Churg-Strauss Syndrome (CSS) Manifesting With Mononeuritis Multiplex Due To The Use Of A Botanical Solution: A Case Report

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Citation


Abstract

Churg Strauss Syndrome (CSS) or allergic granulomatosis is a systemic vasculitic disorder characterized by bronchial asthma, peripheral blood eosinophilia and necrotizing vasculitis involving the medium and small sized vessels with or without granulomas. We present here a case of Churg Strauss Syndrome (CSS) manifesting with mononeuritis multiplex in a 45-year-old male due to the use of a botanical solution.

CASE REPORT

A-45-year old male presented with complaint of numbness in his hands and feet of 2-week duration. He had being apparently well 2 weeks ago when he first noticed numbness in his right thumb and first two digits. He was seen by his primary doctor and a clinical diagnosis of carpal tunnel syndrome was entertained. Over the next 2-weeks he developed numbness on the dorsum and plantar surface of his left foot, followed by numbness in his left hand and finally right foot. On presentation he was weak in his right hand and unable to make a fist. Four days prior to presentation he had developed “black spots” in his right eye and had complaint of blurred vision. Weight loss of 10 pounds over preceding 6 weeks was also documented. Patient had been self treating his allergic rhinitis symptoms with a botanical solution, which he had obtained over the internet through a google online search, at the time of presentation.

Neurological examination revealed a conscious, alert mildly tachypneic man. Right hand grip was 3/5 (Medical research council grading) and he had a right wrist drop. Deep tendon reflexes could not be elicited at the knees, ankles and at the right bicep. There was patchy loss of pain and temperature in no clear dermatomal pattern, joint sense and vibration thresholds were decreased in both the hands and feet (right> left). There was no sensory level on the trunk and plantars were bilaterally down-going. Chest auscultation revealed bilateral expiratory rhonchi.

Unenhanced CT scan of the head revealed a pan-sinusitis (left frontal, bilateral ethamoidal and bilateral maxillary) with air fluid levels. He had an ESR of 62 mm/hr, WBC count of 18.9 with 40% eosinophils. Urine analysis revealed microscopic hematuria and proteinuria. MPO-p ANCA and rheumatoid factor was positive, titer of rheumatoid factor was 1:40. CSF analysis revealed 1 WBC (lymphocyte) with normal glucose and protein. EMG/NCV study revealed a multi-focal axonal neuropathy suggestive of mononeuritis multiplex. Sural nerve biopsy showed epineurial inflammation with mononuclear T cells and necrosis suggestive of vasculitis. He was treated with 1.5mg/kg of prednisone with immediate symptomatic relief of his asthma, resolution of peripheral blood eosinophilia and arrest of neuropathy at the time of this writing.

DISCUSSION

CSS is a systemic vasculitic disorder of presumed allergic/autoimmune etiology characterized by bronchial asthma, peripheral blood eosinophilia and neurological manifestations. The American Academy of Neurology has proposed six criteria for the diagnosis of CSS. The presence of 4 or more criteria yields a sensitivity of 85% and a specificity of 99.7%. These are asthma ( expiratory rhonchi, wheezing), eosinophilia of more than 10% in peripheral blood, para-nasal sinusitis, presence of pulmonary infiltrates, histological proof of vasculitis with extra vascular eosinophils ( lung, renal or nerve) and mononeuritis multiplex or polyneuropathy (, ). Peripheral neuropathy
occurs in relatively large proportion of patients with CSS and may be the presenting symptom of the disease as was in our case (2). Bronchial asthma (respiratory symptoms) may precede the vasculitic phase of the disease by years. When signs and symptoms fully develop, the most prominent are the ones related to pulmonary, cardiac, dermatological, renal and peripheral nerve involvement (2). Rarely patients may present with stroke and other ophthalmologic involvement. The exact etiology of CSS is still not known although an autoimmune etiology is hypothesized due to the presence of hypergammaglobulinemia, elevated IgE levels, rheumatoid factor and ANCA. A CSS like syndrome has been reported as a rare complication in people with asthma who are steroid dependent and treated with leukotriene receptor antagonists (e.g. montelukast and zafirlukast) especially when the steroids are rapidly tapered down. In our patient the syndrome developed about 6 weeks after the use of a botanical solution for presumed upper respiratory symptoms. The herbal solution which he took contained extracts of Mormon Tea (Ephedra trifurca), Licorice Root (Glycyrrhiza glabra), Ginger Root (Zingiber officinale), Eyebright (Euphrasia officinalis), Nettle Root (Urtica dioica), Mullein leaf (Verbascum thapus), Cayenne (Capsicum annum), Bromelain, Quercetin, Citrus bioflavonoids, magnesium aspartate, magnesium ascorbate, magnesium citrate, magnesium gluconate and vitamins B-1, B-2, B-6, B-12 and B-15. Though it is not possible for us to conclusively prove that the use of the herbal remedy was responsible for the development of the syndrome but the temporal profile of the use of the herbal remedy and the patient’s symptoms makes us hypothesize this and thus implicate it as the possible causative agent of CSS in our patient. Alternatively the herbal remedy might have brought out a latent CSS in our patient as the respiratory symptoms preceded his use of the botanical solution. The use of herbal remedies is increasing by the day in the United States. Most of them can be easily obtained over the internet with the click of a mouse and there is precious little FDA control over them. Our case amply illustrates the pressing need for more stringent regulation over the sale and use of these medications.

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