Sir

Neurological manifestations in sarcoidosis, a multisystem granulomatous disease with enhanced cellular immune process at the site of disease activity, occur in 5% to 6% patients with sarcoidosis.1,3

A 52-year-old male developed left-sided lower motor neuron type of facial nerve palsy, which responded to steroid therapy. Two months later, the patient developed dysarthria, difficulty in swallowing and nasal regurgitation of fluids. He gave history of cough, expectoration and wheezing with seasonal exacerbations, responding to bronchodilators and steroids for the past 30 years. He was treated with long-term steroids for allergic bronchopulmonary aspergillosis (ABPA).

Examination revealed residual left VII nerve paresis with evidence of right-sided IX and X nerve palsy and partial involvement of right XI nerve. There was pansensory loss on the trunk involving the D5 to D8 region on the left side. The ankle reflexes were absent bilaterally with mild impairment of touch, joint position and vibration sensation in the upper and lower limbs in a glove and stocking distribution. Nerve conduction studies revealed evidence of symmetrical axonal sensorimotor neuropathy, predominantly involving the lower limbs. Transbronchial lung biopsy and nerve biopsy of the right sural nerve revealed non-caseating granulomas with giant cell formation consistent with the diagnosis of sarcoidosis (Figures 1 and 2). The patient was initially treated with oral prednisolone in the dose of 1mg/kg/day for 4 weeks followed by gradual tapering to a dose of 10 mg every alternate day. Subsequently, chloroquine at a dose of 150 mg b.i.d/day was added with regular monitoring of visual impairment. The patient showed remarkable clinical improvement in bulbar symptoms and truncal pain within 2 weeks of starting the therapy, with gradual improvement thereafter.

Neurosarcoïdosis can manifest in myriad ways including cranial neuropathy, aseptic meningitis, mass lesions, encephalopathy, vasculopathy, seizures, psychiatric manifestations, hydrocephalus, hypothalamic pituitary disorders, myelopathy, peripheral neuropathy and myopathy.3,4,6 The mode of onset of neurosarcoïdosis is variable, but it is usually subacute to chronic. Acute onset neurological disease usually presents with isolated cranial neuropathies and aseptic meningitis, while patients with a chronic onset usually present with parenchymal involvement, hydrocephalus or peripheral nervous system manifestations.7 Cranial neuropathy is the most common manifestation of neurosarcoïdosis occurring in up to 75% of the patients.9 The facial nerve is the most commonly affected cranial nerve and is involved in up to 50% of neurosarcoïdosis patients.9 Though commonly unilateral, bilateral facial palsy can occur simultaneously or sequentially in approximately one-third of the patients.9 Other cranial nerves, especially the IX and X are less commonly affected.9 The pattern of non-cranial peripheral neuropathy includes patchy neuropathy or mononeuropathy, a component of which may be an “intercostal neuritis” with numb patches on the trunk (as seen in the present case), acute Guillain-Barré syndrome (GBS), and chronic sensorimotor, motor or pure sensory neuropathies.3,11,12 Of these, chronic symmetric axonal sensorimotor polyneuropathy is most commonly observed.12 A combination of multiple cranial and non-cranial neuropathies was also observed in our patient. Sensorimotor neuropathy has been attributed to epineural and perineural granulomas with an associated granulomatous vasculitis, producing an axonal degeneration with associated demyelination.12,31 Although non-caseating granulomas are rarely observed in nerve biopsy samples,12 the sural nerve biopsy in our patient revealed these characteristic changes (Figure 2).

Unlike pulmonary sarcoidosis where a period of observation is recommended for mild and asymptomatic cases, neurosarcoïdosis should always be treated.14 Treatment decisions are governed by disease location, clinical severity, time course and morbidity of treatment. Corticosteroids are the
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


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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 ketoadidosis 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 serratius anterior, pectoralis major, supraspinatus, infraspinatus, latissimus dorsi and teres major while on the left side, muscles of the thenar and hypotenar 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 tendon 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.