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# Idiopathic Intracranial Hypertension as an Initial Presentation of Sjögren's Syndrome in Medicine

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**Abstract**

It is uncommon and sometimes misdiagnosed for primary Sjogren's syndrome (pSS) to impact the central nervous system. A highly unusual occurrence is Sjogren's syndrome coupled with concurrent intracranial hypertension. The existence of elevated intracranial pressure in MS patients might be challenging to identify since the symptoms of visual impairment in the two illnesses can be comparable. A severe headache in an MS patient may be an early indicator of the development of intracranial hypertension. Making the right diagnosis requires being able to distinguish between minor variations in how visual acuity, visual field abnormalities, and subjective symptoms are presented. Injurious and irreversible loss of vision may come from failing to recognise and manage rising intracranial pressure.

**Keywords:** Sjögren's syndrome, Idiopathic intracranial hypertension, Papilloedema, Headache.

## 1. Introduction

Raised intracranial pressure without any discernible brain disease is the hallmark of idiopathic intracranial hypertension. The composition of cerebrospinal fluid (CSF) is typical. The most typical symptom is a headache. Papilloedema is the primary symptom of idiopathic intracranial hypertension (1).

Although central nervous system involvement in primary sjogren's disease is rare, peripheral nervous system (PNS) involvement is widely established.

Symptomatic brain lesions, meningitis, myelopathy, and cranial neuronopathy are examples of nervous system involvement (2). Here, we present a case of primary Sjogren's disease in which the first symptom was idiopathic intracranial hypertension.

The lupus erythematosus (SLE) and Sjogren's syndrome may be related. Additionally, some individuals with long-standing Sjogren's disease may acquire systemic symptoms that meet the 1982 criteria for SLE established by the American Rheumatism Association. Here, we present the case of a Sjogren's disease patient also experienced symptoms of intracranial hypertension Sjogren's syndrome, which led to a secondary diagnosis of SLE. Increased intracranial pressure without a growing tumour or hydrocephalus is known as intracranial hypertension Sjogren's syndrome.

This neuro-ophthalmic presentation of SLE is an uncommon occurrence. It is unclear what is causing this elevated intracranial pressure. Our patient did not have the dural sinus thrombosis that other researchers have seen in SLE patients who also had intracranial hypertension.

## 2. Case

A 38-year-old lady with normal blood pressure reported having headaches and blurred vision for a month along with brief visual obscurations.

There was no historical medical history indicating systemic vasculitis, endocrinopathies, migraines, or chronic drug use. Examination results showed a body mass index of 24.6; a consistent 82 beats per minute pulse; and a blood pressure of 120/80 mmHg. Bilateral papilledema was found during a fundus examination, and both eyes' visual acuities were 6/12. The blind spot has become larger with peripheral vision constriction, according to perimetry. Other deficiencies were nonexistent. Investigations showed that the whole hemogram was normal with the exception of the erythrocyte sedimentation rate, which was 60 mm/hour. Tests for liver function, thyroid function, serum creatinine, and blood calcium all fell within the normal range. Retrovirus and syphilis serological testing came out negative.

Serum antinuclear antibody (ANA) was positive (1+), and the ANA profile showed antibodies to SS-A and Ro 52 were also positive. Workup for antiphospholipid antibodies was negative. An ultrasound of the abdomen and an X-ray of the chest did not identify any anomalies. Venogram results from magnetic resonance imaging (MRI) of the brain were normal. Studies on the nerve conduction in the limbs were normal. Examining the cerebrospinal fluid indicated an opening pressure of 42 cm of water, and its routine and meningitis workup analyses were within normal bounds. 6 mm (left eye) and 10 mm (right eye) were found using Schirmer's test.

A little sample of the salivary glands revealed lymphocytic salivarianitis.

Two 250 mg acetazolamide pills taken three times a day were started. The hospital stay helped the symptoms become better. She had counselling from a rheumatologist and was started on 50 mg of azathioprine. The patient had no symptoms at the follow-up appointment, and the fundus exam was unremarkable.

## 3. Discussion

Increased intracranial pressure and papilledema, which are not accompanied by localised neurological symptoms, are the key characteristics of intracranial hypertension Sjogren’s syndrome. Idiopathic intracranial hypertension Sjogren’s syndrome is the most common diagnosis. Since intracranial hypertension Sjogren’s syndrome is a very uncommon manifestation of rheumatological illnesses, it is simple to ignore. However, there are case reports that imply the main sjogren's disease may appear with CNS involvement at first. As in this instance, the diagnosis of Sjogren's syndrome was taken into consideration due to the existence of bilateral papilledema without any discernible disease in the brain and with congruent laboratory and pathology results consistent with primary Sjogrens syndrome. After receiving steroid therapy, the symptoms disappeared, suggesting that the intracranial hypertension was perhaps related to the original sjogren's syndrome's aberrant activity.

It is still unclear what causes primary sjogren's syndrome's intracranial hypertension. Cerebral venous thrombosis has been implicated in a number of cases of intracranial hypertension linked to rheumatological illnesses (3,4). It was thought that the patients' lupus anticoagulant or anticardiolipin antibodies would cause a hypercoagulable condition (3,9). Vasculitis, immune complex precipitation, or even direct antibody damage may be connected to another mechanism (10). The patient's tests for lupus anticoagulant and serum anticardiolipin antibody came back negative, and the MRI scans showed no signs of venous sinus thrombosis.

Increased intracranial pressure and papilloedema are symptoms of intracranial hypertension Sjogren’s syndrome, which is unrelated to specific neurological symptoms. To diagnose this Sjogren’s syndrome, four criteria must be met: elevated cerebrospinal fluid pressure exceeding 200 mm H20, normal cerebrospinal fluid cellular and biochemical composition, symptoms and signs limited to those of elevated intracranial pressure, and normal radiographic findings. The intracranial hypertension condition in our patient is not likely to be considered a symptom of Sjogren's illness. In spite of the fact that it occurs seldom during SLE, intracranial hypertension Sjogren’s syndrome has not been described as a neurological symptom in Sjogren's disease.

It is relatively uncommon for SLE to appear as intracranial hypertension Sjogren's syndrome. Although the majority of instances with intracranial hypertension Sjogren's syndrome are idiopathic, there have been other illnesses linked to it. In a review of published work, The SLE itself may lead to anaemia and hypertension. Blood pressure was within normal range in our patient, and the anaemia was not severe enough to account for the intracranial hypertension condition. The intracranial hypertension Sjogren's syndrome associated with SLE may potentially be influenced by corticosteroids. But in this case, the patient got the same amount of 10 mg prednisone before being admitted to the hospital.

It might be challenging to identify elevated since individuals with both increased intracranial pressure and those with MS may experience visual symptoms and signs (Table 1).

**Table 1. Comparing MS's visual symptoms and signs to elevated intracranial pressure.**

| | MS | ICP |
|---|---|---|
| Vision Clarity | reduced in the afflicted eye | kept until quite late |
| Difference Awareness | Decreased | Decreased |
| Afferent Pupillary Defect in Relation | existent | Unless asymmetric PE, nonexistent. |
| deficiencies in the visual field | Generalised depression first manifests as nerve fibre bundle abnormalities, such as central and cecocentral scotomas, arcuate defects, and altitudinal defects. | Early stages of normal visual fields; larger blind patches, constriction, and defects of nerve fibre bundles, including arcuate faults sparing the centre region |
| Optic Nerve Head Appearance | 1/3 optic nerve head edoema without haemorrhages, 2/3 retrobulbar initially normal | Bilateral optic nerve head edema (absent in only ~ 10% ) with hemorrhage(s) depending on severity |

Both disorders frequently involve headaches. More than 50% of MS patients report having regular headaches, which are often migraines or tension headaches. An ongoing symptom of elevated intracranial pressure is headaches. Idiopathic intracranial hypertension is the model condition that exhibits elevated intracranial pressure in this age group distribution. Although they are vague, headaches are a defining characteristic of this illness. Since the incidence of migraine is said to be higher in both circumstances, the kind of headache may not be able to distinguish between them. However, the existence of pulsatile tinnitus can be used to make a distinction. This condition, which is not normally connected to MS, is seen in 58% of IIH patients. Three women with known Multiple Sclerosis and persistent headaches were the subjects of an investigation by Newman et al. Each time a lumbar puncture was performed, there was evidence of bilateral papilledema linked to elevated intracranial pressure.

Two of the patients in our series showed no papilledema on funduscopic or OCT. The most prevalent ocular motor finding in IIH is horizontal diplopia, whether it is transitory or continuous. Other erroneous localising cranial nerve symptoms, such sensory alterations in the face, may also appear. Multiple cranial nerve deficits, most typically affecting cranial nerves 3, 4, and 6, may manifest in MS. Although it is uncommon to have internuclear ophthalmoplegia with high pressure, it is the characteristic symptom.

Recent developments in diagnostic radiology have greatly improved the capacity to detect people with elevated intracranial pressure using both MRI and MRV. These methods locate the localised lesions that cause changes in cerebral dynamics, but they also frequently locate modest indications of increasing pressure. About 90% of time, greater pressure will be present if the optic nerve changes and the sella is empty. Last but not least, new imaging results have shown localised constriction of the dural venous sinuses. Farb reports that there is a more than 90% incidence of focal constriction using 3D gadolinium enhanced venography.

The disorder known as idiopathic intracranial hypertension is frequent elevated group where young women outnumber males. The Dandy inclusionary criterion are broken when localised abnormalities are present. If causative, it may reasonably be categorised as Digre's pseudotumor type two.

Deficits in the visual field might have overlapping patterns. The most typical first visual field loss. In 37% of damaged eyes, all vision is lost respectively, there are central and centrocecal impairments. Flaws are examples of localized defects that are less frequent. In contrast, localized anomalies fibre are responsible of the visual field observed in intracranial hypertension patients. The most frequent of these, arising from persistent pressure on the optic nerve, are larger blind spots and arcuate bundle abnormalities.

This unusual instance of primary Sjogren's syndrome exacerbated by intracranial hypertension syndrome demonstrates the need of taking Sjogren's syndrome into account when making a diagnosis of intracranial hypertension Sjogren’s syndrome.

## 4. Conclusion

As symptoms of visual impairment may be identical in both illnesses, it can be challenging to recognise increased intracranial pressure in MS patients. In MS patients, persistent headache may be a symptom of developing intracranial hypertension. Establishing a proper diagnosis requires the understanding of minute variations in how visual acuity, visual field abnormalities, and subjective symptoms are presented. An adverse and irreversible loss of vision may occur if the elevated intracranial pressure is not identified and treated.

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