CASE OF IDIOPATHIC HYPERTROPHIC CRANIAL PACHYMENINGITIS ASSOCIATED WITH OPTIC NEUROPATHY AND FACIAL PALSY

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Background: Idiopathic hypertrophic cranial pachymeningitis (IHCP) is a rare disease, which causes chronic progressive inflammationand thickening of the basal dura mater and produces multiple cranial neuropathies, headache, ataxia and seizure.
Case and Result: 69 year-old women presented with progressive bifrontal headache, lt visual loss with periorbital pain and lt facial palsy. Brain MRI revealed thickened, highly enhanced pachymeninges in both frontal supratentorial areas. A subsequent meningeal biopsy showed non-specific chronic inflammations of the pachymeninges. The patient dramatically responded to high dose steroid therapy.She remained free of neurological symptoms after prednisolone was tapered 10mg/day.However, after 2 years headache and visual loss recurred and corticosteroid therapy improved the headache and visual loss.
Conclusion: We report a case of idiopathic hypertrophic cranial pachymeningitis with optic neuropathy and facial palsy.