cornerstone of the therapy for neurosarcoidosis. Steroid therapy is usually started at a high dose and after achieving a clinical response, the dose is gradually tapered. Alternative therapeutic agents are indicated in patients with steroid side-effects or lack of response to treatment or in cases where steroids are contraindicated. These include cyclosporine, azathioprine, hydroxychloroquine, chloroquine and radiation therapy. Chloroquine and hydroxychloroquine have been found to be effective in controlling neurosarcoidosis in patients who fail to respond to corticosteroids or develop serious side-effects, with no evidence of ocular toxicity during the treatment. Clinical manifestations are the best predictors of the course and prognosis in patients with neurosarcoidosis. Cranial neuropathies and aseptic meningitis carry the best prognosis with recovery in up to 90% of cases. Approximately 32% of the patients with neurosarcoidosis, especially those with cranial neuropathies, relapse after the initial neurological episode. Patients with parenchymal disease generally have a prolonged disease course with significant morbidity. Among the peripheral nervous system manifestations, polyradiculitis and acute myopathy tend to respond well to steroids compared to the slowly progressive peripheral neuropathy and myopathy.

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References


Trigger autoimmunity - Development of multiple plexopathy in a patient with chronic idiopathic thrombocytopenic purpura

Sir,

A case of chronic idiopathic thrombocytopenic purpura (ITP) who developed plexopathy, diabetes mellitus and transient disseminated intravascular coagulation (DIC) after splenectomy during hospitalization for the treatment of ITP is presented.

A 42-year-old man diagnosed to be having chronic ITP was admitted for elective splenectomy, for a steroid-responsive but dependent status. His diagnosis was reconfirmed during the preoperative period. He underwent splenectomy. On the third postoperative day, his blood sugar was 348 mg/dl with the presence of urinary ketones, and arterial blood gas (ABG) revealed metabolic acidosis. He was treated with plain porcine insulin, intravenous fluid and electrolytes. His ketoacidosis was controlled with a total of 40 U of plain insulin. During the postoperative period his platelet counts remained at 20-24000/cumm, while peripheral smear revealed fragmented RBCs and thrombocytopenia. Fibrinogen degradation product (FDP) was positive with prolonged prothrombin time (test 21 min; control 13 min). DIC was diagnosed for which he received fresh frozen plasma (FFP) and platelet packs. Multiple blood cultures, urine cultures, cultures from the site of surgery were negative. On the fifth postoperative day he complained of weakness and numbness of the right upper limb. Clinical examination revealed lower motor neuron weakness and wasting of the following muscles: in the upper limb on the right side serratus anterior, pectoralis major, supraspinatus, infraspinatus, latissimus dorsi and teres major while on the left side, muscles of the thenar and hypothenar group, dorsal and palmar interossei, and the lumbricals were involved. He had a loss of all sensory modality over the right forearm, hand, lower 1/3rd of the right arm and the ulnar border of the left hand and forearm. Deep tender reflexes were lost in the right upper limb. Plain radiography, CT thorax and magnetic imaging resonance (MRI) of the spine were normal. CSF study was normal.

Danazol, prednisolone, insulin and physiotherapy were prescribed. At discharge his platelet count was 1,20,000/cumm and perception of sensations and muscle power in both upper limbs was improving. At follow-up after one and a half months, the power also improved, but he required prednisolone and danazol for maintaining a good platelet count.

“Kaleidoscopic autoimmunity” has been reported with various diseases. Our patient developed transient DIC, diabetes mellitus and multiple plexopathy following splenectomy. Multiple plexopathy has a variable presentation. One of the forms is neuralgic amyotrophy (NA) or acute brachial neuritis (ABN) which usually presents as severe pain in the shoulder followed by weakness of shoulder girdle muscles. Moore et al reported a case of lumbosacral plexopathy in a woman with CREST syndrome and vasculitis. The most common mechanism is thought to be viral etiology or immune-mediated. Blood lymphocytes are known to get sensitized to branchial plexus nerves in patients with neuralgic amyotrophy. It is usually unilateral with rare bilateral asymmetrical findings. Sensory symptoms are rare. Sometimes patchy sensory loss may be present. Electrophysiological studies might demonstrate sub-clinical involvement in asymptomatic limbs in up to 25% of patients and very rarely there is a mild lymphocytic pleocytosis or a rise in protein in the CSF. In our patient there was bilateral asymmetrical involvement which is a rare presentation along with weakness without any pain. Our patient also had right common peroneal nerve involvement demonstrated by electrophysiological studies. Prognosis of NA is good with full recovery of strength in 90% of the patients by three years. Plexopathy in our patient was probably immune-mediated. The cause for DIC in our patient was most probably immune-mediated though surgical trauma or diabetic ketoacidosis with severe dehydration contributing to DIC could not be ruled out.

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Multiple sclerosis in a patient with chronic ulcerative colitis

Sir,
We report a patient with chronic ulcerative colitis who developed multiple sclerosis after 5 years of remission from ulcerative colitis.

A 34-year-old lady had recurrent bouts of loose stools mixed with blood and mucus for 8 years and a non-healing ulcer on the lateral aspect of left leg. Investigations revealed ulcerative colitis. Skin biopsy revealed features of pyoderma gangrenosum. She was treated with prednisolone and 5-aminosalicylate following which there was improvement in colonic symptoms and the ulcer over the leg healed. Five years after the treatment the patient complained of diminished vision in both eyes and pain in the orbit on eye movements for 5 days. She was treated with oral prednisolone and showed significant recovery. Three months later, she developed weakness in both lower limbs over 6 days. She was treated with oral steroids and her symptoms ameliorated in 2 weeks. One month after recovery, she was readmitted with complaints of progressive weakness and heaviness in the left upper and lower limb for 15 days. Neurological examination revealed a visual acuity of 6/9 in both eyes and a relative afferent pupillary defect in the left eye. Pronator drift could be elicited in the left upper limb; in the left lower limb the power was Grade 4/5. The deep tendon jerks were Grade 3 in the upper limbs and Grade 4 in the lower limbs bilaterally. Spasticity was demonstrable in the left side with extensor plantar response. MRI brain showed white matter lesions (Figures 1a and 1b). Visual evoked potential was abnormal in both sides. Cerebrospinal fluid (CSF) findings were as follows: cells were absent; protein in the CSF was greater than 12% of the total protein (26.4%). CSF electrophoresis revealed decreased albumin, 35.57% (53.5 – 282.0 mg/dl), serum IgG 1722 mgs/dl (1000 – 2000 mgs/dl) and albumin 3.8 gms/dl (3.5 – 8 gm/dl). The CSF IgG level was greater than 12% of the total protein (26.4%). CSF electrophoresis revealed decreased albumin, 35.57% (53.5 –

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