대한임상신경생리학회지 18(1):11-13, 2016
http://dx.doi.org/10.14253/kjcn.2016.18.1.11
pISSN 1229-6414 eISSN 2288-1026

Case Report

토로사-헌트 증후군으로 오인된 특발성 비후성 두개 경막염

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Idiopathic Hypertrophic Cranial Pachymeningitis Masquerading as Tolosa-Hunt Syndrome

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Idiopathic hypertrophic cranial pachymeningitis (ICHP) is diffuse inflammatory process of the dura mater. ICHP can produce similar presentation with Tolosa-Hunt syndrome (THS) if it involves cavernous sinus. A-29-year old male with persistent headache and no definite neurologic dysfunction was noted. Two weeks later, he complained of ophthalmoplegia, and his symptoms were thought to be manifestations of THS. Brain magnetic resonance images revealed diffuse thickened, enhanced pachymeninges in left tentorium. The patient was diagnosed with IHCP. We report a IHCP patient who showed very similar presentation as THS. (Korean J Clin Neurophysiol 2016;18:11-13)

Key Words: Pachymeningitis, Tolosa-Hunt syndrome, Ophthalmoplegia

Received 11 December 2015; received in revised form 9 May 2016; accepted 17 May 2016.

Idiopathic hypertrophic cranial pachymeningitis (IHCP) is a rare disease, characterized by a fibrosing inflammatory process involving the dura mater.[1] It is mainly diagnosed by exclusion as numerous pathological processes lead to a thickening of the pachymeninges.[2] Brain magnetic resonance imaging (MRI) is aid in diagnosis, although biopsy is needed for definite diagnosis.

IHCP commonly presents with headache and cranial nerves (CN) palsy.[1,3,4] However, the Tolosa-Hunt syndrome (THS) could also be a focal manifestation of IHCP. We report the IHCP case similar to THS.

Case Report

A 29-year-old male presented with headache in the left parietal area, which started 2 weeks earlier. Its nature was compressive and accompanied with left eyeball pain, but without visual symptoms and other neurological abnormality. There was no history of diabetes, hypertension, or other specific medication. Routine evaluation for systemic illness was carried

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* This study was supported by the Wonkwang Grant in 2015.
* The authors have no financial conflicts of interest.

out, as well as brain MRI and cerebrospinal fluid (CSF) examination. All results were normal. His headache did not show meaningful improvement in severity and frequency after treatment with analgesic drugs. Two weeks later, he complained of horizontal and vertical diplopia with periorbital pain, and sudden ptosis with limitation of adduction and infraduction in the left eye were noted. He tended to tilt his head to right side. In the red glass test, his vertical diplopia improved at gaze to upward and left side, suggesting CN IV palsy combined with CN III palsy. Pupils were normal and reacting to light, and fundus was normal. There was no meningeal irritation sign. Deep tendon reflexes were preserved and other neurologic examinations were nonspecific. The patient was diagnosed with THS, and hospitalized for further laboratory, brain MRI, and CSF tests. Laboratory analyses included complete blood count, blood sugar, renal and liver function tests, human immunodeficiency virus, venereal disease research laboratory and tuberculin sensitivity tests, autoimmune markers, thyroid function test and tumor markers tumor markers were within normal limits. CSF study showed a mild increase of white blood cell count (30/mm^3, lymphocyte dominant) and nonspecific findings otherwise, suggesting a mild inflammatory condition. T1- and T2-weighted images revealed mild bulging soft tissues in the left cavernous sinus (Fig. 1A). Enhanced T1 weighted image revealed a thickening and enhancement of the left cavernous sinus and superior orbital fissure (Fig. 1B), and of the left tentorium (Fig. 1C).

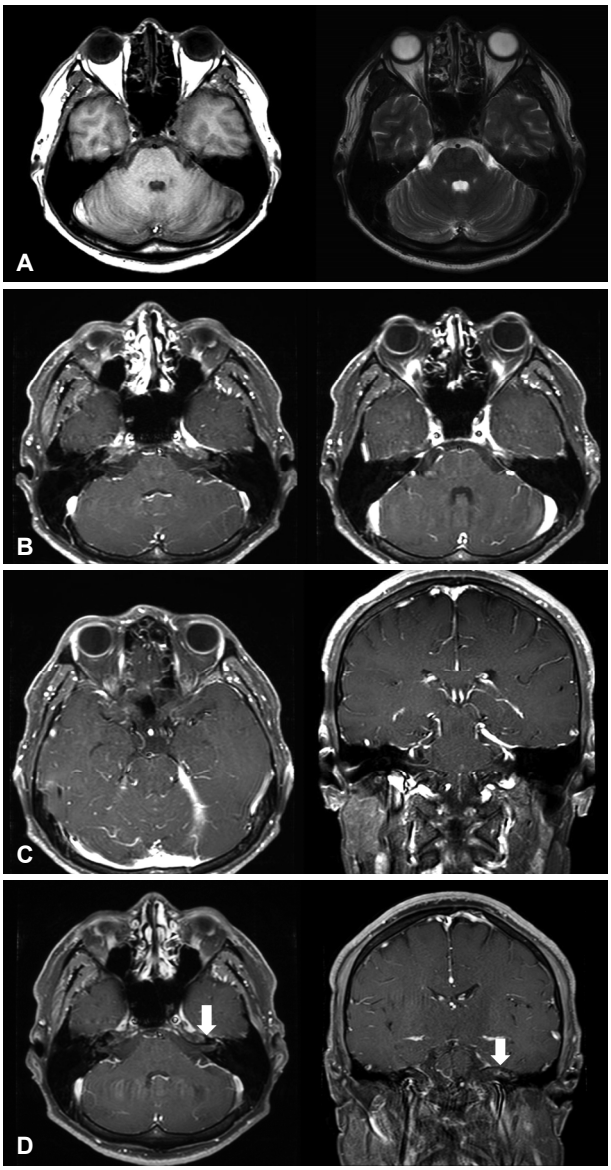

Figure 1. Brain MRI of patient. T1-and T2-weighted images reveled mild bulging soft tissues in the left cavernous sinus left cavernous sinus (A). Enhanced T1 weighted image show a thickening and enhancement of the left cavernous sinus and superior orbital fissure (B), and of the left tentorium (C). Three weeks later, abnormal enhancement is visible in the facial nerve (arrow), suggesting inflammatory facial nerve disease in T1-weighted images (D).

Abnormal convexity of the cavernous wall and stenosis of the ipsilateral intracavernous internal carotid artery were not noticed. The patient was treated with steroid for confirmative diagnosis of IHCP. Three weeks after initiation of steroid therapy, ptosis and extra-ocular movement of left eyeball were improved. However, he complained of acute peripheral facial palsy and intermittent headache. With a follow up MRI, abnormal enhancement was detected in the left facial nerve, suggesting an inflammatory facial nerve disease (Fig. 1D). We continued the steroid therapy and the symptoms were improved.

Discussion

IHCP is a diffuse inflammatory disease causing thickening of the dura mater, which presents with headache and multiple CN palsies. This is caused by compression of the brainstem due to the thickened dura.[1,5] At the onset of the disease, the patient may only complain of common nonspecific symptoms such as headache, nausea and vomiting.[3] CN VIII is the most frequently involved CN, followed by V, VII, X, and XII, that are involved at equal frequency.[3] THS, which is a 'painful ophthalmoplegia' caused by a nonspecific granulomatous in-

flammation of the cavernous sinus, may also, though rarely, be a focal manifestation of hypertrophic pachymeningitis. But the headache in THS should precede paresis of CN III, IV and/or VI by 2 weeks, or develop with it. And the headache is usually localized around the ipsilateral brow and eye. In our case, headache and eyeball pain without ophthalmoplegia had preceded paresis of CN III and IV by 4 weeks. MRI findings were different from the typical characteristics of THS. Therefore, a diagnosis of IHCP is more adequate rather than THS in our patient.

Biopsy is needed for confirmatory IHCP diagnosis, but brain imaging such as computed tomography and MRI can be helpful for detection. On MRI, the thickened dura mater appears isointense or hypointense on T1-weighted images, and hypointense - and often with an hyperintense edge - on T2-weighted images. Contrast administration reveals uniform enhancement of the thickened meninges.[6] However, brain imaging can reveal nonspecific findings, even up to 2 years before there are any significant findings.[3] Therefore, close observation for symptoms and follow up MRI should be performed if needed for patients with normal brain images and suspected IHCP.

Our patient complained only headache and left eyeball pain, without any specific neurological dysfunction. Results from initial brain MRI and CSF study was normal, and all the other tests revealed nonspecific findings. However, after treatment, THS-like ophthalmoplegia was noted. Follow up MRI revealed diffuse left tentorial thickening and enhancement, suggesting IHCP. Therefore, at an early stage of IHCP, brain imaging can reveal nonspecific findings, and nonspecific symptoms such as headache only can be presented. Persistent headache should be carefully observed and considered during follow-up studies.[7] Even without visible ophthalmoplegia, persistent eyeball pain can be considered an early sign of ophthalmoplegia such as IHCP, THS or other cranial nerve dysfunction.

The recurrence of IHCP is well-known.[2,3] In our patients, despite steroid therapy, additional CN VII palsy was also presented. It means that long-term follow-up is needed for IHCP patients, and that other treatment such as immunosuppressant or surgery should be considered for steroid-unresponsive patients.[6]

We suggest that in patients with clinical diagnosis of THS or chronic headache with cranial nerve palsies, neuroimaging is absolutely needed for IHCP detection. In addition, the relationship between THS and IHCP should be discussed and etiologically studied.

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