosler M. Reduced mandibular dimensions and asymmetry in juvenile rheumatoid arthritis. Pathogenetic factors. Arthritis Rheum 1988;31:602-11.
7. Petty RE, Southwood TR, Baum J, et al. Revision of the proposed classification criteria for juvenile idiopathic arthritis: Durban 1997. J Rheumatol 1998;25:1991-4.
8. Larheim TA, Haanes HR, Ruud AF. Mandibular growth, temporomandibular joint changes and dental occlusion in juvenile rheumatoid arthritis. A 17-year follow-up study. Scand J Rheumatol 1981;10:225-33.
9. Hu YS, Schneiderman ED. The temporomandibular joint in juvenile rheumatoid arthritis: I. Computed tomographic findings. Ped Dent 1995;17:46-53.
10. Küseler A, Pedersen TK, Herlin T, Gelineck J. Contrast enhanced magnetic resonance imaging as a method to diagnose early inflammatory changes in the temporomandibular joint in children with juvenile rheumatoid arthritis. J Rheumatol 1998;25:1406-12.
11. Tamai K, Yamamoto M, Yamaguchi T, Ohno W. Dynamic magnetic resonance imaging for the evaluation of synovitis in patients with rheumatoid arthritis. Arthritis Rheum 1994;37:1151-7.
12. Smith B. Anaesthesia in paediatric rheumatology. In: Woo P, White PH, Ansell BM, editors. Paediatric rheumatology update. Oxford: Oxford University Press; 1990:124-30.
13. Seymour RL, Crouse VL, Irby WB. Temporomandibular ankylosis secondary to rheumatoid arthritis. Report of a case. Oral Surg 1975;40:584-9.
14. Gattinara M, Vigano R, Gerloni V, et al. Intraarticular steroid injection after arthroscopic synovectomy enhances the efficacy of the local treatment in active knee synovitis [abstract]. Arthritis Rheum 1998;41 Suppl:S45.
15. Padeh S, Passwell JH. Intraarticular corticosteroids injection in the management of children with chronic arthritis. Arthritis Rheum 1998;41:1210-4.
16. Allen RC, Gross KR, Laxer RM, Malleson PN, Beauchamp RD, Petty RE. Intraarticular triamcinolone hexacetonide in the management of chronic arthritis in children. Arthritis Rheum 1986;29:997-1001.