Hypertrophic pachymeningitis

Hypertrophic pachymeningitis is a chronic Inflammation disease, manifesting as a fibrous thickening of the dura mater.

Hypertrophic pachymeningitis is a rare disorder of diverse etiology. It was first described by Charcot and later by Naffziger and Stern [1)]

It can involve the cranial or the spinal dura or both.

Classification

The condition can be broadly divided into two forms:

Primary or Idiopathic hypertrophic pachymeningitis where no identifiable cause is found

Secondary where identifiable causes co-exist, although their definite relationship to the development of this condition may be debatable.

IgG4-related hypertrophic pachymeningitis

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see Idiopathic hypertrophic cranial pachymeningitis.

Etiology

see IgG4-related hypertrophic pachymeningitis

Early reports were in relationship to tuberculosis or syphilis. Exact etiopathogenesis of this entity is still unknown, but it is speculated to be an autoimmune phenomenon or occur as a direct result of infectious or infiltrative pathology [2)].

Diagnosis

HP can appear as a vanishing tumor, and pathological evaluation is essential for a precise diagnosis. If spontaneous disappearance of tumefactive intracranial lesions is encountered, the possibility of HP should be considered [3)].

Treatment

A 68-year-old man presented at a nearby hospital with a headache and a low-grade fever. A blood test revealed inflammation, as well as elevation of IgG4 level. Magnetic resonance imaging(MRI)revealed diffuse thickening of the dura mater, dominantly in the posterior fossa and cerebellar tentorium. The lesion was enhanced significantly with gadolinium(Gd). An open biopsy was performed to determine pathological diagnosis. Hematoxylin and eosin staining showed infiltration of inflammatory cells, including plasma cells. The infiltrating cells were positive for the IgG4. Post-operatively, the patient was treated with glucocorticoid, and both the inflammation and patient symptoms were improved. In conclusion, IgG4 is related to the etiology of hypertrophic pachymeningitis and glucocorticoid therapy is effective for this disease [4].

Case reports

A 59-year-old man with hypertrophic pachymeningitis (HP), initially presenting as a tumefactive lesion that disappeared spontaneously. He developed headache and left abducens nerve palsy 2 years before admission, and brain magnetic resonance imaging (MRI) revealed a round mass lesion. Meningioma was initially considered, but the lesion disappeared spontaneously along with the symptoms. However, 6 months before admission, left abducens nerve palsy reappeared. Repeated MRI revealed multiple intracranial tumefactive lesions. HP was diagnosed based on the pathological analysis of the biopsied specimen. HP can appear as a vanishing tumor, and pathological evaluation is essential for a precise diagnosis. If spontaneous disappearance of tumefactive intracranial lesions is encountered, the possibility of HP should be considered [5].

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