Intracerebral hemorrhage in a patient with Churg-Strauss syndrome

Sir,

A 45-year-old man was admitted with history of painful, sequential multiple peripheral nerve involvement for four months. On questioning he revealed history of allergic rhinitis with recurrent nasal polyps, bronchial asthma for two years, weight loss, joint pains with swelling and Raynaud's phenomenon of four months duration. Examination revealed skin rash and features of mononeuritis multiplex. Investigations showed eosinophilia (absolute eosinophil count-14600/microliter), raised ESR and leucocytosis. The HIV serology, HbsAg, anti-HBc, antinuclear antibody and antineutrophilic cytoplasmic antibody were negative. Serum creatinine was normal and urine examination was unremarkable. Skin biopsy showed evidence of small vessel vasculitis with eosinophilic infiltrate [Figures 1a and 1b]. Nerve biopsy showed evidence

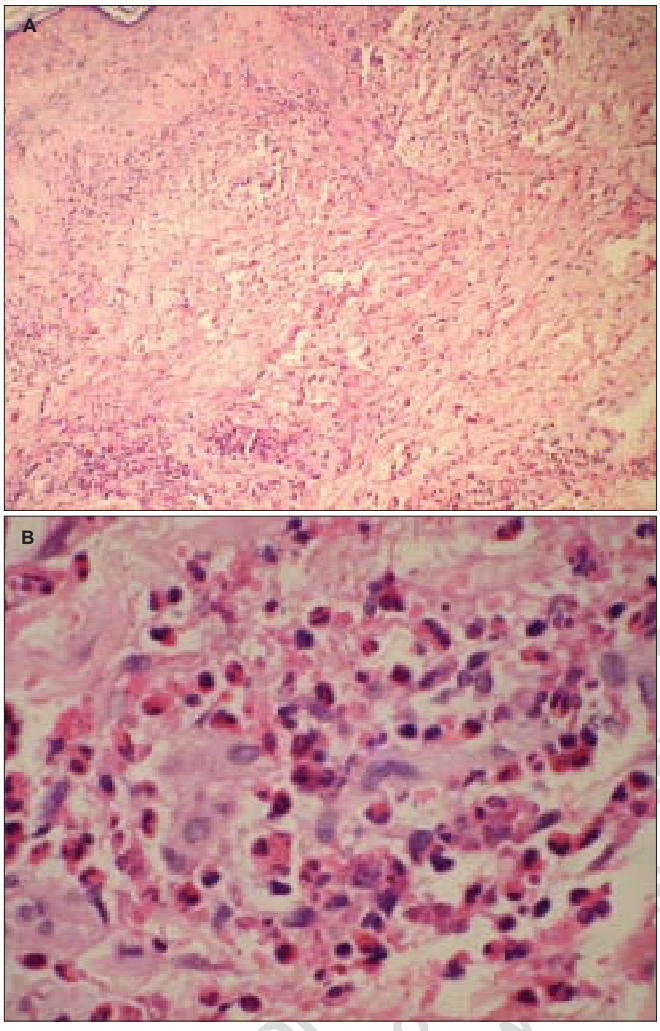

Figure 1: A. Skin biopsy shows normal epidermis but dermis shows marked eosinophilic vasculitis. B. Skin biopsy showing eosinophilic infiltrate surrounding the small vessels

of marked axonal degeneration along with loss of myelin and minimal inflammation. With these features, diagnosis of Churg-Strauss Syndrome (CSS) was made as this patient fulfilled five of the six criteria suggested by the American College of Rheumatology (asthma, eosinophilia, neuropathy, pulmonary infiltrates, paranasal sinus abnormality, extravascular eosinophils on biopsy) and he was started on prednisolone and showed improvement in symptoms. Three months there was worsening of weakness in his upper and lower limbs. Four days before the second admission he developed headache, nuchal pain and drowsiness. His blood pressure was elevated at presentation (210/100). The CT head showed right occipital hemorrhage with intraventricular and subarachnoid extension [Figure 2]. Digital subtraction cerebral angiogram did not reveal any vascular malformation or evidence of vasculitis. He was treated with cyclophosphamide, prednisolone and antihypertensive drugs and showed improvement in weakness, headache and general condition.

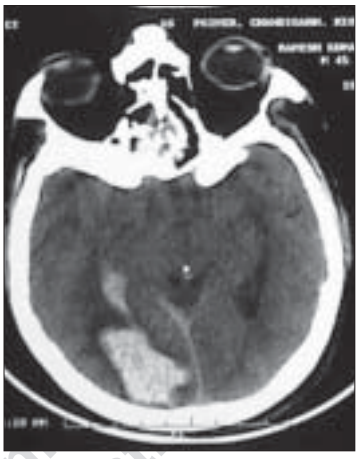

Figure 2: Non-contrast CT head axial section showing right occipital hemorrhage with intraventricular and subarachnoid extension

Neurological involvement is seen in 60-80% cases of CSS.[1,2] Most of the cases, however, have affliction of the peripheral nervous system in the form of mononeuritis multiplex. Involvement of the central nervous system has been reported in 6.3-27.0% of CSS cases.[1,2] Cerebrovascular events have been reported, with a frequency of 6.2-6.4%.[1,2] In the series reported from Mayo clinic,[1] 29 out of 47 patients had neurological involvement. Of this, 17 had multiple mononeuropathy and only three had involvement of the central nervous system in the form of cerebral infarction. No patient had ICH in this series. In another series of 96 patients with CSS,[2] only 8.3% patients had central nervous system involvement in the form of infarcts or cognitive decline. Chang *et al*[3] reported the first case of pathologically documented vasculitis involving the choroid plexus causing massive intraventricular and subarachnoid hemorrhages in CSS. Multiple intracerebral hemorrhages in both hemispheres have been reported in a patient with clinicopathologically diagnosed CSS.[4] Findings on peripheral nerve/muscle biopsy have been reported in 24 cases of CSS by Vital *et al*.[5] Fifteen patients exhibited eosinophils either in extra-vascular infiltrates or in vessel walls and six of them had necrotizing vasculitis. Granulomas were found in only three cases. The clinical diagnosis of CSS was supported by the biopsy findings in 15 out of the 24 patients. However, in our case nerve biopsy had shown axonal degeneration with loss of myelin and only minimal inflammation.

The etiology of intracerebral hemorrhage is not fully understood. A close relationship between hypertension and cerebral hemorrhage in patients with CSS is known and is thought to be the cause of ICH by some authors.[5] However, in patients with SAH and intraventricular hemorrhage, vasculitis has been documented on autopsy or findings consistent with vasculitis have been found on cerebral angiography.[3]

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