Successful application of high dose intravenous immunoglobulins in Sjogren's syndrome associated arthritis

High dose intravenous immunoglobulins (IVIg) have become an important treatment in a number of autoimmune disorders.1 Both controlled and uncontrolled studies have demonstrated beneficial effects of IVIg in rheumatic diseases such as dermatomyositis,2 rheumatoid arthritis,3 and systemic lupus erythematosus.4 We report our experience with IVIg in a patient with Sjogren's syndrome.

A 39 year old man presented for the first time in 1992 with a three year history of painful swelling of both parotid glands without signs of infection. A keratoconjunctivitis sicca which had been confirmed by Schirmer's test required the use of tear substitutes. In addition, the patient had developed an incapacitating, non-erosive arthritis affecting the metacarpophalangeal and proximal interphalangeal (PIP) joints, the knees, and the ankles. Serological evaluations revealed high titres of anti-nuclear antibodies (ANA), anti-SJLa (anti-SJL-A), and anti-La(SB-B) antibodies, and increased titres of rheumatoid factor. Serum IgG was increased to 30.4 g/l without IgG subclass imbalance.5 The patient fulfilled four of the six 1992 criteria and the diagnosis of Sjogren's syndrome.6 Treatment with NSAID was initiated because of persisting arthritis, but without success. Prednisolone was effective in doses up to 30 mg per day, but had to be withdrawn because of signs of iatrogenic Cushing's syndrome, including hyperviscosity and impotence. Treatment with methotrexate was refused by the patient because of the risk of impaired fertility.

We therefore decided to institute IVIg treatment. The patient received 30 g of a sulphonated IVIg (Venimmun®, Behringwerke AG, Marburg, Germany) on each of days 1 to 4 and 21 to 24. Except for one period of headaches, the IVIg were well tolerated and led to a remarkable improvement in the arthritis, as manifested by a reduction in the cumulative PIP joint circumference from 562 mm before treatment to 550 mm at day 30, and a simultaneous increase in grip strength from 80/180 mm Hg (left/right) to 280/300 mm Hg. In addition, a distinct resolution of the patient's parotid gland swelling was noted. Serologically, IVIg treatment resulted in an increase in serum IgG to 56.0 g/l at day 5 and 41.1 g/l at day 30. While titres of ANA, anti-SS-A (anti-La), and anti-La(SB-B) antibodies were not affected, the erythrocyte sedimentation rate decreased from 32 mm/1st h to 5 mm/1st h within 50 days, accompanied by a parallel decrease in C reactive protein from 10 to 8.5 mg/l. The beneficial effects lasted for a total of three months. During month 4, moderate joint pain recurred, but it was controlled adequately by NSAID for the ensuing two months. However, at the end of month 5 a severe relapse of arthritis occurred. IVIg treatment was reinstituted, followed again by a distinct relief of joint swelling and pain.

Our observations suggest a beneficial effect of IVIg in a patient with Sjogren's syndrome associated arthritis, which is one of the most common extradural disease manifestations of this symptom complex.7 Although spontaneous remission cannot be excluded definitely, this seems to be unlikely because the clinical improvement closely followed IVIg administration and was reproducible in a flare of the disease. To our knowledge, this is the first description of the application of high dose IVIg in Sjogren's Syndrome. As many patients with this condition present with increased IgG levels, they are considered at risk of developing symptoms of hypergammaglobulinemia8; however, despite high IgG levels before treatment, no major side effects occurred in our patient. Although our observation in a single patient requires confirmation in others, it does suggest an additive therapeutic option for selected Sjogren's syndrome patients with extradural manifestations in whom conventional treatment fails or is contraindicated.


Asymmetrical nodular osteoarthritis in a patient with a hemiparesis

A 69 year old woman presented to our department complaining of pain and swelling affecting the fingers of the right hand and which had developed during the last 10–15 years. There were no symptoms arising from the left hand, which was affected by a hemiparesis, a consequence of the surgical resection of a brain tumour at the age of 12. On examination, the patient had a left hemiparesis with a mild pyramidal weakness and loss of sensation to light touch. She had Heberden's and Bouchard's nodes affecting all the interphalangeal joints of the right hand, but none on the left (fig 1).

The patient had worked full time as a clerk for 41 years, during which time she was predominantly right handed, but used her left hand for light manual tasks. She gave a very clear description of a similar arthritis affecting her mother who, she recalled, had marked symmetrical deformity of her proximal (PIP) and distal interphalangeal (DIP) joints.

Radiographs confirmed the diagnosis of nodular generalised osteoarthritis (OA) with osteophytic, lipping, sclerosis, and joint space narrowing of the affected joints (fig 2). Serological tests were negative and erythrocyte sedimentation rate was within normal limits for her age.

Osteoarthritis is the commonest joint disorder in Western populations, and the hand is one of the most frequently affected joints.5 In Nodular generalised OA affects the joints of the hand in a symmetrical manner, the most frequent joint groups involved being the DIP and thumb base. In a recent population study,6 the tendency for involvement of nodular generalised OA of the hand will be found to be symmetrical. Asymmetrical OA of the hands has been described occasionally in similar circumstances.9,10 Stecher and Karnosh described a woman with a median nerve injury of the hand who later developed Heberden's nodes in all her fingers except those supplied by the injured nerve.9 Stecher also described it in a hand paralysed by poliomyelitis,4 and in 1935 Coste and Forrestier reported a case that occurred after a cerebral accident.10
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