# Sensorineural hearing loss due to Idiopathic Hypertrophic Pachymeningitis: a case report.

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## INTRODUCTION

Hypertrophic Pachymeningitis (HP) is a rare progressive fibro-inflammatory disorder resulting in a focal or diffuse thickening of the intracranial and/or spinal dura mater. It can be secondary to infection, autoimmune disease or cancer but mostly is idiopathic (Idiopathic Hypertrophic Pachymeningitis, IHP). Its clinical presentation can be heterogeneous including headache, cranial nerve palsy, ataxia, seizures and myelopathy, occurring either alone or in combination.

## CASE REPORT

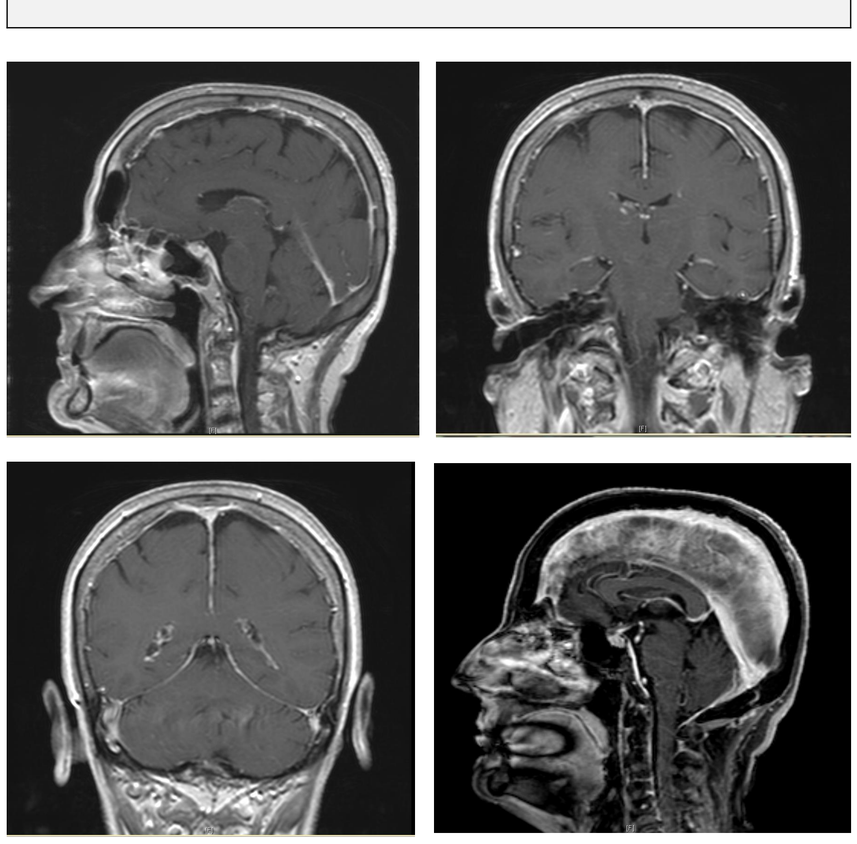
We describe the case of a 74 year-old female patient who was referred to our hospital in January 2016 because of progressive hearing impairment characterized by tinnitus and hearing loss, onset for the first time about ten years before, and detection at brain magnetic resonance imaging (MRI) of diffuse thickening and abnormal enhancement of the dura mater. At admission she also complained onset of headache.

Firstly we performed again brain MRI: it confirmed the presence of diffuse dural thickening in the craniocervical region including both acoustic channels. Neurological examination revealed mild right oral muscular weakness and body imbalance without visual control. Audiologic evaluation confirmed bilateral sensorineural hearing loss, especially in the left ear. Laboratory test showed only elevated erythrocyte sedimentation rate. Evaluation for Lyme disease, syphilis, tuberculosis, cysticercosis and fungi were negative in both serum and cerebrospinal fluid (CSF). Tests for HIV and HTLVI-II resulted negative. No pathological results were found in thyroid function, C-reactive protein, B2 microglobulin, angiotensin converting enzyme, rheumatoid factor, autoimmune antibodies and plasma IgG4 rate. Cancer screening, including tumor markers and thoracic and abdominal computed tomography (CT) scans, excluded presence of tumor. CSF analysis was normal; cultures and cytology for carcinomatous cells were negative. A meningeal biopsy showed only mild fibrosis and low inflammatory infiltrates.

Based on these results, a diagnosis of IHP was done and steroid therapy was started. After three months, during follow-up visit, we detected progressive improving of hearing impairment and headache.

**ETIOLOGY OF THICKENED ABNORMALLY ENHANCING DURA MATER ON GADOLINIUM MRI [1]**

- **Idiopathic cranial or spinal pachymeningitis**
- **Intracranial hypotension**
  - Spontaneous
  - Post spinal fluid drainage
- **Infections**
  - Lyme disease
  - Syphilis
  - Mycobacterium tuberculosis
  - Fungal infections
  - Cysticercosis
  - Human T-cell lymphotrophic virus I.
  - Malignant external necrotizing otitis due to Pseudomonas
- **Systemic autoimmune/vasculitic disorders**
  - Wegener granulomatosis
  - Rheumatoid arthritis
  - Sarcoidosis
  - Behcet disease
  - Sjogren syndrome
  - Temporal arteritis
- **Malignancy**
  - Dural carcinomatosis
  - Metastatic disease in adjacent skull bone
  - Lymphoma
  - Meningioma
- **Trauma**



**Brain MRI T1-weighted images following gadolinium infusion**

## CONCLUSIONS

HP is a rare condition and brain MRI has a key role in the diagnostic work-up. When MRI is suggestive for the disease all specific etiologies have to be evaluated; if all of these are excluded the diagnosis of HIP can be performed. This case highlights the importance of considering HP also in patients with progressive sensorineural hearing loss or hearing impairment especially when it is accompanied by other cranial neuropathy or headache. Corticosteroid therapy is usually effective so HP can be a cause of a potentially treatable sensorineural hearing loss.

**References:**

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2 - Lim E-J, Kim S-H, Lee S-H, et al. Reversible Sensorineural Hearing Loss due to Pachymeningitis Associated with Elevated Serum MPO-ANCA. Clinical and Experimental Otorhinolaryngology. 2011;4(3):155-158.

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