Letters to the Editor

more commonly among men in their 50s. Back pain (19.1%) was the most common rheumatic manifestation. Myalgia (15.5%) was also frequently observed, as was arthralgia without arthritis (10%). Peripheral arthritis (13.6%) was also present, but generally as monoarthritis. In contrast, saccroilitis was only observed in a single patient. This patient was a man in his early twenties, who had positive blood cultures for *Enterococcus faecalis*, and experienced a progressive improvement of his saccroiliac pain following antibiotic therapy with ampicillin and gentamicin.

It is worth noting that in our series of patients with IE and rheumatic manifestations microhematuria was observed in almost 60% of the patients, versus 27% in IE patients without rheumatic manifestations (2). In summary, unexplained peripheral synovitis, arthralgia or low back pain along with unexplained microhematuria may be possible warning signs for the presence of IE. Awareness of these complications may be useful to avoid inappropriate delay in the diagnosis of this severe disease.

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References


Macrophage activation syndrome as the initial manifestation of systemic onset juvenile idiopathic arthritis

Sirs,

The macrophage activation syndrome (MAS), a clinical syndrome caused by excessive proliferation and activation of well-differentiated macrophages, has been associated with a heterogeneous group of conditions that include drugs, infections, and neoplastic and rheumatologic diseases. MAS has been described in association with systemic onset juvenile idiopathic arthritis (soJIA), with various triggering events such as bacterial or viral illness or the use of nonsteroidal antiinflammatory drugs (NSAIDs), gold salts, sulfasalazine or methotrexate (MTX) (1). Because of the life-threatening nature of this syndrome, prompt recognition is mandatory and treatment with high doses of corticosteroids and cyclosporine A in MAS associated with soJIA has been advocated (2). We present the case of a 2-year-old boy who developed MAS syndrome shortly after the start of a febrile arthritis retrospectively diagnosed as soJIA and only 4 days after treatment with salicylates was initiated.

The patient was admitted to our hospital after 8 days of high spiking daily fever reaching 39.5°C and microhematuria of the right wrist. On admittance, physical examination showed a slight swelling over the ulnar area of the right wrist with normal general, neurological and ophthalmologic examinations. An evanescent erythematous maculopapular rash over the trunk and upper extremities was evidenced. Laboratory data showed a white blood cell count of 17.4 x 10^9/l (72% granulocytes), Hb 10.6 g/dl, hematocrit 31%, and platelet count 486 x 10^9/l. Serum urate, creatinine, calcium, phosphorus, alkaline phosphatase, transaminases, and -glutamyltranspeptidase were within the normal range. The prothrombin time (PT) and partial thromboplastine time (PTT) were normal. Rheumatoid factor and antinuclear antibodies were negative. C-reactive protein was elevated to 216 mg/l. Ferritin was elevated to 56,807 g/L with normal serum iron level and binding. Serologic tests for the usual bacterial or viral diseases were negative. An abdominal echography showed hepatomegaly with normal kidneys and spleen. A thorax radiograph was normal. Bone radiographs showed soft tissue swelling. The electrocardiogram was normal.

On day 3 after admission acetylsalicylic acid was initiated (100 mg/kg/day). Melena appeared on day 6 together with maintained fever, 77 x 10^9/l platelets, Hb 8.70 g/dl, and 8.2 x 10^9 leucocytes. PT and PTT were prolonged at 23.3 seconds (normal 11-15) and 57 seconds (normal 25-40), respectively, with low fibrinogen 1.09 g/L. GPT was elevated to 83 U/L, GOT to 77 U/L and LDH to 9,570 U/L. Hypoalbuminemia 30 g/L was noted. Triglycerides were 2.27 g/L. Platelets reached 26 x 10^9/Lo/L that platelets and fresh plasma were necessary. A gastroscopic disclosed 4 erosions over the antrum and a hyperaemic fundus suggesting NSAID gastropathy. Saliycylates were stopped. Bone marrow aspiration showed the presence of abundant macrophages with phagocytosis (Fig. 1).

MAS was diagnosed and glucocorticoids 1 mg/kg/day were initiated. The corticosteroid dose was gradually increased up to 3.5 mg/ kg/day to control the systemic disease and fever. After 10 days of corticosteroid therapy, platelets and a coagulation test were normal and the patient was discharged. Two months later, because of tenosynovitis of both shoulders and malaise, oral MTX up to 0.6 mg/kg/day was administered with resolution of the joint symptoms. Presently corticosteroids have been stopped. No MAS recurrence was found after MTX introduction.

MAS is a clinical syndrome characterised by persistent unremitting fever, lymphadenopathy, hepatosplenomegaly, mental status changes, easy bleeding, depression of the three blood cell lines, low erythrocyte sedimentation rate (ESR), and elevated serum liver enzyme values (1). The pathologic finding on bone marrow aspiration is the presence of numerous well-differentiated macrophages actively phagocytosing hematopoietic elements (3). SoJIA is a multisystemic disease in which extra-articular features are prominent and include fever, rash, lymphadenopathy, hepatosplenomegaly, cardiitis and laboratory abnormalities such as anemia, leukocytosis, thrombocytosis and hyperferritinemia. MAS has been des-
Celiac disease with a “crowned” odontoid process

Sirs,

Celiac disease is now considered an autoimmune disease. Recently tissue transglutaminase (tTG), a widely distributed protein in human organs, has been taken into consideration as an autoantigen of celiac disease (1). The ingested gliadin might modify tTG unmasking cryptic epitopes, leading to the production of anti-endomyosal antibodies; the presentation of these self-peptides in association with specific HLA class II molecules by antigen presenting cells may activate the T cell population, priming and then perpetuating an autoimmune process (2). The complete elimination of gliadin by diet may stop these events.

Celiac disease may present in adult subjects with an atypical clinical picture represented by extra-intestinal manifestations such as osteopenia (3), recurrent abortions (4), neurological and psychiatric disorders (5) or arthralgias (6). The diagnosis of all forms of celiac disease is relevant, because untreated patients are at risk for the development of complications, particularly malignancies (7). Here we describe a case of celiac disease characterized by the unusual involvement of the atlantoaxial joint.

A 55-year-old woman was referred to our unit for arthromyalgias lasting for 15 years. The patient had had 4 pregnancies; 2 of these ended with premature delivery and subsequent death of the newborn. The physical examination did not reveal any relevant finding. ESR, C-reactive protein, a full blood count, CPK level, liver and renal function tests were normal; rheumatoid factor and antinuclear antibodies were negative. Since childhood the patient had presented two or three bowel movements per day, but she had never attached importance to this aspect, considering her bowel habits completely normal. Based on this finding we carried out a serological screening for celiac disease; antigliadin antibodies were positive (IgG 77.3 mg/l, normal < 18 mg/l; IgA 51.1 mg/L, normal < 3 mg/l), as were IgA anti-endomysium antibodies at the dilution of 1:10. Histologic examination of the duodenal mucosa showed atrophy of the villi, increased intra-epithelial lymphocytes and infiltration of the lamina propria with plasma cells and lymphocytes.

X-rays and magnetic resonance imaging of cervical spine showed minute calcifications in the form of a rosary around the odontoid process (Fig. 1); neither subluxation in any direction nor pannus near the odontoid process were observed.

Wrist, knee and pelvis x-rays were normal; moreover, the conditions frequently associated with celiac disease such as hyperparathyroidism, hemochromatosis and hypomagnesemia were excluded. A gluten-free diet caused normalization of the patient’s bowel habits and the progressive disappearance of arthromyalgias. After 9 months both IgA anti-endomysium and anti-gliadin were negative, while IgG anti-gliadin was still positive. Histologic examination of the duodenal mucosa after 1 year of a gluten-free diet showed regeneration of the villi and a strong reduction of lymphocyte infiltration.

In our patient longstanding arthromyalgias disappeared after a gluten-free diet; therefore we think that her arthromyalgias were caused by unrecognized celiac disease. The radiographic examination revealed minute calcifications in the form of a “crown” around the odontoid process. To our knowledge celiac disease has never been associat-

References

Letters to the Editor