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Hypertrophic Pachymeningitis (HP)-A Rare presentation of Sjogren's Syndrome

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ABSTRACT

Inflammation and thickening of the dura are symptoms of Hypertrophic Pachymeningitis (HP). There are several different causes of HP, ranging from idiopathic to secondary causes include infections, autoimmune disorders, malignancies, and connective tissue disorders. The Sjogren's syndrome occasionally presents with HP. This case study demonstrates that Sjogren's syndrome can produce HP.

Key words: Inflammation, Connective tissue disease, Sjogren's syndrome

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INTRODUCTION

Inflammation and thickening of the dura mater are referred to as hypertrophic Pachymeningitis (HP), which can be idiopathic or related to a wide range of diseases. Clinically, HP can cause headache and cranial nerve abnormalities, although it can also go unnoticed for years. HP may coexist with underlying conditions as infections, cancers, and autoimmune disorders [1]. The most frequent cause of autoimmune disease-related HP is IgG 4/multifocal fibro sclerosis, followed by ANCA-related (microscopic polyangiitis and granulomatosis with polyangiitis) HP [1,2]. There have also been reports of correlations with relapsing polychondritis, giant cell arteritis, sarcoidosis, mixed connective tissue disease, and undifferentiated connective tissue disease [3-6]. HP in Sjogren's syndrome (SS) is uncommon, albeit [2]. We present the example of a patient with primary SS and HP.

CASE REPORT

A 60-year-old man who had been complaining of headaches for two months and an intermittent fever for a month was admitted to this hospital. The headache was diffuse and of a moderate intensity. There were no chills or rigors and the fever was low grade and sporadic. No history of seizures, diplopia, vomiting, weakness, coughing, expectoration, hearing loss, or arthralgia. On examination, the temperature in the axilla was 38.6°C. Vitals in the submandibular and anterior triangle, there were 4-5 distinct, non-tenders, mobile, and firm cervical lymph nodes. Examination of the heart, lungs, abdomen, and nerve system did not reveal any abnormality. Meningeal signs were not present. The ENT exam was normal. Laboratory investigations revealed as follows (Table 1).

The CSF study revealed; TLC 14 cells/HPF, all was lymphocytes. CSF sugar- 48 mg/dL, Protein 50 mg/dL, 15 mg/dL of IgG, and IgG index of 1.09 (0.70). Mycobacterium tuberculosis, bacteria, and any fungi were not identified in CSF cultures. CSF Cytology found no indication of cancer. Thus, the CSF examination was unable to identify the headache's underlying cause. A thicker, abnormally enhanced dura mater seen on a brain MRI after gadolinium contrast suggested Hypertrophic Pachymeningitis (HP) (Figure 1).

Table 1: Investigations of the patient.

Investigations	Patient's values	Reference values
Haemoglobin (Hb)	12.6 g/dL	12.2-14.6 g/dl
White blood cells	8,400/µL	6,000-11,000/ µL
Platelet count	2,25,000/µL	1,50,000-4,50,000/µL
C-reactive protein (CRP)	6.8 mg/dL	<1mg/dl
Blood urea	16 mg/dl	9-20mg/dl
Serum Creatinine	0.8 mg/dl	0.6-1.2 mg/dl
Random Blood Glucose	94 mg/ dl	80- 120 mg/dl
Anti-SS-A antibodies	498AU/ml	<100AU/ml
Anti-SS-B antibodies	134AU/ml	<100AU/ml
anti-neuronal cell antibodies	178 U/ mL	<18 U/ml

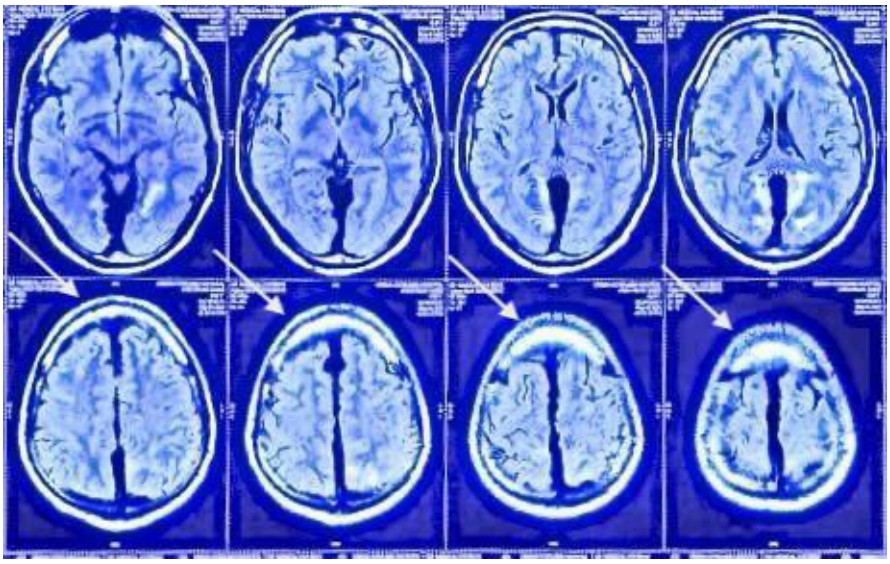

Figure 1: MRI Brain showing thickened and abnormally enhanced dura mater (white arrows).

Supplementary investigations to investigate underlying disease were done as follows IgG, 2612 mg/dL; IgG4, 25.8 mg/dL; anti-nuclear antibodies, 1,2350 (speckled); anti-SS-A antibodies, 498; anti-SS-B antibodies, 134; and anti-neuronal cell antibodies, 178 U/ mL (18 U/ mL). Tests for ds-DNA antibodies, PR3-ANCA antibodies, MPO-ANCA antibodies, ribonucleoprotein antibodies, RNA-polymerase III antibodies, cardiolipin antibodies, and anti-NR2 antibodies came back negative. Anti-NR2 antibodies were also sent for CSF, and it was discovered that they were positive, measuring 2.56 U/mL compared to a normal range of 0.27 U/ml. Anti-neuronal antibody levels in CSF were also elevated, coming up at 0.19 U/ mL compared to the expected 0.1 U/ml. There was a suspicion of Sjogren's syndrome. In order to confirm Sjogren's syndrome, more tests were carried out. An eye examination was performed. Where both the fluorescein eye stain test and the Schirmer's test came out positive. The gum-chewing test was positive. Salivary scintigraphy was performed to check for involvement of the salivary glands and revealed no abnormalities.

The patient met the requirements for a Sjogren's Syndrome diagnosis as per the 1999 revisions to the criteria published by the Japanese Ministry of Health and Welfare. Schirmer's test, fluorescein staining test, and anti-SS-A and anti-SS-B antibody results are all positive [7]. Therefore, a diagnosis of linked Hypertrophic Pachymeningitis to Sjogren's syndrome was determined based on the aforementioned findings.

Prednisolone 40 mg/day or 0.8 mg/kg was administered orally to the patient. The therapy had a good effect on the patient. Five days after starting prednisolone medication, the patient's symptoms, including fever and lymphadenopathy, subsided. His headache issue also vanished ten days later. Following treatment, a repeat MRI revealed a reduction in dura thickness. Within two to three weeks, oral corticosteroids were gradually decreased to 10 mg/day.

DISCUSSION

The term Hypertrophic Pachymeningitis (HP) refers to inflammation and dura mater thickening. HP has a wide range of systemic inflammatory disorders that are linked to it, including sarcoidosis, Wegener's granulomatosis, Sjogren's syndrome, rheumatoid arthritis, and IgG4-related disease [1-3,8]. Syphilis, Lyme disease, and tuberculosis are examples of infectious causes [9,10]. It is important to distinguish between actual neoplastic invasion and dural thickening, which is considered as a reactive alteration in response to a tumour in the bone beneath. It could also be the result of intracranial hypotension, which is probably brought on by compensatory meningeal interstitial edema in response to a reduced CSF space volume [11]. Idiopathic HP is the term used to describe a state when no underlying disease mechanism has been found.

The symptoms of cranial nerve deficits and headache were the most common HP presentations. Idiopathic HP can also appear with headache and cranial nerve dysfunction [1]. Although practically all cranial nerves have been shown to be impacted, the vestibulocochlear nerve has been reported to be the most frequently affected cranial nerve deficit in idiopathic HP [1,12,13].

Initial MRI findings that indicated dural thickness and enhancement helped to diagnosis HP. There was a lot of variance in the specific enhancement patterns, such as nodular, regular, or irregular, within these modifications. In cases of idiopathic HP, the posterior fossa was frequently afflicted, but generally, placement was highly diverse. The occipital/temporal lobe, the spinal column, the posterior fossa, and the tentorium are also involved. Also possible is focal dura involvement.

In terms of treatment, steroid medication is typically used as the first line of protection against idiopathic or autoimmunity-related HP. Additional immunosuppressant therapy may be necessary in some

cases of idiopathic HP, as well as ANCA- and IgG4-related HP. Steroid medication was used to successfully treat the current case, preventing a recurrence. According to Nakano et al., treating HP in SS may only require moderate doses of steroids [14]. Aseptic meningitis is another condition of the central nervous system connected to Sjogren's syndrome in addition to hypertrophic Pachymeningitis. To differentiate between meningitis and HP, contrast-enhanced MRI is required, and, if practical, a dural biopsy should be carried out.

CONCLUSION

Several autoimmune and viral diseases have been linked to Hypertrophic Pachymeningitis (HP), however one possible cause of hypertrophic Pachymeningitis is Sjogren's syndrome. According to this case study, when a patient presents with Hypertrophic Pachymeningitis (HP), Sjogren's Syndrome is also to be considered though it is rare.

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