

IgG4-Related Hypertrophic Pachymeningitis Presenting with Features of Intracranial Hypertension: A Rare Case Report

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Abstract

Background:

IgG4-related disease (IgG4-RD) is a systemic fibroinflammatory disorder that can involve virtually any organ system. Hypertrophic pachymeningitis is a rare neurological manifestation characterized by localized or diffuse dural thickening and enhancement. Early diagnosis is essential because the disease responds well to immunosuppressive therapy.

Case Presentation:

We report the case of a 40-year-old male who presented with symptoms suggestive of raised intracranial pressure. Magnetic resonance imaging (MRI) of the brain revealed diffuse pachymeningeal thickening and enhancement involving the cerebral convexities, falx cerebri, tentorium, and cavernous sinus regions. Initial laboratory evaluation demonstrated elevated serum IgG4 levels (approximately 5 g/L). Extensive investigations excluded infectious, neoplastic, and other autoimmune etiologies. Based on the clinical, radiological, and serological findings, a diagnosis of IgG4-related hypertrophic pachymeningitis was established.

Management and Outcome:

The patient was treated with corticosteroid therapy, resulting in significant clinical improvement and normalization of serum IgG4 levels on follow-up.

Conclusion:

IgG4-related hypertrophic pachymeningitis should be considered in patients presenting with unexplained dural thickening and features of intracranial hypertension. Prompt diagnosis and treatment with corticosteroids can result in excellent clinical outcomes.

Keywords:

IgG4-related disease, hypertrophic pachymeningitis, intracranial hypertension, pachymeningeal enhancement, corticosteroids.

Introduction

Hypertrophic pachymeningitis (HP) is an uncommon inflammatory disorder characterized by localized or diffuse thickening of the dura mater. The disease may occur secondary to infections, autoimmune disorders, neoplasms, or systemic inflammatory conditions. Among the autoimmune causes, IgG4-related disease (IgG4-RD) has emerged as an important and increasingly recognized etiology.

IgG4-RD is a chronic immune-mediated fibroinflammatory disease characterized by tumefactive lesions, lymphoplasmacytic infiltration rich in IgG4-positive plasma cells, and often elevated serum IgG4 concentrations. Neurological involvement is uncommon and most frequently presents as hypertrophic pachymeningitis. Because of its rarity and varied presentation, diagnosis is often delayed.

We report a rare case of IgG4-related hypertrophic pachymeningitis presenting with features of intracranial hypertension, which demonstrated an excellent response to corticosteroid therapy.

Case Report

A 40-year-old male presented with complaints of persistent headache of several weeks' duration, associated with symptoms suggestive of raised intracranial pressure. There was no history of fever, seizures, cranial nerve palsies, connective tissue disease, tuberculosis, malignancy, or previous neurosurgical intervention.

On examination, the patient was conscious, oriented, and hemodynamically stable. Neurological examination revealed clinical features suggestive of intracranial hypertension without focal neurological deficits.

Considering the persistent symptoms, a detailed radiological and laboratory evaluation was performed.

Investigations

Hematological and Biochemical Parameters

- * Hemoglobin: 15.0 g/dL
- * Total leukocyte count: 9,800/mm³
- * Platelet count: 392,000/mm³
- * ESR: 13 mm/hour
- * HbA1c: 5.65%

Thyroid Profile

- * Free T3: 2.64 pg/mL
- * Free T4: 1.31 ng/dL
- * TSH: 4.66 μ IU/mL

Cerebrospinal Fluid Analysis

Lumbar puncture revealed:

- * CSF protein: 30.35 mg/dL
- * CSF glucose: 75 mg/dL
- * CSF culture: No growth
- * Gram stain: Negative

No evidence of infectious meningitis was identified.

Immunological Evaluation

Serum IgG4 concentration was found to be elevated, measuring approximately 5 g/L, supporting the diagnosis of IgG4-related disease.

Neuroimaging Findings

MRI Brain with Contrast

MRI demonstrated extensive sheet-like and nodular pachymeningeal enhancement involving:

- * Bilateral cerebral convexities
- * Falx cerebri
- * Bilateral temporal regions
- * Bilateral cavernous sinuses
- * Superior tentorial leaflets

Additional findings suggestive of intracranial hypertension included:

- * Bilateral optic nerve tortuosity
- * Prominent perioptic cerebrospinal fluid spaces
- * Posterior globe flattening
- * Partial empty sella

Magnetic resonance venography excluded cerebral venous sinus thrombosis.

Management and Outcome

The patient was diagnosed with IgG4-related hypertrophic pachymeningitis and initiated on corticosteroid therapy.

Treatment included:

- * Intravenous corticosteroids followed by oral prednisolone taper
- * Symptomatic management of headache
- * Measures to reduce intracranial pressure
- * Regular clinical and biochemical follow-up

The patient demonstrated significant symptomatic improvement following corticosteroid therapy. Follow-up laboratory evaluation revealed normalization of serum IgG4 levels, indicating an excellent therapeutic response. Clinical symptoms resolved substantially, supporting the diagnosis of IgG4-related hypertrophic pachymeningitis.

Discussion

IgG4-related disease is a systemic fibroinflammatory disorder that can affect multiple organ systems, including the pancreas, salivary glands, kidneys, retroperitoneum, and central nervous system. Hypertrophic pachymeningitis represents one of the rare neurological manifestations of this disease.

Patients commonly present with headache, cranial neuropathies, visual disturbances, seizures, or symptoms of raised intracranial pressure. MRI with gadolinium contrast remains the imaging modality of choice and typically demonstrates diffuse or focal dural thickening with intense enhancement.

The diagnosis of IgG4-related hypertrophic pachymeningitis relies on a combination of:

- * Characteristic clinical presentation
- * Typical neuroimaging findings
- * Elevated serum IgG4 levels

- * Exclusion of alternative etiologies
- * Favorable response to corticosteroid therapy

Our patient had extensive pachymeningeal involvement with radiological features of intracranial hypertension. The significantly elevated serum IgG4 level (~5 g/L) and subsequent normalization following corticosteroid treatment strongly supported the diagnosis of IgG4-related hypertrophic pachymeningitis.

Corticosteroids remain the first-line treatment and usually produce rapid clinical and radiological improvement. However, relapse can occur, necessitating long-term follow-up and, in some cases, steroid-sparing immunosuppressive agents such as azathioprine, rituximab, or mycophenolate mofetil.

Conclusion

IgG4-related hypertrophic pachymeningitis is a rare but treatable cause of chronic headache and intracranial hypertension. Elevated serum IgG4 levels combined with characteristic MRI findings and an excellent response to corticosteroid therapy can establish the diagnosis. Early recognition and prompt treatment are essential to prevent irreversible neurological complications and improve patient outcomes.

References

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