**Case Report**

# Pseudotumor Cerebri Syndrome as the Initial Presentation of Primary Sjögren's Syndrome: A Case Report

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Pseudotumor cerebri syndrome comprises intracranial hypertension with usual accompaniment of papilledema without ventriculomegaly, brain tumor, or brain dysfunction.[1,2,3] Primary Sjögren's syndrome is a common autoimmune connective tissue disease that may present with neurologic symptoms as its first manifestations. While central nervous system involvement in primary Sjögren's syndrome is rare,[4] we report a case with pseudotumor cerebri syndrome as the initial presenting symptoms.

**Key words:** pseudotumor cerebri, Sjögren's syndrome, papilledema, intracranial hypertension

## Case Report

A 38-year-old woman complained of subacute progressive worsening of headache accompanied by visual obscuration for one month. She had vertex and occipital pulsatile headache together with nausea, vomiting and photophobia. There were no significant systemic illness, long-term medication use, or history suggestive of autoimmune diseases.

Physical examination revealed an obese female with a body height of 155 cm, a body weight of 105 kg and a body mass index (BMI) of 43.6. Neurologic examination findings were grossly normal except poor visual acuity. There were no cognition, cranial nerve, motor or sensory deficits. Ophthalmic examination revealed poor visual acuity with a bilateral decimal visual acuity of 0.3, increased bilateral intraocular pressure (27 mmHg) and bilateral papilledema.

Serum laboratory investigations including hemogram & electrolytes were normal. Brain MRI, MR angiography and venography revealed widening of sella, compatible with the empty sella sign of intracranial hypertension (Fig. 1) with mild protrusion of the optic nerve. Lumbar puncture revealed an opening pressure of 230 mm $H_2O$, a normal leukocyte count but an elevation of total protein (214 mg/dL). Cerebrospinal fluid (CSF) analyses, including bacterial culture, cryptococcus antigen and tuberculosis DNA, were all normal. Further autoimmune survey revealed an elevated erythrocyte sedimentation rate (ESR) of 71 mm/hr, positive ANA with a titer of 1:1280 with a speckled pattern and positive anti-SSA antibody. Further review of her symptoms revealed mouth dryness before her headache. The Schirmer's test gave a result of 3 mm and

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2 mm in the right and the left eye, respectively. Salivary scintigraphy showed severe xerostomia (Fig. 2).

With the diagnosis of pseudotumor cerebri syndrome, we prescribed acetazolamide for her headache. She was also diagnosed with primary Sjögren's syndrome for which the rheumatologist was consulted.

## Discussion

Pseudotumor cerebri syndrome primarily affects children and adults younger than age 50. The female preponderance manifests after puberty. When no secondary cause is identified, the syndrome is termed idiopathic intracranial hypertension, which most commonly affects women of childbearing age who are obese.[1]

Of all the symptoms of pseudotumor cerebri syndrome, headache is the most common and present in approximately 80% to 90% of patients at diagnosis. Also, it is frequently the initial symptom.[1] Visual symptoms include transient obscurations of vision, diplopia, subjective visual loss and visual distortion. Prolonged elevated intracranial pressure may cause bone erosion at the skull base with a subsequent empty sella and CSF rhinorrhea or otorrhea. Persistent headaches are a major source of morbidity and contribute greatly to a decreased quality of life. With proper diagnosis and treatment, most patients have a good visual outcome, but severe visual loss may still occur in up to 10% of patients.[1]

In 2013, Friedman et al. proposed a set of revised diagnostic criteria for the pseudotumor cerebri syndrome.[2] To diagnose pseudotumor cerebri syndrome based on these criteria, a patient with papilledema should also give normal neurologic examination results except for cranial nerve abnormalities, normal brain parenchyma without evidence of hydrocephalus, mass, or structural lesion and no abnormal meningeal enhancement on neuroimaging studies, normal CSF composition and elevated lumbar puncture opening pressure ($\geq$ 250 mm $H_2O$ in adults in a properly performed lumbar puncture). The diagnosis of pseudotumor cerebri syndrome is definite if the patient fulfills all criteria above. The diagnosis is considered probable if all criteria above are met but the measured CSF pressure is lower than that specified for a definite diagnosis.

Pseudotumor cerebri syndrome can be subdivided into either primary or secondary. Primary pseudotumor cerebri includes idiopathic intracranial hypertension,[2] while secondary pseudotumor cerebri syndrome can be further categorized into one (a) that causes cerebral venous hypertension such as venous sinus stenosis or thrombosis, (b) that causes changes in CSF composition such as infectious leptomeningitis and other high CSF protein conditions, and (c) that is assumed to cause

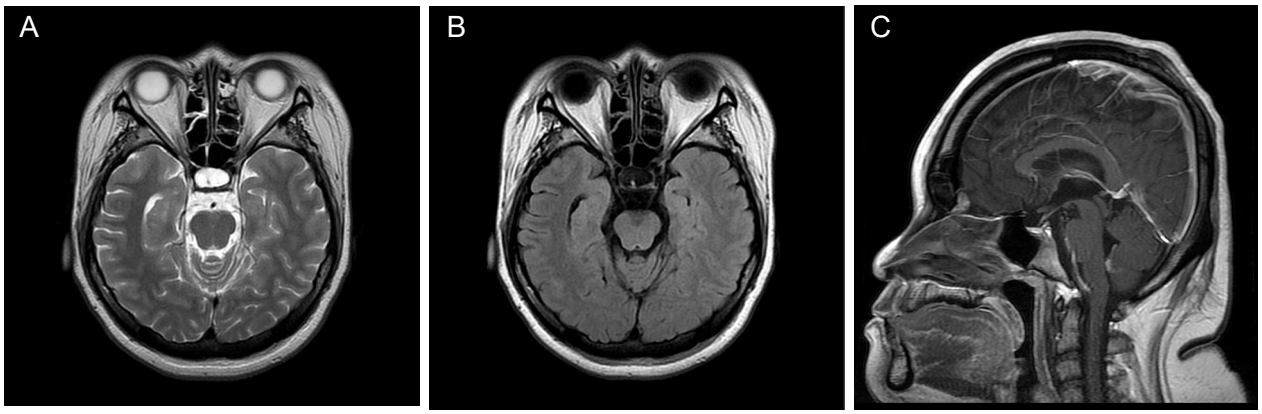


*Fig. 1 Empty sella sign of intracranial hypertension: (A) T2-weighted axial MRI (B) T2-weighted fluid-attenuated inversion recovery axial MRI (C) postcontrast T1-weighted sagittal MRI.*

high arachnoid villous resistance such as hypercapnic respiratory failure, vitamin A excess, corticosteroids or tetracyclines exposure.[3]

Sjögren's syndrome is one of the more prevalent autoimmune inflammatory disorder that primarily affects connective tissues. Neurologic manifestations from the involvement of both peripheral and a smaller part the central nervous system may also be significant. The estimated prevalence of neurologic symptoms is about 8.5% – 70% among patients diagnosed with primary Sjögren's syndrome.[4]

Albeit rare, patients with Sjögren's syndrome who had psendotumor cerebri syndrome as the sole initial presentiation have been previously reported.[5,6,7] In our case, ophthalmic examination revealed papilledema. Neurologic examination findings were grossly normal. Brain MRI also showed normal brain parenchyma without hydrocephalus. On the other hand, a widened sella and mild intraocular protrusion of the optic nerve could be clues to the diagnosis of pseudotumor cerebri.[2] In our patient, CSF analysis showed normal composition except for the elevation of total protein which was observed in primary Sjögren's syndrome patients with active central nervous system complications.[8] The lumbar puncture opening pressure was 230 mm $H_2O$ (< 250 mm $H_2O$). Based on these findings, the diagnosis of pseudotumor cerebri syndrome was considered probable though not definite. Together with the symptom of dry mouth, positive anti-SSA antibody, positive Schirmer's test (< 5 mm in 5 minutes) and salivary scintigraphy revealing severe xerostomia, primary Sjögren's syndrome was diagnosed.

The pathophysiological mechanism of pseudotumor cerebri syndrome in primary Sjögren's syndrome remains unclear. In cases associated with rheumatic diseases, the possible mechanism of increased intracranial pressure includes cerebral venous thrombosis, medication side effects, vasculitis, immune complex precipitation and direct antibody injury.[5] CSF hyperproteinemia is also believed to block arachnoid CSF drainage, thereby elevating CSF pressure.[9] In our case, magnetic resonance imaging showed no venous thrombosis and there was no medication-related side effect, but elevation of CSF protein was noted. CSF hyperproteinemia and other immune responses in primary Sjögren's syndrome might be the causes of pseudotumor cerebri syndrome.

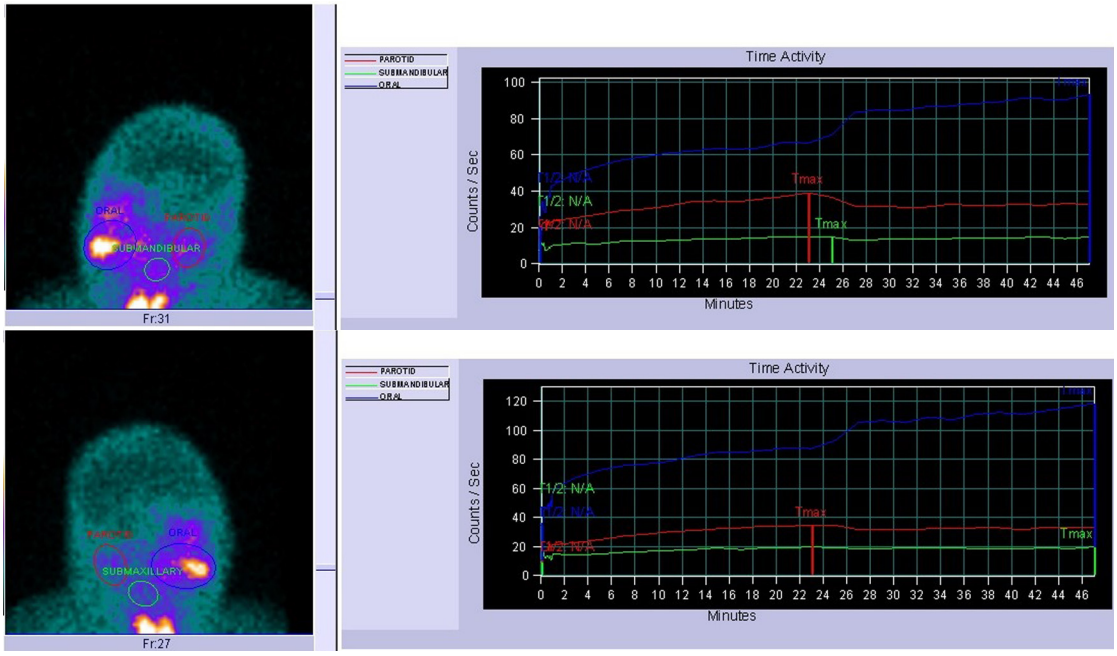


*Fig. 2 Salivary scintigraphy showing severe xerostomia.*

## Conclusion

Considering the risk of neurological deficits including severe visual loss, appropriate diagnosis of pseudotumor cerebri syndrome is essential. Even in the obese women of childbearing age who are most commonly affected by idiopathic intracranial hypertension, primary Sjögren's syndrome should be considered in the differential diagnosis of pseudotumor cerebri syndrome.

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