A rare initial neurological presentation of Sjogren's syndrome

Sir,

There are only few prior reports of intracranial hypertension syndrome as the presenting symptom of primary Sjogren's syndrome (pSS).[1,2] We describe one such patient. A 38-year-old normotensive woman presented with headache of 18 months duration with increased severity since 2 months accompanied by transient visual obscurations and tinnitus in both ears. There was neither any history to suggest systemic vasculitis, endocrinopathies, migraines in the past nor any longstanding medication intake. Examination revealed body mass index–23.6; pulse–80/minute and regular; blood pressure–120/80 mmHg. Neurologically, her cognition and speech were normal. Fundus examination revealed bilateral papilledema; visual acuity-6/12 in both eyes. Visual field charting revealed enlargement of the blind spot with peripheral visual constriction. There were no other deficits. Investigations revealed normal complete hemogram except the erythrocyte sedimentation rate being 62 mm/hour. Blood calcium, liver function tests, thyroid profile and serum creatinine were in normal range. Serological tests for syphilis and retrovirus were negative. Serum antinuclear antibody (ANA) was positive and ANA profile revealed positivity for antibodies to SS-A and Ro 52. Antiphospholipid antibody workup was negative. Chest X ray and ultrasound examination of abdomen did not reveal any abnormality. Magnetic resonance imaging (MRI) brain including venogram was normal. Nerve conduction studies of limbs were normal. Cerebrospinal fluid examination revealed opening pressure of 38 cm water and its analysis (routine and meningitis workup) was within normal limits. Schirmer's test revealed 6 mm (left eye) and 10 mm (right eye). Lip mucosal biopsy was done and histopathological examination revealed hyperplastic stratified squamous epithelium with spongiosis, lymphocytic exocytosis, upper portions of subepithelial stroma showed dense lymphoplasmacytic infiltration along with melanophages. She was treated with parenteral Methyl prednisolone 1 gram per day for 3 days followed by tapering oral steroids. Acetazolamide 250 mg two tablets three times daily were initiated. Symptoms improved during the hospital stay. Rheumatologist consultation was sought and she was started on Azathioprine 50 mg. At follow-up, the patient was asymptomatic and fundus examination was normal.

Sjogren's syndrome was detected in our patient by the "benign intracranial hypertension" type of initial presentation which is rarely reported in the literature. She had no other systemic symptoms or manifestations of pSS. A report has suggested that central nervous system symptoms can be the initial manifestation of pSS when mild complaints of sicca symptoms are overlooked.[2] No associated conditions linked to benign intracranial hypertension syndrome were seen in our patient. The pathophysiological mechanism of intracranial hypertension in pSS remains unknown. The exact causal mechanism of this increased intracranial pressure is not clearly understood though various mechanisms have been proposed (e.g. venous thrombosis, medication side effects, vasculitis, immune complex precipitation or even direct antibody injury).[1-4] This case expands the spectrum of presentation of pSS that has an important impact in the long-term timely management of such patients.

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