Hypertrophic pachymeningitis mimicking hemicrania continua

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Introduction

Hemicrania continua (HC) is a primary headache characterized by a unilateral, moderate, fluctuating but enduring pain, with exacerbations marked by migrainous and cranial autonomic symptoms.

Although HC diagnosis is based on clinical history, neurological examination and absolute response to indomethacin, HC misdiagnosis is frequent and likely due to its relative rarity, autonomic signs paucity and distinct similarities to migraine attacks during exacerbations. In this regard, it is crucial to consider that a clinical phenotype similar to primary HC may be subtended by other neurological, neoplastic or systemic disorders.

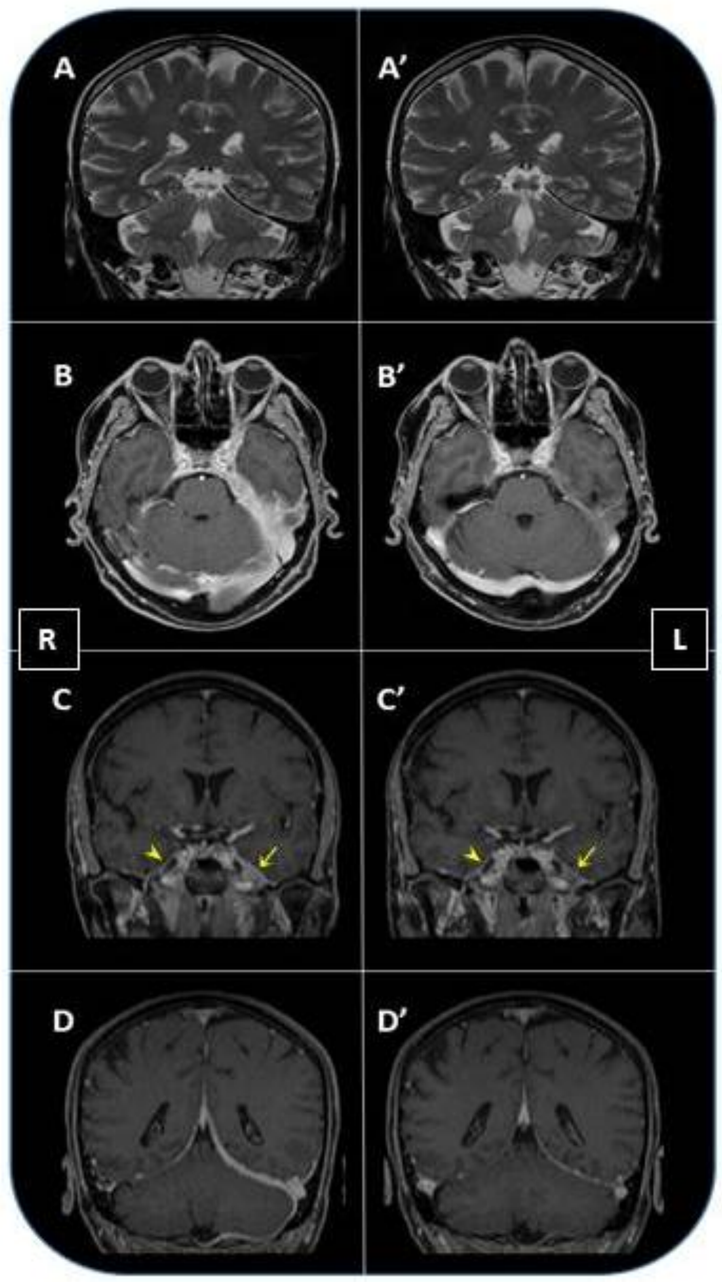
Case report

We report a case of a 62-years-old man experiencing a moderate, persistent headache, with throbbing exacerbations of about 2-3 hours, strictly localized in the left orbital and temporal regions associated with trigeminal-autonomic symptoms ipsilaterally to pain. This clinical picture and the absolute responsiveness to indomethacin administration conducted to the diagnosis of HC. Later, the patient developed diplopia caused by sixth cranial nerve palsy ipsilateral to headache. Clinical, laboratory and neuroimaging findings supported the diagnosis of idiopathic hypertrophic pachymeningitis (IHP).

Conclusion

IHP is a rare fibrosing inflammatory disorder leading to a localized or diffuse dura mater thickening. IHP clinical manifestations are a progressively worsening headache, signs related to possible cranial nerves involvement or venous sinus thrombosis.

Here we report, for the first time, a HC phenotype subtended by IHP in which left pain and autonomic signs were respectively due to ipsilateral pachymeningeal inflammation and fibrous encasement of trigeminal nerves (see figure).



MRI sequences pre (A-D) and post-therapy (A'-D') and both before (A-A') and after (B-D and B'-D') gadolinium administration.

A and A' images revealed marked dural thickening, diffusely involving left tentorium, with a characteristic low T2 signal intensity. In this context, both the physiologic flow void as well as the slow flow phenomena are absent in the ipsilateral sigmoid sinus. After the gadolinium contrast examination (B-D) intense and uniform enhancement of the mastoid-petroclival and tentorial dural thickening is observed. This MRI finding is present also in the left middle cranial fossa, along the temporal lobe, spreading to the left cavernous sinus and involving the ipsilateral dura of the trigeminal mandibular branch (V3), markedly enlarged (arrow in C and C') compared to right (arrowhead on C and C'). Post-therapy MR images (A'-D') revealed decrease of dural thickening without changes of the slow flow phenomena at level of sigmoid sinus

Reference

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