Pathologies related to lymphocyte infiltration and inflammatory cells are classified, according to their topography, into idiopathic orbital pathology, idiopathic hypertrophic pachymeningitis, Tolosa-Hunt syndrome and lymphocytic hypophysitis.

Hypertrophic craniocervical pachymeningitis (HCP) is a rare disease causing chronic inflammatory hypertrophy of the cranial and spinal dura mater.

Idiopathic hypertrophic pachymeningitis (IHP) is a rare disease, and it is characterized by chronic progressive inflammatory fibrosis and thickening of the dura mater with resultant compression of the spinal cord or neural structure without any identifiable cause. It can occur in the intracranial or spinal dura mater alone or as a craniospinal form. The spinal form is rarer than the cranial form and the craniospinal form is extremely rare.

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Bang et al., report a rare case of IHP in the craniocervical junction involving both the cranial and spinal dura mater and discuss the diagnosis and management of the disease [1].

A 78-year-old man presenting with a rare case of HCP and summarized the clinical features, laboratory evaluations and treatment of the case.

In this case, the HCP involved the intracranial dura and high cervical regions, manifesting as lower cranial nerve palsies, headache, and neck pain, developing over 7 months. Magnetic resonance imaging revealed thickening of the dura in the craniocervical region with peripheral enhancement. Steroid therapy was commenced and the symptoms improved rapidly.

HCP can be diagnosed by MRI and laboratory investigations. In this case corticosteroid treatment was effective, although care must be taken when slowly reducing the dose. This case highlights HCP as a cause of progressive cerebellomedullar and cervical spinal cord symptoms [2].

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see Idiopathic hypertrophic cranial pachymeningitis.

1)
Bang JH, Cho KT, Kim EJ. Idiopathic Hypertrophic Pachymeningitis in the Craniocervical Junction. Korean J Spine. 2015 Sep;12(3):169-72. doi: 10.14245/kjs.2015.12.3.169. PubMed PMID: 26512276; PubMed Central PMCID: PMC4623176.
2)
Zhu R, He Z, Ren Y. Idiopathic hypertrophic craniocervical pachymeningitis. Eur Spine J. 2015 May;24 Suppl 4:S633-5. doi: 10.1007/s00586-015-3956-4. PubMed PMID: 25893340.