Hypertrophic Tumefactive Pachymeningitis Presenting as Left Visual Loss

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Case History

43 year old lady presented with acute onset of visual loss involving the left eye evolving over a period of 24 hours. It was not associated with drooping of the left eye, double vision, pain, facial lag or swallowing difficulty. She had no complaints of weakness, sensory or bladder disturbance. It was not preceded by fever, joint pain or similar complaints before. On examination her visual acuity in the left side was present only for lights, absent color vision and fundus was normal. Other cranial nerves and rest of the neurological examination was unremarkable. A provisional diagnosis of left optic neuropathy of retrobulbar nature was made and she was investigated. Her complete blood count, Renal profile, Liver function tests, Lipid profile, Chest X-ray PA, Ultrasound abdomen was normal. ANA, DSDNA, Serum calcium, ACE, CANCA, PANCA was normal. VEP of the left eye was absent and right eye was normal. Her CSF analysis showed elevated protein. C.T Brain revealed left temporal lobe hypo density which did not enhance with contrast. MRI Brain with contrast was done which revealed isointense lesion involving the dura in T1, hypointense in T2, Flair showing evidence of temporal lobe edema, and contrast enhanced MRI showing diffuse multicentric enhancement of dura, encroaching the optic chiasma and encasing the left optic nerve (Figures 1, 2, 3, 4). A diagnosis of hypertrophic pachymeningitis was made. The enhancement of the dura was mimicking multifocal meningioma. Patient was started on Methyl prednisone 1gm IV OD for 3 days followed by oral

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tapering for 30 days. She improved dramatically. She completely regained her vision on day 5. Follow up MRI was done 2 months later which showed regression of lesions involving the frontal and parietal dura (Figures 5, 6). Similarly the lesions involving the optic nerve had regressed but the dural enhancement involving the sphenoid wing adjacent to temporal lobe remained the same (Figures 7, 8). Absence of bony involvement, vascularity, response to steroids and regression of parietal and optic nerve lesions made multifocal meningioma unlikely. She was advised biopsy but she refused. She is on regular follow up.

**Discussion**

Hypertrophic pachymeningitis is an extremely rare fibrosing inflammatory process involving the dura mater. Pachymeningitis could be primary or secondary. Secondary could be due to granulomatous, paraneoplastic and autoimmune etiologies. If no etiology is identified it becomes idiopathic pachymeningitis, a diagnosis of exclusion. The disease could be focal and multicentric. A multicentric presentation as seen in our case can cause tumefactive enhancement which is a rare radiological appearance. In contrast enhanced MRI the enhancement is usually diffuse, nodular and tumefactive. Hypertrophic pachymeningitis presenting as visual loss is rare and cranial neuropathies are due to involvement of cranial nerves at skull base by the enlarged dura.

Other documented neurologic complications of IHP include venous sinus thrombosis, obstructive hydrocephalus and cerebral edema. Dural biopsy will be the investigation of choice to differentiate primary from secondary forms. This case is being presented to highlight the rare case of Idiopathic tumefactive pachymeningitis presenting as left optic neuropathy.

**References**