Multiple cerebral abscesses in a woman with giant cell arteritis

Sirs,

Giant cell arteritis (GCA) is effectively suppressed by glucocorticoids; however, many side effects, such as an increase in susceptibility to infections and diabetes, may appear (1, 2). We present an elderly female patient with GCA who developed a rare complication - multiple cerebral abscesses - 2 months after corticosteroid therapy was started.

A 73-year-old woman consulted us in May 1998 because of headache and weight loss. Her ESR was 81 mm/hr. A temporal artery biopsy showed GCA findings. Prednisone therapy was started (40 mg/day) with a progressive improvement in the patient's symptomatology and a marked decrease in the ESR to within normal limits (6 mm/hr). One month later, hyperglycemia (325 mg/dl) was detected, and oral antidiabetic therapy was prescribed.

Two months after the GCA diagnosis was established, the patient was admitted to hospital because of intense weakness and progressive difficulty in left arm movements. She did not report headache or fever. A decrease in vision in the left eye, difficulty with memory, and a motor deficit in the left superior limb were observed. Hyperglycemia (318 mg/dl) and an increased ESR (38 mm/hr) were the only laboratory abnormalities found. A urine culture revealed *Staphylococcus aureus.*

Chest X-ray and echocardiogram were normal. A brain CT scan showed multiple areas of decreased attenuation at the right frontal and temporal lobes, surrounded by edema. CT scan with contrast material revealed a multi-compartmental ring-enhancing process at the same site (Fig. 1). The different sizes and the proximity of the lesions were more suggestive of an infectious process than of cerebral metastasis from the radiological point of view.

She required insulin, while metronidazole (1 g three times daily) plus cefotaxime (2 g six times daily) were given as empiric antibiotic treatment for the cerebral abscesses.

The patient did not present any symptom or sign of septicemia and her GCA was in remission. However, during the hospital stay she had a progressive worsening of vision in the left eye and amphotericin B treatment was added for presumed mucormycosis.

Stereotaxic aspiration from brain abscesses was performed. Gram stains of purulent material (5 cc) removed during this procedure tested positive for Gram coccus and negative for Gram bacillus. Nevertheless, culture of the purulent material was negative for bacterial and fungal organisms.

Amphoteridn was interrupted and vancomycin (1 g three times daily) was added, showing a progressive disminution of neurological symptom and signs. Four weeks after starting vancomycin therapy, CT scan revealed that the lesions had decreased in size, and 8 weeks later the patient was asymptomatic. CT scan performed at this time showed that the lesions had almost completely disappeared.

Multiple cerebral abscesses and iatrogenic diabetes developed in this patient after 2 months of high dose corticosteroid therapy. A case of fungal brain abscess, which was treated with steroid therapy because the clinical picture mimicked GCA symptoms, has been described (3). Temporal artery biopsy and the excellent response to corticosteroids confirmed GCA in our patient. Cerebral abscesses are a rare complication, which to our knowledge has not been reported as a consequence of steroid therapy in patients with GCA.

The infectious origin in this patient is unknown. Brain abscesses were multiple and localized around the right medium cerebral artery, suggesting a hematogenous dissemination (4). A urine infection was detected, but the patient did not have any symptom or sign of septicemia. However, our patient was diabetic and elderly, and both circumstances might heve masked symptoms of infectious disease. Stereotaxic aspiration demonstrated negative Gram bacillus and positive Gram coccus. Both have been described in immunocompromised hosts (5-7).

In conclusion, we present a patient with GCA and multiple cerebral abscesses which showed a good response to medical treatment. Corticosteroids might have been the main cause of this rare complication.

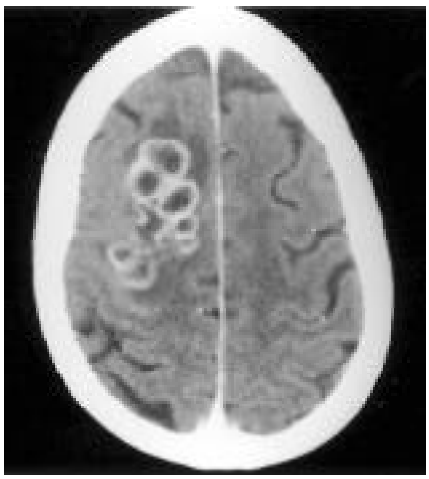

Fig. 1. CT scan with contrast showing multiple images suggestive of brain abscess.

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Juvenile arthritis in Turner's syndrome

Sir,

I was quite interested to read Dr. F. Zulian's article in the Journal (1). The authors described only European and North American patients with Turner's syndrome, however. I would like to add a case of Japanese chronic arthritis (polyarticular type) in Turner's syndrome to your patient group. I believe that this represents the first Japanese case to be reported in the literature. She had 45X/46XX chromosome mosaicism, and the sex-determined region of Y:SRY was negative. The growth plates of both wrist joints were still open at age 22 years.

Our patient's case was complicated by non-insulin dependent diabetes mellitus and Hashimoto's thyroiditis, although these two conditions were well controlled. She first developed arthritis at 14 years of age, with morning stiffness, swelling, and pain in the PIP and wrist joints. Her PIP joints showed progressive deformity over 2 years. She had

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.

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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.

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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