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Review Article

Hypertrophic Pachymeningitis: A Literature Review

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ABSTRACT

Hypertrophic pachymeningitis is a condition characterized by the thickening of dura mate, where tough outer membrane surrounding brain and spinal cord thickens due to ongoing inflammation, resulting in scarring. There are two main types of hypertrophic pachymeningitis: idiopathic hypertrophic pachymeningitis (IHP) and secondary hypertrophic pachymeningitis (SHP). IHP occurs without a known cause and is believed to involve an abnormal immune response. On the other hand, SHP is associated with specific underlying conditions such as autoimmune diseases. Common symptoms include headaches, neurological problems, and signs of increased pressure within the brain. To diagnose hypertrophic pachymeningitis, doctors typically use imaging tests such as MRI and CT scans, as well as a tissue biopsy to confirm the condition.

INTRODUCTION

Hypertrophic pachymeningitis is a rare inflammatory condition characterized by the thickening of dura mater, outermost layer of meninges that surrounds brain and spinal cord. This thickening results from chronic inflammation and fibrosis of dura mater, leading to various neurological symptoms. The condition can be idiopathic or secondary to other underlying diseases, such as autoimmune disorders, infections, or malignancies. Infection, tumor, neurosarcoidosis, granulomatosis with polyangiitis, Sjogren syndrome, Systemic lupus erythematosus, Giant-cell arteritis, Behcet's syndrome, Relapsing polychondritis and IgG4-related disease are all common causes of secondary cases. The pathophysiology of idiopathic hypertrophic pachymeningitis is

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unclear but based on some hypotheses, it is believed that it is an autoimmune disorder that arises from infiltrative or infectious etiologies [6]. The first report of idiopathic Spinal HPM was published by Charcot and Joffroy in 1869 [7] with involvement of the spinal meninges, and intracranial idiopathic hypertrophic pachymeningitis was reported in 1918 [8]. Compression of anatomic structures by the meninges, thickened by inflammatory infiltration is the most important cause of the clinical features like headache, cranial nerve palsies and ataxia [9]. In hypertrophic pachymeningitis, dura mater becomes abnormally thickened due to accumulation of inflammatory cells, fibrous tissue, and blood vessels. This thickening can exert pressure on underlying brain structures, causing symptoms such as headaches, cranial nerve palsies, visual disturbances, and focal neurological deficits. The clinical presentation of hypertrophic pachymeningitis can vary widely depending on the location and extent of dural involvement. Based on the site affected, hypertrophic pachymeningitis can be divided into two main types: cranial hypertrophic pachymeningitis and spinal hypertrophic pachymeningitis. Cranial hypertrophic pachymeningitis primarily involves thickening of dura mater surrounding the brain, leading to symptoms related to increased intracranial pressure and neurological deficits. On the other hand, spinal hypertrophic pachymeningitis affects the dura mater around the spinal cord, resulting in symptoms such as back pain, weakness, and sensory abnormalities in the limbs. The clinical presentation can vary from mild symptoms to severe neurological deficits depending on the extent of spinal cord compression. Patients with hypertrophic spinal pachymeningitis often present with symptoms such as chronic back pain, radicular pain, sensory disturbances, motor deficits, and bowel or bladder dysfunction.

Diagnosis of hypertrophic pachymeningitis typically involves a combination of clinical evaluation, imaging studies (such as MRI or CT scans), and sometimes dural biopsy for histopathological examination. Treatment strategies aim to reduce inflammation, alleviate symptoms, and prevent disease progression. Corticosteroids, immunosuppressive agents, and sometimes surgery may be considered depending on the underlying cause and individual patient factors. Steroids and other immunosuppressive agents are among the therapeutic strategies proposed, along with radiotherapy and surgical removal of the affected tissue [10]

ETIOLOGY

The etiology of hypertrophic pachymeningitis can be categorized into two main types. Idiopathic hypertrophic pachymeningitis (IHP) occurs without a known cause and is thought to involve an abnormal immune response leading to inflammation and thickening of the dura mater. On the other hand, secondary hypertrophic pachymeningitis (SHP) is linked to specific underlying conditions like autoimmune diseases, infections, or malignancies. These conditions trigger the inflammation and thickening of the dura mater as part of the primary disease process. Understanding the etiology is crucial for determining the appropriate treatment approach based on the underlying cause. Clinical manifestations in the majority of patients are similar. The most common symptoms of cranial IHP are headache and cranial nerve palsy. IHP may also manifest with chronic headache resembling migraine. Headache, attributed to focal dural inflammation is a universal symptom, and at times may be the only symptom for many years. Intraparenchymal involvement in IHP is rare

PATHOPHYSIOLOGY

The pathophysiology of hypertrophic pachymeningitis is a complex process that involves a series of immune responses and

inflammatory mechanisms within the dura mater. This condition is characterized by chronic inflammation leading to thickening and fibrosis of the protective membrane surrounding the brain and spinal cord. In idiopathic hypertrophic pachymeningitis (IHP), an abnormal immune response is thought to be a central factor. This response triggers the activation of immune cells and the release of inflammatory mediators within the dura mater, resulting in tissue damage and scarring. Genetic predispositions and environmental factors may also contribute to the development of IHP. Secondary hypertrophic pachymeningitis (SHP) is associated with underlying conditions such as autoimmune diseases, infections, or malignancies. In SHP, the primary disease process initiates an inflammatory response that affects the dura mater, leading to thickening and fibrosis. The specific mechanisms vary depending on the underlying condition but often involve dysregulation of the immune system and chronic inflammation.The pathophysiology of hypertrophic pachymeningitis involves a cascade of events that ultimately lead to the thickening and fibrosis of the dura mater. Understanding these underlying processes is crucial for developing targeted treatment strategies to effectively manage the condition. Treatment typically involves addressing the underlying cause, reducing inflammation, and managing symptoms to improve quality of life.

DIAGNOSIS

1. **Clinical Evaluation-**

The doctor will start by taking a detailed medical history and performing a physical examination. Symptoms of hypertrophic pachymeningitis can include headaches, visual disturbances, cranial nerve palsies, and signs of increased intracranial pressure.

2. **Imaging Studies-**

Imaging studies such as MRI (Magnetic Resonance Imaging) or CT (Computed Tomography) scans are commonly used to visualize the thickening of the dura mater. These imaging tests can help identify the location and extent of the thickening and any associated complications like compression of adjacent structures.

3. **Biopsy-**

In some cases, a biopsy may be necessary to confirm the diagnosis. A small sample of the affected dura mater may be taken and examined under a microscope to look for characteristic changes consistent with hypertrophic pachymeningitis.

Other diagnosis are ANA, ANCA Profiles etc.

Hypertrophic pachymeningitis can lead to various complications due to the thickening of the dura mater, the outermost layer of the brain's protective covering. Some detailed complications include compression of nearby structures like nerves or blood vessels, resulting in neurological symptoms such as weakness, numbness, or vision changes. Additionally, chronic inflammation can lead to persistent headaches, cognitive issues, and in severe cases, permanent damage to the brain or spinal cord.

TREATMENT

Therapeutic strategies for HP include steroids, azathioprine, methotrexate, rituximab, radiotherapy, surgery and observation. Early treatment is an important point in management of these patients because of its association with improved outcome in terms of neurological recovery [19]. Ventriculoperitoneal shunt and antiepileptic drugs may be needed for symptom therapy. Corticosteroids such as prednisolone could decrease the dural thickening as documented with MRI and resulted in dramatic reduction or even complete remission of symptoms in some patients. Patients may become steroid dependent. Surgical techniques and ventriculoperitoneal shunt have been used with variable success. An

empirical treatment with antituberculous drugs may be warranted in selected patients.

CONCLUSION

Hypertrophic spinal pachymeningitis, a rare condition marked by the thickening of the dura mater enveloping the spinal cord, manifests with a spectrum of symptoms including persistent back pain, nerve-related discomfort, sensory abnormalities, motor impairments, and potential issues with bowel and bladder function. The diagnostic process typically involves a comprehensive clinical assessment, advanced imaging techniques like MRI or CT scans to visualize the thickened dura mater and assess spinal cord compression, and occasionally a biopsy for definitive confirmation and to rule out other conditions. The prognosis of hypertrophic spinal pachymeningitis hinges on various factors such as the underlying cause, extent of spinal cord involvement, and the individual's response to treatment. Early detection and appropriate therapeutic interventions play a pivotal role in shaping outcomes and averting potential complications in affected individuals. Treatment strategies commonly encompass a combination of medications like corticosteroids and immunosuppressants, surgical interventions to alleviate spinal cord compression, and adjunctive measures such as physical therapy to enhance functional recovery. By incorporating details on the etiology, clinical presentation, diagnostic modalities, and prognostic considerations, a comprehensive understanding of hypertrophic spinal pachymeningitis can be attained. The intricate interplay of these facets underscores the importance of a multidisciplinary approach in managing this complex condition and optimizing patient care.

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