The Turkish Journal of Pediatrics 1997; 39: 277-280

A CASE OF JUVENILE ANKYLOSING SPONDYLITIS AND CROHN'S DISEASE*

*Erbil Ünsal MD**, *Yasemen Eroğlu MD***, *Benal Büyükgebiz MD****
*Ali Küpelioğlu MD*****, *Necla T. Çevik MD****

SUMMARY: Ünsal E, Eroğlu Y, Büyükgebiz B, Küpelioğlu A, Çevik NT. (Departments of Pediatrics and Pathology, Dokuz Eylül University Faculty of Medicine, İzmir, Turkey). A case of juvenile ankylosing spondylitis and Crohn's disease. Turk J Pediatr 1997; 39: 277-280.

Juvenile ankylosing spondylitis (JAS) is a chronic inflammatory arthritis of the peripheral and axial skeleton, frequently accompanied by enthesitis. About four percent of patients with JAS have ulcerative colitis or Crohn's disease. Crohn's disease is the more common of the two and is diagnosed in 26 percent of patients with chronic spondyloarthropathy. In this paper, a 14-year-old male patient is presented as a typical case of juvenile ankylosing spondylitis and Crohn's disease with low back pain, morning stiffness, limited motion in anterior and lateral flexion and extension, left sacroiliitis, ankylosis in the apophyseal joints of the lumbar vertebrae, abdominal pain, bloody diarrhea, characteristic histopathologic changes of colonic involvement such as lymphoid follicles, fissures, submucosal polymorphonuclear cell infiltration and definite ganglion cells. The current therapy with mesalazin, having fewer side effects than sulfosalazin, and its applicability in combination with naproxen sodium is also discussed. *Key words: ankylosing spondylitis, Crohn's disease, mesalazin.*

Juvenile ankylosing spondylitis (JAS) is a genetically based, chronic inflammatory arthritis of the peripheral and axial skleton, frequently accompanied by enthesitis, and not characterized by rheumatoid factor (RF) or antinuclear antibody (ANA) seropositivity. The sacroiliac joints are eventually affected in all patients with JAS, and radiologic evidence of inflammation of these joints is required for a definitive diagnosis[1]. JAS has distinctive features such as familial aggregation, association with HLA B27, enthesopathy, sacroiliitis and onset mostly after the age of 10 years[2]. About four percent of patients with JAS evaluated by radiology and sigmoidoscopy have ulcerative colitis or Crohn's disease. Crohn's disease is the more common of the two and is diagnosed in 26 percent of patients with chronic spondyloarthropathy[3]. In this paper, a case having both JAS and Crohn's disease is presented.

* From the Departments of Pediatrics and Pathology, Dokuz Eylül University Faculty of Medicine, İzmir.

** Instructor in Pediatrics, Dokuz Eylül University Faculty of Medicine.

*** Professor of Pediatrics, Dokuz Eylül University Faculty of Medicine.

**** Professor of Pathology, Dokuz Eylül University Faculty of Medicine.

Case Report

A 14-year-old male patient was referred to the Pediatric Rheumatology Unit suffering from low back pain, morning stiffness, abdominal pain and bloody diarrhea three times a day. He had had gastrointestinal system complaints for three years and low back pain for two. In the previous two months he had had three fainting and syncope attacks. Physical examination revealed tenderness and pain of the left sacroiliac joint. He also had limited motion in anterior and lateral flexion and extension. He had no signs or symptoms of uveitis. In the laboratory evaluation, the hemoglobin level was 9.7 g/dl with microcytosis and hypochromia, C-reactive protein was strongly positive, erythrocyte sedimentation rate (ESR) was 60 mm/hr, and RF and ANA were negative. X-ray examination showed left sacroiliitis with ankylosis in the apophyseal jounts of the lumbar vertebrae (Fig. 1). Intestinal barium enema was not helpful because of colon gases. Colonoscopy was not available. A diagnostic approach by rectal biopsy revealed lymphoid follicles, fissures, submucosal polymorphonuclear cell infiltration and definite ganglion cells compatible with Crohn's disease (Fig. 2). The patient was given mesalazin 750 mg and naproxen sodium 500 mg a day. In the first month of therapy, significant regression was obtained in the number and the bloody character of stools as well as in the low back pain.

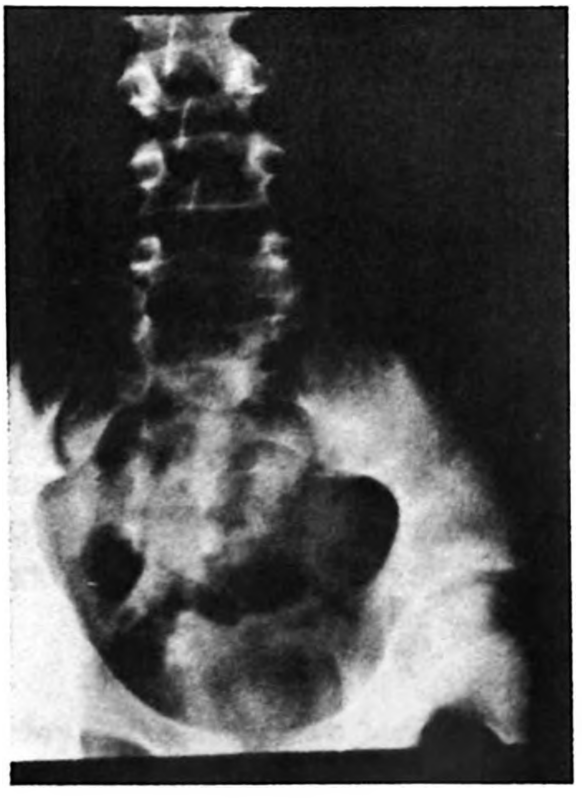

Fig. 1: Ankylosis in the apophyseal joints of the lumbar vertebrae with closed left sacroiliac joint.

Fig. 2: Marked lymphoid tissue in the mucosal layer and hyperplasia of the lymphoid follicle is clearly seen at the right. There is also a fissure tract of the mucosa in the middle. (Hematoxylen + Eosin x 4).

Discussion

Crohn's disease is a chronic inflammatory disease that is characterized by unpredictable exacerbations and remissions in disease activity. The cause is entirely unknown. The currently held hypothesis is that it occurs as a result of a genetically determined response which is probably immunologically mediated. Other theories include alterations in autoprotection due to abnormal metabolism of the prostoglandin-leukotriene system or changes in the mucus layer[4,5]. Increased permeability of the gut wall to known and unknown antigenic material together with a defective local immune regulation are implicated to be sufficient to initiate joint inflammation in genetically predisposed individuals. In Crohn's disease, articular manifestations may precede intestinal signs by years[1]. In this patient, after a year of passing bloody stools, low back pain occurred with limitation of motion in anterior flexion, lateral flexion and extension caused by ankylosis in the apophyseal joints and left sacroiliitis. Findings in the rectal biopsy established the diagnosis of Crohn's disease associated with ankylosing spondylitis.

In a clinical, radiological and immunogenetical study of 51 Crohn's patients, rheumatological disorders were found in 16 (31%) of them[6]. Spondylitis clinically and radiologically indistinguishable from idiopathic ankylosing spondylitis was detected in three to six percent of patients with inflammatory bowel disease in one prospective study[7]. In chronic spondyloarthropathies, and especially in ankylosing spondylitis, gastrointestinal infection also plays a role in which the inflammation of joints is triggered by specific bacterial pathogens.

The therapy of Crohn's disease has been the focus of several multicentered studies. Symptomatic therapy with prednisone, sulfosalazine, metronidazole, azathioprine or 6-mercaptopurine, together with nutritional support are the main goals. Sulfosalazine and its derivative mesalazin, the latter having fewer, side effects, have been shown to be useful in inducing remission in patients with colonic involvement[4]. This patient has been well controlled by mesalazin and naproxen sodium. Interestingly, nonsteroidal antiinflammatory drugs have not been found to be associated with gut lesions[3]. Therefore, in our opinion, a combination of the two drugs is recommended to control symptoms.

REFERENCES

1. Russell AS. Ankylosing spondylitis. In: Klippel JH, Dieppe PA (eds) Rheumatology. St. Louis: CV Mosby Co, Yearbook Europe Limited 1994; 23.1-24.5.
2. Hussein A. HLA-B27-associated spondyloarthritis in childhood. Monatsschr Kinderheilkd 1987; 135: 185-194.
3. Leirisalo-Repo M, Turunen U, Stenman S, et al. High frequency of silent inflammatory bowel disease in spondylarthropathy. Arthritis Rheum 1994; 37: 23-31.
4. Kirschner BS. Inflammatory bowel disease in children. Pediatr Clin North Am 1988; 35: 189-208.
5. Daum F. Pediatric inflammatory bowel disease. In: Silvelberg A, Daum F (eds). Textbook of Pediatric Gastroenterology (2nd ed). Philadelphia: Yearbook Medical Publishers Inc; 1988: 392-418.
6. Modena V, Amoroso A, Frattasio C, et al. HLA antigens and clinical manifestations in Crohn's disease. Clin Exp Rheumatol 1988; 6: 221-225.
7. Gravallese EM, Kantrowitz FG. Arthritic manifestations of inflammatory bowel disease. Am J Gastroenterol 1988; 83: 703-709.