Isolated Arthritis of the Temporomandibular Joint as the Initial Manifestation of Juvenile Idiopathic Arthritis

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ABSTRACT. ***Objective.*** To describe characteristics of patients with juvenile idiopathic arthritis (JIA) presenting with isolated arthritis of the temporomandibular joints (TMJ).

Methods. Patients with JIA with isolated TMJ arthritis from 4 large tertiary pediatric rheumatology centers were included. Demographic and clinical data were analyzed using descriptive statistics.

Results. Fifty-five patients were identified (65% bilateral presentation). Six patients developed arthritis in other joints (median time 6 mos); 4 patients developed uveitis, all prior to arthritis. At last followup, 9% were still taking antirheumatic medications.

Conclusion. JIA TMJ arthritis can occur in isolation, and is probably underdiagnosed. Care providers including dentists and orthodontists should be aware of this presentation. (First Release September 1 2017; J Rheumatol 2017;44:1632–5; doi:10.3899/jrheum.170263)

Key Indexing Terms:
JUVENILE IDIOPATHIC ARTHRITIS TEMPOROMANDIBULAR JOINT

Juvenile idiopathic arthritis (JIA) is the most common chronic joint disease in childhood. It affects up to 150 in 100,000 children worldwide[1]. The involvement of the temporomandibular joint (TMJ) in pediatric arthritis has been increasingly described in the last decade[2,3,4]. Multiple studies have shown TMJ arthritis being present in a large proportion of patients with JIA, with frequencies depending on the method of examination used and the JIA subtypes studied[5,6].

TMJ arthritis has traditionally been described in the context of an already established diagnosis of JIA, defined by the presence of arthritis in other joints. To date, the published literature on isolated arthritis of the TMJ consists of 2 case reports[7,8]. It seems unlikely that isolated cases of TMJ arthritis are such rare events.

The aim of our study was to determine characteristics of patients with JIA presenting with isolated TMJ arthritis in a multinational cohort of pediatric rheumatology centers.

MATERIALS AND METHODS

The databases of 4 tertiary care centers for pediatric rheumatology were searched for all patients with a diagnosis of JIA and isolated arthritis of the TMJ. Patients with other TMJ conditions or arthritis in any other joint determined by clinical examination and/or imaging findings prior to diagnosis of TMJ arthritis were excluded. A retrospective chart survey was used to extract demographic, clinical, laboratory, imaging, and therapeutic data (Table 1). Data on time to initial symptoms and orthodontic treatment were available for 14 and 38 patients, respectively. Data were analyzed using descriptive statistics, and for comparison of patients with and without involvement of other joints; the chi-square test and Student t test were used. The study was approved by the ethics committee of the medical faculty at the Ludwig-Maximilian University, Munich, Germany (Reference no. 730-15) and the institutional review boards of the respective institutions.

RESULTS

In total, 55 patients with isolated TMJ arthritis at disease onset were identified [89% female, 33% (17/52) antinuclear antibody (ANA)-positive]; of these, 5 were followed in Germany, 16 in Denmark, 4 in the United States, and 30 in Canada. Their mean age at diagnosis was 13.4 ± 2.8 yrs (range 6.0–18.2 yrs; Table 1). Diagnosis was made based on typical findings in various imaging modalities, including synovitis on gadolinium-enhanced magnetic resonance imaging (MRI) in 44/55 patients (80%), and condylar deformations by panoramic radiograph in 20/55 patients (36%) or cone-beam computed tomography (CT) in 18/55 patients

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Accepted for publication June 12, 2017.

Table 1. Demographic and clinical data of the patient cohort. Data are n (%) except where otherwise indicated.

	Cohort, n = 55
Demographics	
Female : Male (% female)	49:6 (89.1)
Referring physician	
Primary care provider/pediatrician	16 (29.1)
Dentist/orthodontist	33 (60.0)
Other specialty	6 (10.9)
Mean age in yrs at diagnosis of TMJ arthritis (± SD)	13.44 ± 2.76
Affected side	
Left	11 (20.0)
Right	8 (14.5)
Both	36 (65.5)
Disease characteristics	
Subtype according to ILAR	
Oligoarthritis	53 (96.4)
Psoriatic arthritis	2 (3.6)
Positive antinuclear antibodies	17/52 (32.7)
Positive HLA-B27	4/34 (11.8)
Positive rheumatoid factor	0/50 (0)
Other affected joints/eyes	
Patients with involvement of other joints	6 (10.9)
Time to involvement of other joints, median, range, mos	6.0 (0.5–17.2)
Patients with uveitis	4 (7.3)
Time to develop arthritis post uveitis diagnosis, median, range, mos	28.9 (9.5–51.4)
Treatment	
Nonsteroidal antiinflammatory medications	22 (40)
Methotrexate and/or leflunomide	18 (33)
Biologic medications	5 (9)
Intraarticular TMJ steroid joint injections	19 (35)

TMJ: temporomandibular joint; ILAR: International League of Associations for Rheumatology.

(33%; Figure 1). Thirty-six patients (65%) presented with bilateral TMJ involvement.

Referring physicians. Median time between initial symptoms and diagnosis of TMJ disease was 24 months (range 0–56 mos). The referring physician to pediatric rheumatology was the pediatrician or primary care provider in 16 patients (29%), a dentist or orthodontist in 33 patients (60%), and physicians of other specialties in 6 patients (11%).

Other joint manifestations and uveitis. Six patients (11%) developed arthritis in additional joints after a median of 6 months (range 0.5–17.2 mos). Compared to patients with persistent isolated TMJ arthritis, patients who subsequently developed arthritis in other joints were of similar age at onset (13.6 ± 2.6 yrs vs 12.4 ± 3.7 yrs; p = 0.47) and were similar in ANA frequency (2/6 vs 15/49; p = 0.823), but had a significantly longer disease duration (47.4 ± 7.6 mos vs 29.3 ± 25.4 mos; p = 0.001).

Four patients (7%) were diagnosed with uveitis, all prior to diagnosis of TMJ arthritis [median time to diagnosis of TMJ arthritis: 28.9 mos (range 9.5-51.4)]; none developed arthritis in additional joints. ANA was present in 3/4 (75%) of patients. All patients were treated with topical corticosteroids prior to TMJ arthritis diagnosis. Three patients also received disease-modifying antirheumatic drugs including methotrexate and azathioprine; of these patients, 1 was treated with tumor necrosis factor inhibitors prior to diagnosis and 2 at time of diagnosis of TMJ arthritis. No isolated patients with TMJ arthritis developed uveitis during followup. Two of the patients with uveitis were referred to rheumatology by an ophthalmologist.

Orthopedic/orthodontic and antirheumatic treatment. Treatment of isolated TMJ arthritis was variable. There were 18/38 patients (47%) who received orthopedic/orthodontic treatment with a distraction splint, while 11/38 patients (29%) received other orthodontic treatments, including braces and orthognathic surgery. Medications used are listed in Table 1. Twenty-one patients (38%) did not receive any antirheumatic treatment.

Disease course and outcome. After diagnosis of isolated TMJ arthritis, 47/55 patients (85%) were followed for longer than 1 month. Median followup time in these patients was 26 months (range 1–84 mos). At last followup, 4/47 patients (9%) were taking antirheumatic medications, 12/47 patients (22%) had clinical signs of TMJ arthritis, and 4 additional patients had gadolinium-enhanced MRI demonstrating activity.

DISCUSSION

Our study describes the largest cohort of patients, to our knowledge, with isolated TMJ arthritis, which in most cases was the sole manifestation of JIA. Although TMJ arthritis is a frequent feature of JIA[3], it is rarely the initial symptomatic manifestation of the disease[6,9]. About one-tenth of patients in this cohort developed clinical evidence of arthritis in other joints during the study period. The only difference in these patients was a longer disease duration, similar to an increased risk of more joint involvement with increased disease duration in JIA independent of the TMJ[3,5]. In our study, age at diagnosis or presence of ANA was comparable for all patients irrespective of involvement of additional joints. Over one-third of patients did not receive antirheumatic treatment; imaging studies demonstrated findings consistent with chronic but not active arthritis.

Only 2 reports in the literature describe cases of isolated TMJ arthritis. The first describes a 15-year-old girl who presented with bilateral erosions of the mandibular condyles, developing within 8 months[7]. She was treated successfully with arthroscopic synovectomy and intraarticular steroid injections. The second report presents a 5-year-old girl with severe bilateral TMJ arthritis with effusion, as well as cortical erosions of both the condyle and the glenoid fossa[8]. The patient improved with arthrocentesis. Neither patient developed arthritis in additional joints during followup. These previous reports appear to suggest that isolated TMJ presen-

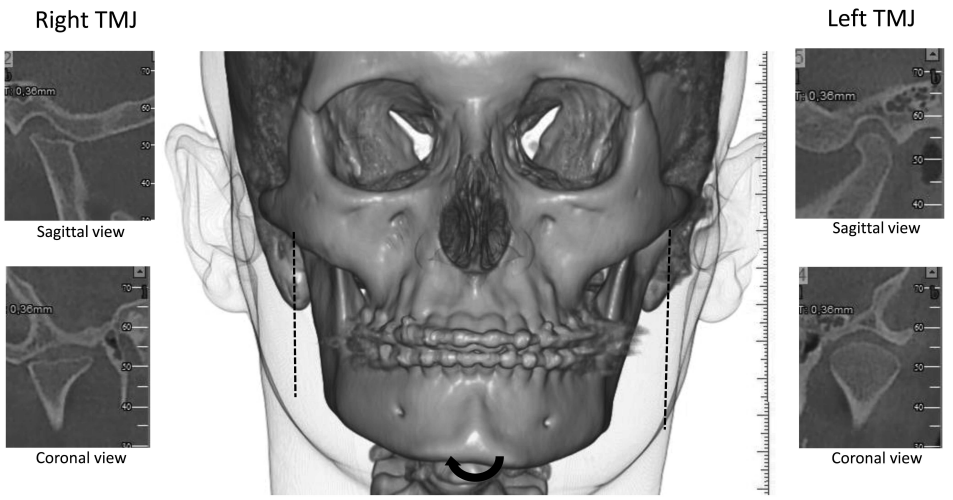

Figure 1. Fifteen-year-old girl with isolated arthritis in the right TMJ causing functional disabilities and moderate orofacial symptoms. Right TMJ: radiological signs of pronounced TMJ deformity with condylar flattening and erosive surfaces changes. Left TMJ: normal condylar outline and shape of TMJ components. Full-face cone-beam computerized tomography displays pronounced arthritis-induced dentofacial asymmetry; reduced vertical mandibular development on the right side compared to left side (dotted lines). The interside difference in vertical development leads to an abnormal mandibular growth-dependent rotation, indicated by the black arrow. TMJ: temporomandibular joint.

tation is uncommon. However, because TMJ arthritis in children with established JIA is often clinically silent, it is likely that a large number of patients with isolated TMJ arthritis were, and continue to be, overlooked.

The large variance in patient numbers from the different tertiary care centers may illustrate the varying levels of collaboration of pediatric rheumatologists with other specialties, and the importance of the first referral being directed to a dental versus medical specialty.

Diagnosis of JIA on the basis of TMJ changes, both by MRI findings of synovitis and effusion, and by radiographic findings of condylar resorption, is difficult[10,11]. The majority of patients in our study were diagnosed by gadolinium-enhanced MRI showing typical findings of inflammation; however, a small number was diagnosed on the strength of typical findings of joint deformations and abnormal/asymmetrical dentofacial development on cone-beam CT or radiograph studies, or by clinical assessment. The known frequency studies of TMJ arthritis in JIA are in patients with involvement of other joints[3,5,6,10]. Idiopathic condylar resorption (ICR), a condition of unknown origin occurring mostly in adolescent females, is also associated with flattening and thinning of the condylar heads and other mandibular changes similar to those observed in JIA[12,13]. Classified as a subgroup of conditions in the same order of joint diseases as systemic arthritides, ICR can be considered a low-inflammatory condition[14]. A suggested possible etiology of ICR could be repetitive microtrauma of the joint components caused by disc displacement with or without reduction as well as TMJ hypermobility[15]. In a recent study, 72% of a group of patients with ICR had bilateral TMJ involvement, but without other joints involved, indicating a systemic causality[16]. At this point it remains unknown whether JIA-related isolated TMJ arthritis and ICR are 2 distinct conditions, and whether ICR therefore constitutes a differential diagnosis to isolated TMJ arthritis. In fact, the question could be raised whether contemporary diagnostics fail to distinguish between these 2 conditions, with MRI findings for TMJ arthritis and ICR still remaining based on expert opinion. The consequence could be a failure in optimal treatment of isolated TMJ arthritis. Ours is a retrospective study based on a limited number of JIA patients with a condition difficult to diagnose with reasonable certainty. A limitation to our study is that no single standardized approach to interpretation of MRI of the TMJ exists. The overall strength of our study is the large catchment area of the 4 centers, working in close collaboration with orthodontists and maxillofacial surgeons.

Isolated TMJ arthritis may be the first or only manifestation of JIA and is probably not as rare as previously reported. Many patients are initially seen by dental or nonrheumatologic medical specialists, who may not be familiar with the condition, and the apparent diagnosis will be ICR or other TMJ disorders. Further studies are necessary to determine the true frequency of isolated TMJ arthritis in JIA and explore other possible causes for isolated TMJ arthritis and optimal therapy.

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