Pseudotumor Cerebri Secondary to Dural Sinus Thrombosis in Neurosarcoidosis

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Pseudotumor cerebri as a presenting manifestation of neurosarcoidosis is rare. This case reported herein had pseudotumor cerebri secondary to dural sinus thrombosis as a presenting feature of neurosarcoidosis without known systemic sarcoidosis. Proper systemic steroid therapy resulted in ocular and systemic recovery. Neurosarcoidosis should be considered in the differential diagnosis of pseudotumor cerebri in patients with unusual neurological findings.

Key Words: Pseudotumor cerebri—Dural sinus thrombosis—Neurosarcoidosis

CASE REPORT

A 35-year-old man presented in 1991 with bilateral papilledema and visual loss in his left eye. He has had severe occipital headaches and convulsions controlled by medical treatment since 1985. But the etiology of these symptoms had not been disclosed. At 2 months before admission he developed horizontal diplopia and left gaze palsy, and it resolved in 2 months. At 3 months prior to admission, polydipsia and polyuria developed.

In neuro-ophthalmologic examination, his visual acuity was 20/20 OD, 20/200 OS, and color vision was 10/12 and 2/12 with Ishiara plates, respectively. An afferent pupillary defect was found in his left eye. Ocular movements were normal, but he had double vision in extreme left gaze. Slit-lamp examination was normal. On ophthalmoscopy both optic discs showed elevation with blurred margins and some hyperemia. The elevation of left optic disc was more apparent. In Goldmann perimetry, the blind spots were found enlarged.

A lumbar puncture (LP) was performed. The cerebrospinal fluid (CSF) was clear and contained slightly elevated protein (62 mg/dl) and decreased glucose (41 mg/dl) levels. The opening pressure of CSF was 340 mm H2O. Repeated LP’s revealed the persistence of elevated CSF pressure. IgG level was elevated in CSF and serum. Microbiologic studies in CSF were negative. Renin angiotensin-converting enzyme (ACE) level was determined to be normal in CSF and serum.

Chest radiography was normal. Gallium-67 scan showed bilateral perihilar involvement. Hypercalcemia (11.4 mg) and high alkaline phosphatase levels were found. Serum antidiuretic enzyme (ADH) level was normal. Mantoux test was negative with 5 IU PPD and Brucella and syphilis tests were also negative. Bronchoscopic examination with bronchoalveolar lavage (BAL) was performed. No pathologic changes were present and cytologic examination of BAL fluid remained normal.

Neurosarcoidosis occurs in approximately 5% of patients with systemic sarcoidosis. Intracranial sarcoidosis may rarely develop in a patient without known systemic disease. The most recognized clinical characteristics are cranial nerve palsies, aseptic meningitis, hypothalamic and pituitary lesions, space-occupying masses, peripheral neuropathy, and spinal cord involvement. We report a patient with neurosarcoidosis presenting the syndrome of pseudotumor cerebri secondary to dural sinus thrombosis and meningeal involvement.

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Computed tomography (CT) revealed left occipital infarct. Magnetic resonance imaging (MRI) of the brain (Fig. 1) demonstrated diffuse leptomeningeal enhancement following GD-diethylene triamine pentaacetic acid (Gd-DTPA). A localized focus of increased T2 signal in the periventricular white matter (Fig. 2) were noted. Pituitary gland enlargement, left occipital and subtemporal infarct areas as well as sagittal sinus thrombosis were visualized. With digital subtraction angiography, the diagnosis of sagittal sinus thrombosis was also confirmed.

The diagnosis of pseudotumor cerebri secondary to sagittal sinus thrombosis with neurosarcoidosis was considered. After systemic steroids (80 mg/day) were given and repeated LPs were performed, papilledema rapidly decreased. The patient’s symptoms improved rapidly, and his color vision ameliorated to normal levels with steroid treatment.

DISCUSSION

Sarcoidosis can cause a variety of neurologic manifestations and pose difficult diagnostic and management problems, especially when the neurologic signs and symptoms are the first presentations (1,2). Pseudotumor cerebri as a presenting feature of neurosarcoidosis is very rare (3,4). This report describes a patient with pseudotumor cerebri and meningeal sarcoidosis. The patient’s investigations showed the evidence of dural sinus thrombosis with Gd-enhanced MRI and digital subtraction angiography confirmed the diagnosis. We believe that dural sinus thrombosis is the mechanism responsible for the increased intracranial pressure in this patient. MRI also demonstrated localized periventricular white matter lesion as well as leptomeningeal involvement and infarct areas. The diagnosis of neurosarcoidosis was considered with these findings. Seizures, abducens palsy, and diabetes insipidus were the other features of this clinical picture. CSF findings and hypercalcemia supported the diagnosis.

Neurosarcoidosis should be considered as a diagnosis in any patient with pseudotumor cerebri who develops unusual neurological findings. Enhanced MRI seems very helpful in demonstrating the full extent of intracranial abnormalities in patients with neurosarcoidosis (5,6).

REFERENCES