Unusual rhombencephalitis in relapsing polychondritis

Sir: Relapsing polychondritis is a rare disorder, resulting from chronic immunologically mediated inflammation of various cartilaginous structures. The nervous system is affected by a broad range of dysfunctions, as well as idiopathic cerebral spinal pleocytosis, often unresponsive to immunosuppressive treatment.

We report here an unusual presentation of this disease that presented as a diagnostic challenge.

The patient, a 73 year old woman, with an unremarkable history and no treatment presented in February 1990 with a painful redness 3–4 mm in diameter at the base of the nose, which was treated with amoxycillin and clavulanic acid for two weeks without improvement. In mid-March conjunctivitis of the right eye was successfully treated with steroid and antibiotic ointment. At the beginning of April low grade fever, odynophagia, and hoarseness appeared, followed two weeks later by neck pain without meningeal signs, which disappeared with diclofenac treatment. An upper right eyelid paresis and disturbance of the level of consciousness led to hospital admission.

The patient was acutely ill, drowsy, and slow in mentation, but fully orientated. Her temperature was 38 °C. Neurological examination showed a drop of the right eyelid but no other sign of Horner’s syndrome, a slight unilateral facial weakness, conjunctival inflammation of both eyes that was more marked on the right side, and a slight stiffness of the neck without any other meningeal sign. No other physical abnormality was noted.

The erythrocyte sedimentation rate was 140 mm/h, haemoglobin 132 g/l, packed cell volume 0.4, white cell count 25.9×10⁹/l (neutrophils 91%), and platelet count 512×10⁹/l. Results of routine blood analyses were otherwise normal. A lumbar puncture yielded turbid cerebrospinal fluid evocative of bacterial infection (table). Gram staining and tests for pneumococcus, meningococcus, Streptococcus agalactiae group B, and Haemophilus influenzae type b antigens (agglutinins) were negative. All cultures remained sterile. A cerebral computed tomographic scan was normal.

A diagnosis of bacterial rhombencephalitis was considered likely, and amoxycillin 12 g/d was given intravenously. No improvement occurred, but two days later a second lumbar puncture showed a slight improvement of the inflammatory cerebrospinal fluid parameters (table). On the seventh day cartilaginous inflammation developed on one ear, then on the other and on the nose. The diagnosis of polychondritis was retained, and treatment with prednisone 100 mg/d by mouth was started. Fever disappeared immediately, with rapid subsequent general and neurological improvement.

On the 14th day signs of uveitis of the right eye appeared and cyclophosphamide 100 mg/d was added. On the 17th day a sudden left hearing loss developed, leading to complete deafness by the end of May 1990. The patient left the hospital after 38 days, in an improved condition. Cyclophosphamide was reduced to 50 mg/d after one month and prednisone gradually tapered to an alternate day maintenance dose of 5 mg. One year later both treatments were stopped, without recurrence of symptoms. She is now well, 21 months after discharge from hospital, has not required any treatment for nine months, but has not recovered hearing in the left ear.

This case report illustrates a dramatic neurological involvement in relapsing polychondritis that mimicked an infectious disease of bacterial origin. Retrospectively, the history was suggestive of relapsing polychondritis, but the alteration of consciousness prevented the patient from giving an adequate description when admitted to hospital, and the first laboratory investigations were further misleading. The initial cerebrospinal fluid values probably reflected the metabolic consequences of the marked pleocytosis due to the flare of relapsing polychondritis.

This report underscores the importance of early diagnosis and initiation of immunosuppressive treatment, because the condition progresses in flares with risk of important sequelae if specific treatment is delayed, as illustrated here by the hearing loss.

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