

Ayça TÖREL ERGÜR¹
Melih AKYOL²
Esin YILDIZ³

Infective Panniculitis in a Child With Juvenile Rheumatoid Arthritis and IgA Deficiency

Departments of ¹Pediatrics, ²Dermatology,
³Pathology, Faculty of Medicine, Cumhuriyet
University, Sivas-TURKEY

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In dermal and mucosal defects, the most important natural defence mechanism against infections is secretory IgA (1). Juvenile rheumatoid arthritis (JRA) accompanied by chronic arthritis together with immunodeficiency syndromes is rarely seen (2). A case of pauciarticular disease type I associated with infective panniculitis and IgA deficiency is reported in this study.

Case

A 12-year-old girl came to Cumhuriyet University Medical Faculty with the complaints of joint pain, weakness, weight loss and a wound on the right lower leg in March 1998. These symptoms (excluding the wound) had occurred for the last three years. On physical examination, the blood pressure was 110/70 mmHg with a pulse rate of 80 beats per minute. She was 146 cm (5%p) in height and 35 kg (5%p) in weight. Her general condition was bad. Pupilla light reflexes were +/-, pupillaries were isochoric. Heart beats were rhythmic, and there was a 1/6 systolic murmur on the apex. In the abdominal examination, bowel sounds were noted to be normoactive, her liver was 2 cm palpable at the midclavicular line, and the spleen was nonpalpable. Examination of the skin revealed erythematous lesions, eroded areas and defects of tissue markedly swollen, particularly on the right lower leg (Figure 1). Pathergy test was negative. She had bilateral arthritis of the knee joints. The examination of other systems yielded normal results. Blood analysis showed anemia (Hb: 8.2 g/L), a sedimentation rate of 84 mm/h, white cell count of 3400/mm³, and a mild thrombocytosis (444,000/mm³). Hypochromic microcytic anemia was found in the peripheral smears. In addition, laboratory studies showed

negative rheumatoid factor (7 IU ↓), mild positive antinuclear antibody test (1:80), positive C-reactive protein (36 mg/l), negative antiDNA and a markedly decreased level of IgA (4.93 mg/dl). The other immunoglobulin levels were within the normal limits. C3, C4 complement levels, hepatitis A, B, C, D, E antigens and antibodies, amylase, and the other biochemical analyses were normal. No abnormalities were observed in the chest roentgenogram or electrocardiogram. No microorganisms were cultured from repeated swabs. A skin biopsy obtained from the wound region showed prominent edematous changes and inflammatory cell infiltration in the reticular dermis. In addition, lobular panniculitis was accompanied by a massive cellular infiltration consisting of numerous neutrophils and occasional eosinophils (Figures 2, 3). There was acanthosis of the whole epidermis. On the other hand, there was neither periarterial or perivenular vasculitis nor endothelial proliferation. Stainings for microorganisms, including those for acid-phase bacteria, were negative.

In the light of the above findings, the patient diagnosed with type I pauciarticular JRA according to modified JRA criteria (3) was treated by nonsteroidal anti-inflammatory drugs and antibiotics. The erythematous swelling of the affected leg regressed moderately in a short time and within two weeks the ulcers became clean and were totally healed. No relapse occurred following the treatment.

Neutrophilic panniculitis in a patient with rheumatoid arthritis was described by Newton et al. in 1988 (4). Only three cases of panniculitis associated with rheumatoid arthritis have previously been reported (5). However, all of the cases covered in their study were adults, there



Figure 1. The clinical appearance of the lesion.

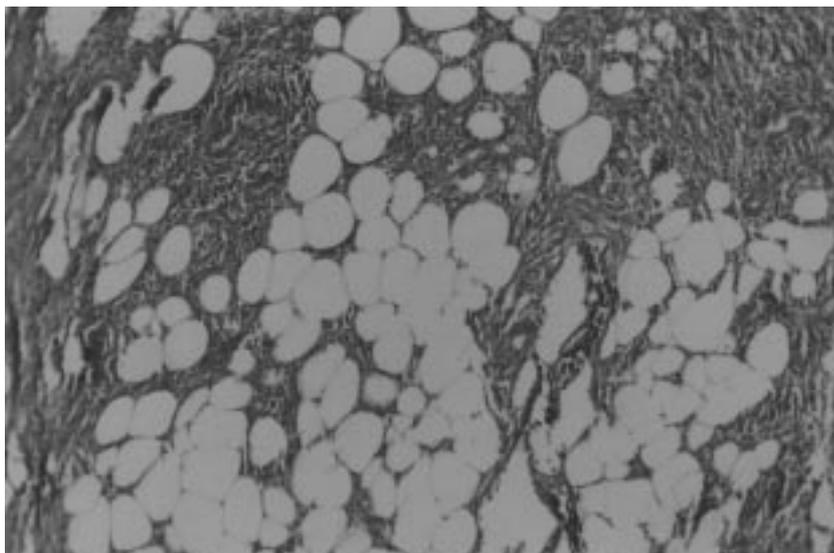


Figure 2. The histopathologic appearance of the specimen of the lesion (HE, x 75).

were no children. Moreover, all of the cases had panniculitis, leukocytoclastic immune-complex vasculitis. Kuniyuki et al. postulated that the occurrence of panniculitis in rheumatoid arthritis associated with immunological changes was the result of the cumulation of immune complexes (5). Kuniyuki et al. agreed with Winkelman and Anley (6,7).

In our case, the histopathologic examination showed no vasculitis, endothelial proliferation, fibrinoid necrosis or leukocytoclasia. Panniculitis was mixed septal/lobular and intensive polymorphonuclear leukocytosis (PMNL) was also present. These findings indicated no association with leukocytoclastic immune-complex vasculitis as

described in previous publications (8). The usage of antibiotics and anti-inflammatory drugs without the administration of cortison and/or immunosuppressive agents resulted in the healing of the wound supporting the hypothesis that the disease was infective panniculitis rather than immune-complex vasculitis. In our case determination of the infective panniculitis indicated that other factors instead of immune-complexes may play a role in the pathogenesis of panniculitis in JRA.

In panniculitis, the differential diagnosis is also made on the systemic lupus erythematosus (SLE), scleroderma, dermatomyositis and other connective tissue disorders, pyoderma gangrenosum, tuberculosis and sarcoidosis (8).

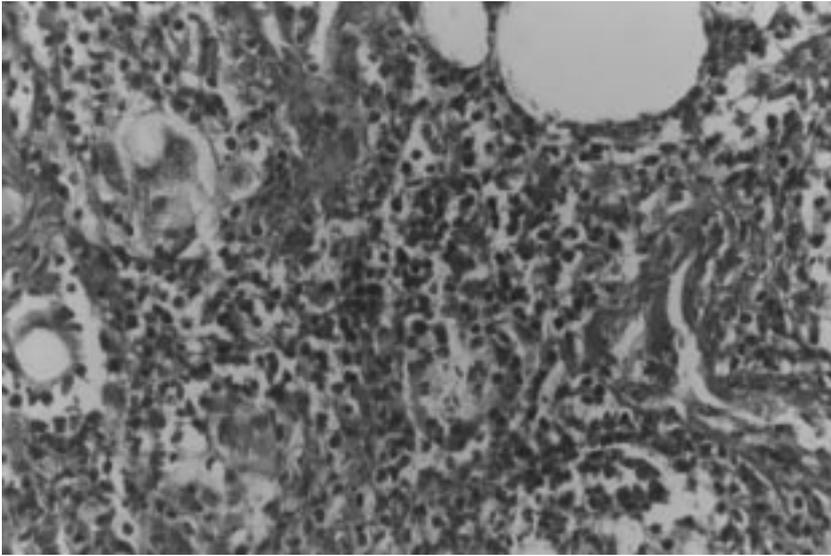


Figure 3. Lobular panniculitis accompanied by a massive cellular infiltration (HE, x 150).

Considering the histopathologic examination of the lesion and also its location our case was not likely to be connective tissue disorder panniculitis. In connective tissue disorder panniculitis, there is epidermal atrophy and intensive lymphocytic infiltration. Frequently they take place on the trunk symmetrically. In tuberculosis and sarcoidosis, the histopathological characteristics of the lesion are granulomatous appearance and widespread lymphocytic infiltration (8). Pyoderma gangrenosum can produce ulcers that are difficult to distinguish from those of an ulcerating panniculitis. Biopsies taken from the lesions of pyoderma gangrenosum show pustules and lymphocytic inflammation in the dermis (9). However, there was extensive neutrophilic infiltration in histopathologic examination of the lesion in our case.

Moreover, pyoderma gangrenosum responds to immunosuppressive treatments and pathergy reaction is positive (10), whereas the ulcers were totally healed by the treatment with antibiotics in our case.

In conclusion, this report presenting a case of JRA with IgA deficiency together with developing infective panniculitis was considered interesting since no previous reports on this subject were found in the literature.

Correspondence author:

Ayça TÖREL ERGÜR

Örtülüpınar Mah. Selçuklu Sok.

Orhan Kurt Sitesi Yakın Apt.

No: 6/2 58030 SİVAS

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