swelling and pain of the right elbow, both wrists, the right knee, and the right ankle at 16 years of age. Her ESR was 69 mm/hr, CRP 4.35 mg/dl, and platelet count 402 x 10^9/l. RF and ANA were negative. IgG was 1,810 mg/dl, IgA 363 mg/dl, and IgM 153 mg/dl. She had no ocular involvement.

Before a diagnosis of Turner's syndrome is made in a patient, the SRY status should be examined. SRY-positive cases are defined as mixed gonadal dysgenesis and must be excluded from the group of Turner's syndrome. However, Zulian *et al.* did not describe the SRY status in their patients, so SRY-positive patients with mixed gonadal dysgenesis could have been included in their study.

Turner's syndrome may be complicated by several autoimmune diseases, such as Hashimoto's disease. Chronic arthritis is rare in comparison with the other complications. Patients with Turner's syndrome are characterized by short stature; their growth plates are often not closed at 20 years of age despite a growth hormone deficiency and they are not adult in terms of bone age. Childhood or juvenile status is usually defined by the chronological age. However, we should perhaps instead consider juvenile arthritis according to a different concept of age such as the biological age. In most patients with Turner's syndrome the growth plate at the wrist joint remains open after 16 years of age because of bone pre-maturation linked to gonadal insufficiency. Thus, arthritis in Turner's syndrome patients over 16 years of age does not strictly speaking represent adult rheumatoid arthritis. "Juvenile arthritis in Turner's syndrome" rather than the terms JRA or JCA used by the authors, may be the most suitable term from this viewpoint.

Y. INAMO, *MD, PhD*

Divisions of Pediatric Rheumatology and Endocrinology, Department of General Pediatrics, Nihon University Nerima-Hikarigaoka Hospital, 2-11-1 Hikarigaoka, Nerima-ku 179-0072, Tokyo, Japan

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Reply

Sir,

Dr. Y. Innamo has reported the first Japanese patient with juvenile arthritis (JA) and Turner's syndrome (TS). We would have been glad to include this patient in our series, but when we conducted the study it was restricted to 66 pediatric rheumatology centers in Europe and North America.

He also suggested the need to define the SRY status of the patients with JA and TS reported in our study (1). As reported in the literature, the SRY test is not routinely carried out in all cases of TS (2). It has been recently introduced to detect a possible cryptic Y-sequence in patients with the 46X0 karyotype. These patients are, in fact, at risk of developing gonadoblastoma and have to be closely followed (3, 4). They have the phenotypic appearance of TS, however, and do not represent cases of the clinical entity of mixed gonadal dysgenesis, as proposed by Dr. Innamo (5).

In our retrospective study, the patients were collected from several centers and many of them were diagnosed before SRY testing became available. It is therefore possible that some of them were SRY-positive. This does not change the significance of our findings, however, particularly the evidence that the incidence of JA in TS is higher than expected, probably due to immune, genetic and/or hormonal reasons.

It is difficult to demonstrate that "JA is rare in comparison to other complications in TS" because our study was conducted in pediatric rheumatology centers and not in endocrinological units where patients with Hashimoto's thyroiditis (6) or IDDM (7) are followed.

Finally, Dr. Innamo suggested that the term juvenile arthritis and not juvenile chronic arthritis or juvenile rheumatoid arthritis be used to define this group of patients. We agree with him; in fact the title of our paper and the terminology defined in the Material and Methods section clearly anticipated his comment.

F. ZULIAN, *M.D.*

Dipartimento di Pediatria, Università di Padova, via Giustiniani 3, 35128 Padova, Italy.

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Behçet's disease with a cecal perforation

Sirs,

Behçet's disease (BD) is a type of systemic vasculitis characterised by recurrent aphtous ulcers of the mouth and genitalia, various skin lesions, arthritis, and panuveitis. Arteries and veins of all sizes, the central nervous system, and the gastrointestinal tract (GI) are involved with serious consequences (1). We describe a patient with BD who presented with a right lower abdominal mass, and was subsequently found to have cecal perforation with abscess formation.

A 21-year-old male was admitted with a 2-month history of recurrent right lower abdominal pain. He had suffered over the last 3 years from recurrent oral and genital ulcers, pustular lesions on the trunk, and attacks of anterior uveitis. Ten months earlier he experienced an episode of hematochezia. Colonic ulcerations were demonstrated radiologically and a treatment regimen consisting of prednisone, colchicine and sulphasalasine was commenced with a diagnosis of Behçet's colitis. He became symptomless on this therapy until 4 months before admission when he was diagnosed with pulmonary tuberculosis. At that time anti-tuberculous therapy was started, while sulphasalasine and steroid were stopped.

On physical examination his abdomen was soft, but a firm, tender mass was palpated in the right lower quadrant. He had multiple oral ulcers and an active ulcer on the scrotum together with scarred ulcers. On admission the only abnormal blood test was leucocytosis of 14,000/mm3 with 90% neutrophils. Ultrasonographic examination revealed a heterogeneous solid mass in the right iliac fossa (diameter 3 x 6 cm).

The patient underwent an exploratory laparotomy which revealed matted loops of bowel adherent to the pericecal abscess and to the surrounding tissues. The distal ileum and the proximal part of the right-sided colon were resected, and an end-to-side ileocolostomy was performed.

Macroscopically, there was a perforation at

the caecum 3 cm in diameter. There were also deep, oval shaped ulcers at the ascending colon and ileocecal valve. Microscopic examination showed sharply demarcated ulcers that penetrated through the mucosa into the submucosa or superficial muscularis propria. The base of the ulcers was lined with nonspecific granulous tissue and an overlying inflammatory exudate. The walls of many venules adjacent to the ulcers in the submucosa were infiltrated by lymphocytes (Fig. 1). The patient was discharged on steroid and sulphasalasine therapy and became asymptomatic.

The frequency of GI involvement in BD shows geographical differences. It is high in patients from Japan (50-60%), in contrast to patients from Turkey (0-5%) (2). The typical manifestation is round or oval ulcers which can be seen throughout the alimentary tract, but which are most commonly seen in the terminal ileum and the caecum (75% of patients) (2).

We have identified 11 reported cases of intestinal perforation in patients with BD (3-7). The site of the perforation was the ileum in 2, the caecum in 7 and other parts of the colon in 2 patients. All of the reported cases showed free perforation with generalised peritonitis, whereas ours did not. The chronic use of antibiotics (anti-tuberculous drugs) may have been the cause of abscess formation in our case.

The presence of active pulmonary tuberculosis entailed a further consideration of intestinal tuberculosis in the differential diagnosis. The absence of granuloma and negative staining for acid fast bacilli in the colonic specimens and regional lymph nodes excluded this diagnosis in our patient, however (8).

Some reports have suggested the similarity between Behçet's colitis and inflammatory bowel disease. (7) Our patient's colitis, which was characterised by a right colonic predominance, rectal sparing and discontinuous involvement, was different from that seen in ulcerative colitis. The right-sided involvement is consistent with Crohn's disease, but the absence of transmural inflammation, granuloma, thickening of the bowel wall, and luminal stenosis makes this diagnosis unlikely. Sulphasalasine has been suggested to be of benefit in gastrointestinal BD (9). The exacerbation of our patient's symptoms seen after the withdrawal of sulphasalasine could support this hypothesis.

Fig. 1. An ulcer of the colon extending into the submucosa and a thrombosed vessel adjacent to the submucosa (hematoxylin and eosin, magnification x 200).

I. SIMSEK[1] O. GÜNHAN[3]
A. DINC[2] F. KOCABALKAN[1]
M.R. MAS[1]

[1]Department of Internal Medicine, [2]Department of Rheumatology, [3]Department of Pathology, Gülhane Military Medical Faculty, Ankara, Turkey
Please address correspondence and reprint request to: Dr. Ismail Simsek, Gülhane As. Tip Akademisi, Iç Hastaliklari B.D, 06018 Etlik (Ankara), Turkey.
E-mail: ismail.simsek@isbank.net.tr

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Remitting seronegative symmetrical synovitis with pitting edema and primary Sjögren's syndrome

Sir,

Remitting seronegative symmetrical synovitis with pitting edema (RS3PE) is characterised by symmetrical synovitis involving predominantly the wrists and flexor digitorum tendon sheaths and associated with marked edema of the dorsum of the hands (1). RS3PE may be the inaugural form of different rheumatic diseases of the elderly (2, 3), malignancies (4-6) and myelodysplastic syndromes (6, 7). Primary Sjögren's syndrome (pSS) is an autoimmune disease characterised by chronic lymphocytic infiltration of the salivary and lachrymal glands that, although most common in middle-aged adults, may have an onset in advanced age, with a spectrum of clinical and immunological features similar to that in younger patients (8). To our knowledge, however, there is only one report of RS3PE preceding SS (2). We would like to report an elderly patient in whom RS3PE antedated the onset of pSS by many months.

A 74-year-old man suffering from long-standing blood hypertension treated with 20 mg of enalapril daily, presented in August 1997 with a 4-month history of fatigue and fever, as well as pain and swelling of the wrists, hands and feet; he did not complain of shoulder symptoms. Physical examination showed swelling of the flexor digitorum tendons at the wrists, as well as swelling and tenderness of the metacarpophalangeal joints and marked bilateral pitting edema of the dorsum of the hands and feet.

Laboratory tests revealed an elevation of the erythrocyte sedimentation rate (ESR) (110 mm/hr, Westergren), C-reactive protein (CRP) (5.6 mg/dl) (nv < 0.50 mg/dl), fibrinogen (610 mg/dl) (nv < 400 mg/dl), α_2-globulins (15.4%) (nv < 12%) and white blood cell count (14,500/mm^3). Rheumatoid factor (RF), antinuclear (ANA), anti-native DNA and anti-ENA autoantibodies were negative. In addition, hepatitis B and C virus and tumor markers were also negative.

Radiographs of the hands, wrists and feet demonstrated diffuse osteopenia and soft tissue edema, but no bone erosions. Ultrasonography of the hands showed an anechoic area surrounding the flexor and extensor tendons indicative of tenosynovitis. A diagnosis of RS3PE was made and 15 mg of prednisone daily was started, resulting in complete remission of the clinical picture after a few days. One month later we began tapering the prednisone dose every other day and by 1.25