Idiopathic hypertrophic pachymeningitis (IHP) is a rare inflammatory disease, usually involving the dura mater of skull base, tentorium, and falx. Resultant nerve encasement and ischemia produce multiple cranial nerve palsy and headache. As numerous pathological entities cause thickening of the pachymeninges, when the evaluation fails to reveal a cause, IHP is considered. With the recent development in MRI technique, variable causes of pachymeningeal lesions have been reported. However, the pathogenesis and clinical significance of pachymeningeal enhancement on MRI is still poorly understood. To our knowledge, there is no case with IHP presenting as an isolated abducens nerve palsy in Korea. We describe the clinical and MRI findings in a patient whose abducens nerve palsy was probably caused by IHP.

**CASE REPORT**

A previously healthy 55-year-old woman presented with diplopia for 1 month. The neurological examination did not reveal significant alteration, except the limitation in lateral gaze of the right eye. MRI showed symmetric dural thickening with well enhancement in both inferior frontal area and asymmetric focal dural thickening in the right skull base adjacent to the cisternal segment of the abducens nerve (Fig. 1-A, B). A provisional diagnosis of hypertrophic pachymeningitis was entertained. She underwent general hematology testing including ESR, CRP, blood counts, glucose, fibrinogen, angiotensin converting enzyme, liver, kidney and thyroid function; immunological markers (ANA, p–ANCA and c–ANCA, cryoglobulins, anti–phospholipid antibody, anti–cardiolipin antibody, rheumatoid factor); tumor markers (AFP, CEA, CA-125). Investigation for hepatitis B– and C–virus, cryptococcus, mycobacterium tuberculosis, HIV, parasite antibody and herpes virus were performed on serum. There was no evidence of any systemic inflammatory disease, although ESR was increased to 46 mm/hr. No evidence of conditions known to cause pachymeningeal thickening was found. Cerebrospinal fluid examinations were normal. Also, chest, abdomen and pelvic CT revealed no abnormal findings. However, meningeal biopsy was...
not performed. She had improvement in diplopia after initiating high dose intravenous corticosteroid treatment. She was switched to oral prednisolone with gradual reduction to 10 mg daily. Follow-up MRI, performed at 8 weeks after the initial study, showed complete resolution of the previously enhancing lesions (Fig. 1–C, D). She is currently symptom-free with only low doses of steroid.

DISCUSSION

The diagnosis of IHP can be made only after the exclusion of several other possible causes of dural thickening, including so-called spontaneous intracranial hypotension, infectious disease including tuberculosis, syphilis, HTLV–I, and fungal infections, inflammatory disease including sarcoidosis, Tolosa–Hunt syndrome, rheumatoid arthritis, Wegener’s granulomatosis, and neoplastic disease, and systemic fibrosing disorders. Therefore, systemic evaluation and extensive diagnostic procedures including dural biopsy are essential in approaching pachymeningeal enhancement to search for treatable causes. But it was impossible for us to perform the biopsy in this patient as she did not agree with the aggressive diagnostic procedure. Biopsy of the dural lesion should be considered and is recommended if the patient clinically deteriorates or the neuroimaging worsens despite treatment. Although we did not have a biopsy in this case, she has been followed for 1 year and did not develop any signs of infection or other causes for the pachymeningitis.

MRI is the method of choice for diagnosis of dura mater disease, as well as for monitoring disease activity and its responsiveness to therapy. Most common site of IHP are the tentorium, falx, clivus, parasellar, and cavernous regions. MRI features are considered characteristic, but not pathognomonic of IHP. In IHP the gadolinium-enhanced MRI used to evaluate the localization, extent, and pattern of abnormal dura and to monitor disease progression as the abnormal area appear to correlate with the pathologic inflammatory process. In this case, MRI findings with asymmetric focal dural thickening in the right skull base adjacent to the cisternal segment of the abducens nerve are well correlated with clinical features. Also, follow-up MRI was improved with clinical status recovery after steroid therapy.

Clinical presentation is inconstant and nonspecific. The disease commonly presents with longstanding headache. When the dura mater at the skull base in the posterior fossa was affected, the lower cranial nerve palsy occurred. The clinical features appeared to be associated with the location of the abnormal dura observed on gadolinium-enhanced MRI. Although studies on IHP have described the seventh nerve followed by the lower cranial nerves as the most frequently affected, others have described preferential dysfunction in the
cranial nerves that serve ocular motor function particularly with parasellar involvement. IHP frequently progresses if untreated and although steroid treatment improves the patients, the benefit may be temporary and partial. She was a good responder to steroid therapy, in agreement with the majority of the previous reports. In conclusion, although the gold standard for the diagnosis of IHP is biopsy of the thickened dura mater, the case was labeled as idiopathic since no identifiable cause was found. We describe a patient with IHP who presented with isolated abducens nerve palsy and had characteristic MRI features.

REFERENCES