

Diagnosis Of Juvenile Rheumatoid Arthritis

Ronald W. Chapman, MD, MPH

Family physicians commonly see children with musculoskeletal complaints, and often these complaints are related to sports injuries and physical activities that are a part of childhood. Juvenile rheumatoid arthritis is important in the differential diagnosis of a child with prolonged musculoskeletal complaints. The following case highlights the importance of a complete history and physical examination in the diagnosis of juvenile rheumatoid arthritis.

Case Report

An 11-year-old girl came to the office complaining of a several-month history of bilateral thumb pain and swelling. The symptoms began during volleyball season. She was noted by family members to be an athlete and excelled in a number of organized sports. Volleyball season had ended several weeks before her visit, yet the thumb symptoms persisted. Further history revealed complaints of bilateral ankle and foot swelling and pain. Symptoms were noted to be worse in the mornings and associated with joint stiffness. There was no recent history of trauma, streptococcal infection, antibiotic use, or tick bite. Family history was negative for rheumatic disease. Review of symptoms was unremarkable, and there was no history of fevers, joint redness, or visual changes.

On physical examination she weighed 78 pounds and stood 4 feet, 9 inches. Her temperature was 98.6°F, her blood pressure was 110/70 mmHg, and her pulse was regular at 72 beats per minute. In general she was well developed, well nourished, and in no distress. She had no gait disturbance. Rheumatologic examination of the axial skeleton showed nontender sacroiliac joints and normal findings on back and neck examination. Upper extremity examination showed bilateral fusiform swelling and tenderness of the first and second proximal interphalangeal joints.

Lower extremity examination showed trace, non-pitting edema of the dorsum of both feet. All joints were noted to have full range of motion and were without redness or obvious deformities.

Given the presentation of polyarticular involvement, laboratory and radiologic information was obtained. Radiographs of her feet showed periarticular loss of bone density consistent with juvenile rheumatoid arthritis. A chemistry panel showed a slightly elevated cholesterol level of 187 mg/dL (acceptable <170 mg/dL). Results of a complete blood count and urinalysis were normal. Her erythrocyte sedimentation rate was elevated to 33 mm/h (normal 0 to 15 mm/h). The rheumatoid arthritis latex fixation test was positive with a titer of 1:160. The antinuclear antibody and Lyme titers were negative.

The patient's condition was diagnosed as polyarticular seropositive juvenile rheumatoid arthritis. She was prescribed salicylates with the goal of a serum level of 20.0 to 30.0 mg/dL. Eventually the child achieved steady, therapeutic levels on eight 325-mg aspirin per day. After some initial improvement, at 9 months the patient had a relapse and was referred to a rheumatologist. After an unsuccessful month on tolmetin, 200 mg four times a day, and because she developed a valgus subtalar deformity of her right ankle, she was prescribed aurothioglucose, 40 mg intramuscularly every week. In the subsequent 4 months of weekly gold injections and close observation, the patient has responded well with decreased joint swelling and stiffness.

Discussion

The estimated prevalence of juvenile rheumatoid arthritis is 0.5 per 1000 children.^{1,2} Between 60,000 and 250,000 children in the United States have juvenile rheumatoid arthritis.³ Many terms have been used to describe this disease, including Still disease,⁴ juvenile arthritis, juvenile chronic polyarthritis, and chronic childhood arthritis.⁵ Some clinicians use the term juvenile arthritis in a global sense and juvenile rheumatoid arthritis for those patients who are rheumatoid factor seropositive.

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From the Shasta Cascade Family Practice Residency Program, Redding, California. Address reprint requests to Ronald W. Chapman, MD, MPH, Shasta Cascade Family Practice Residency Program, 2480 Sonoma Street, Redding, CA 96001.

Juvenile arthritis is classically divided into three subgroups: polyarticular, pauciarticular, and systemic. These subgroups are further divided into polyarticular rheumatoid factor seropositive and seronegative and pauciarticular types I and II. The critical differences are the presence of systemic symptoms, the type and number of joints involved, and the results of the rheumatoid factor and antinuclear antibody studies. Table 1 gives a concise overview of the differences between the subgroups.

Juvenile arthritis is a clinical diagnosis made by careful history and physical examination to rule out other more common conditions. One important criterion for the diagnosis of juvenile arthritis is its chronicity. The diagnosis requires persistent signs and symptoms of arthritis for more than 6 weeks. The polyarticular subgroup of patients has joint stiffness, swelling, and loss of motion, which can be gradual or sudden. Morning stiffness, as in this case report, is typical and often the only complaint. Any synovial joint can be involved, often symmetrically, as were the patient's feet and fingers in the case report. Iridocyclitis,⁶ sacroiliitis, and arthritis are characteristic of the pauciarticular subgroup. Patients with systemic juvenile arthritis have fevers, rash, arthritis, and lymphadenopathy and can have multiple organ involvement.

The differential diagnosis of pediatric musculoskeletal complaints is extensive. It is critical to exclude the possibility of a bacterial arthritis or osteomyelitis by arthrocentesis in a child with fevers and a swollen, tender, and red joint. History,

level of suspicion, and appropriate radiologic studies should rule out accidental and nonaccidental injuries. A complete blood count will help exclude several diagnoses, such as leukemia in patients who can have musculoskeletal involvement.⁷

Other useful studies include a urinalysis, clinical chemistry measurements, and Lyme titer measurements in endemic areas. The sedimentation rate is usually elevated during the inflammatory process. A positive rheumatoid factor occurs in only 5 percent of patients with juvenile arthritis, mostly in older children with polyarticular disease. Antinuclear antibodies are more often positive than is the rheumatoid factor and are found with polyarticular disease and pauciarticular type I. Radiologic changes can include osteoporosis, soft tissue swelling, and periostitis of affected joints.

Aspirin is the first line of treatment for juvenile arthritis. Salicylate blood levels should be maintained at 20 to 30 mg/dL and is usually attained with doses of 100 mg/kg of aspirin for children 25 kg or less and doses of 2 to 3 g for older, heavier children. The only nonsteroidal anti-inflammatory drug (NSAID) approved for use in juvenile arthritis is tolmetin and is an appropriate second-line agent. Careful observation is necessary to be aware of gastrointestinal side effects. Weekly intramuscular gold injections are used if aspirin or NSAIDs are ineffective. Gold therapy requires regular complete blood counts and close monitoring to be alert for toxicity, looking for rash, mucosal ulcers, leukopenia, thrombocytopenia, anemia, and

Table 1. Subgroups of Juvenile Rheumatoid Arthritis.*

Subgroup	Percent of Patients	Sex (%)	Age at Onset	Joints Involved	Rheumatoid Factor	Antinuclear Antibody	Prognosis (%)
Polyarticular RF negative	20-25	Female (90)	Childhood	Any, multiple	Negative	25% positive	Severe arthritis (10-15)
Polyarticular RF positive	5-10	Female (80)	Late childhood	Any, multiple	Positive	75% positive	Severe arthritis (50)
Pauciarticular type I	35-40	Male (80)	Early childhood	Large joints	Negative	90% positive	Ocular damage (10) Polyarthritis (20)
Pauciarticular type II	10-15	Male (90)	Late childhood	Large joints	Negative	Negative	Spondyloarthropathy (unknown)
Systemic	20	Male (60)	Childhood	Any, multiple	Negative	Negative	Severe arthritis (25)

RF — rheumatoid factor

*Adapted from *Nelson Textbook of Pediatrics*. 13th ed. Philadelphia: WB Saunders, 1987.

proteinuria. Other less widely used therapies include corticosteroids, chloroquine, hydroxychloroquine, and methotrexate.⁸⁻¹⁰ Physical and occupational therapy are also important to maintain muscle strength, coordination, and joint flexibility.

Overall the prognosis for juvenile arthritis is good, as 75 percent of patients have long-term remission without sequelae. Patients with polyarticular involvement who are positive for the rheumatoid factor have the poorest prognosis. Rarely is juvenile arthritis life-threatening. Children and their families should be encouraged to lead a normal life. The family physician can play an important role in the diagnosis and management of this rheumatologic disease of childhood.

References

1. Hoekelman RA. Primary pediatric care. St. Louis: C.V. Mosby Company, 1987.
2. Gare BA, Fasth A. Epidemiology of juvenile chronic arthritis in southwestern Sweden: a 5-year prospective population study. *Pediatrics* 1992; 90:950-8.
3. Gewanter HL, Roghmann KJ, Baum J. The prevalence of juvenile arthritis. *Arthritis Rheum* 1983; 26:599-603.
4. Juvenile and adult systemic-onset Still's disease. *Lancet* 1990; 336:92.
5. Nelson WE. Nelson textbook of pediatrics. 13th ed. Philadelphia: W.B. Saunders, 1987.
6. American Academy of Pediatrics. Section on Rheumatology and Section on Ophthalmology. Guidelines for ophthalmologic examinations in children with juvenile rheumatoid arthritis. *Pediatrics* 1993; 92:295-6.
7. Ostrov BE, Goldsmith DP, Athreya BH. Differentiation of systemic juvenile rheumatoid arthritis from acute leukemia near the onset of disease. *J Pediatr* 1993; 122:595-8.
8. Rosenberg AM. Advanced drug therapy for juvenile rheumatoid arthritis. *J Pediatr* 1989; 114:171-8.
9. Rose CD, Singen BH, Eichenfield AH, Goldsmith DP, Athreya BH. Safety and efficacy of methotrexate therapy for juvenile rheumatoid arthritis. *J Pediatr* 1990; 117:653-9.
10. Gianini EH, Brewer EJ, Miller ML, Gibbs D, Passo MH, Hoyerall HM, et al. Ibuprofen suspension in the treatment of juvenile rheumatoid arthritis. Pediatric Rheumatologic Collaborative Study Group. *J Pediatr* 1990; 117:645-52.