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LETTER TO THE EDITOR

"Encased brain" in hypertrophic pachymeningitis

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Dear Editor:

A 4-year-old girl presented with a history of acute-onset drooping of the left eye lid for 10 days. There was no associated fever, cough, reduced appetite, recent-onset motor weakness, seizure, or alteration in consciousness. She had been diagnosed with tubercular meningitis with vasculitis, left hemiparesis, and communicating hydrocephalous at the age of 1.5 years (Fig. 1a–d) and took a prolonged course of anti-tubercular therapy for 2 years. At 3 years of age, she underwent right ventriculoperitoneal shunt placement for symptomatic hydrocephalous. Subsequently, she had remained well with residual hemiparesis.

On examination, she had mild pallor, left-sided ptosis and incomplete ophthalmoparesis (upward and inward movement restriction), poor gag reflex (bulbar weakness), left-sided facial paresis (upper motor neuron), and residual left hemiparesis. The rest of the systemic examination was unremarkable. A clinical diagnosis of either a relapse or a new complication of the primary central nervous system tuberculosis was considered. Cerebrospinal fluid analysis showed absence of cells, elevated proteins (190 mg/dL), reduced glucose (39 mg/dL), and absence of organisms on gram stain, culture, and PCR-based testing. Magnetic resonance imaging (MRI) of the brain showed fresh mid-brain lesions, left-sided subdural collection, diffuse hypertrophic pachymeningitis, and encephalomalacic changes in the right basal ganglia (Fig. 2a–g). She was re-initiated on anti-tubercular therapy (isoniazid, rifampicin, ethambutol, and pyrazinamide) due to hypoglycorrhachia and new-onset lesions. Additionally, intravenous pulse corticosteroids were administered to reduce the associated pachymeningitis causing entrapment neuropathy.

Hypertrophic pachymeningitis is a rare condition characterized by localized or diffuse thickening of the dura mater secondary to inflammatory or infectious conditions including central nervous system tuberculosis, syphilis or fungal infections, shunt, trauma, and tumors, or the cause may remain unknown [1, 2]. The underlying pathogenesis is probably immune-mediated or infiltrative [3]. Chronic inflammation of usually inert dura mater by lymphocytes and plasma cells is the main pathological finding [2]. Sometimes, chronic intracranial hypotension may cause pachymeningeal enhancement secondary to compensatory dilatation of dural veins [4]. Brain encasement may result in compression of intracranial structures and presents as headache, cerebellar dysfunction, or cranial neuropathy. MRI is the investigation of choice and delineates the characteristic dural enhancement and/or thickening. Nerve entrapment may occur anteriorly at the level of cavernous sinus causing superior orbital fissure syndrome and paresis of II–VI cranial nerves, or posteriorly at the falcotentorial junction entrapping V to XII cranial nerves, with involvement of VIII nerve being the commonest. In the index case, ocular and bulbar symptoms could be attributed to the combined effect of midbrain tuberculomas as well as encasement of oculomotor, glossopharyngeal, and vagus nerves. Steroid therapy is the mainstay of treatment and

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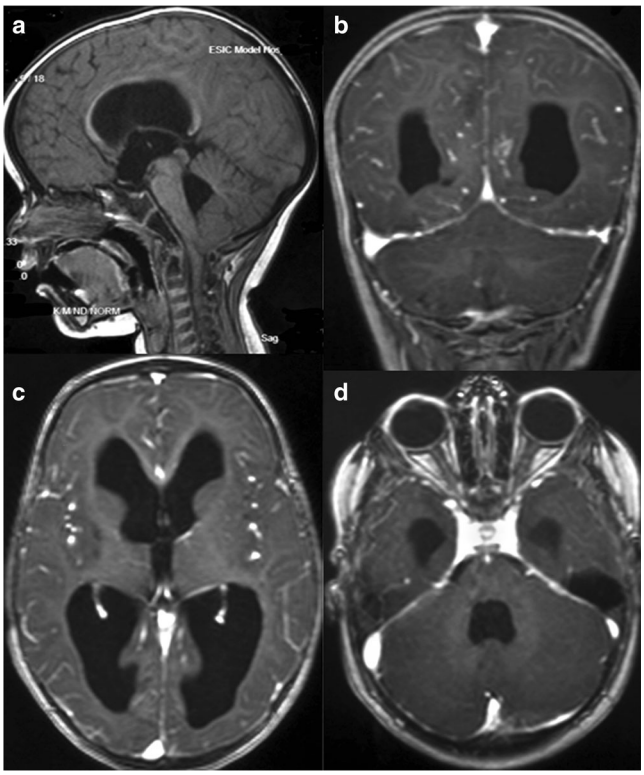

Fig. 1 Contrast-enhanced T1-weighted magnetic resonance images of the brain: **a** sagittal, **b** coronal, and **c**, **d** axial sections showing diffuse leptomeningeal enhancement with moderate communicating hydrocephalous at 1.5 years of age

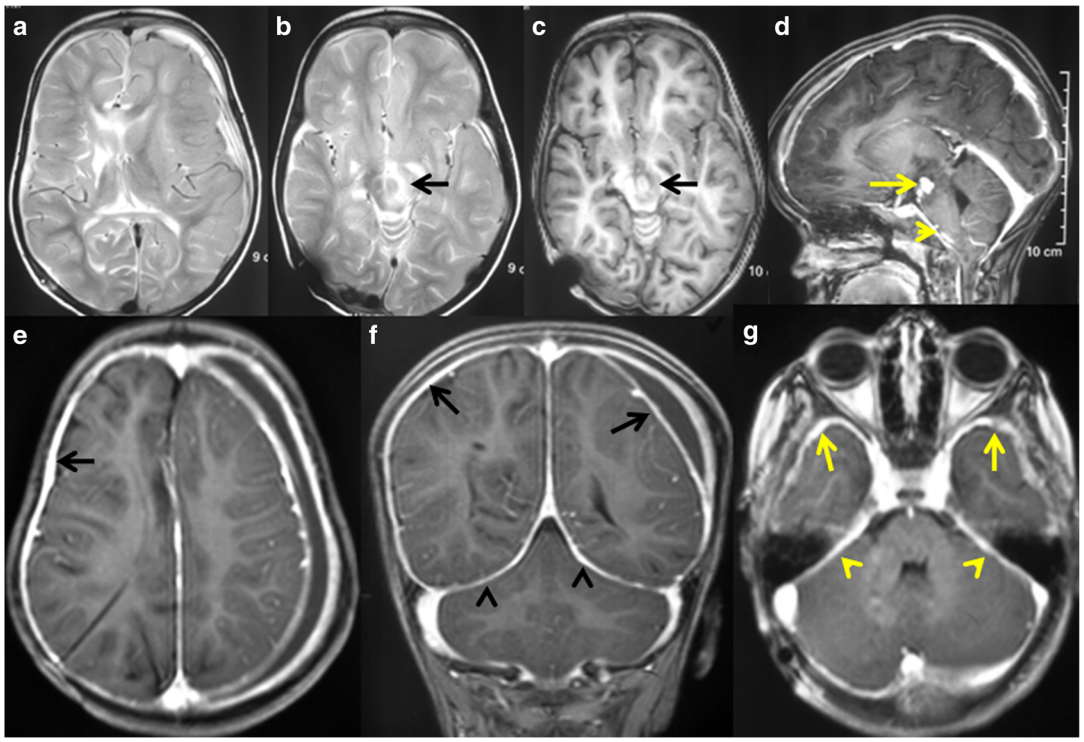

Fig. 2 MRI of the brain at 4 years of age. T2-weighted axial sections (**a**, **b**) showed encephalomalacic changes in the right putamen and globus pallidus. The midbrain showed hyperintense signal changes (arrow), which were hypointense on T1-weighted sequence (**c**) (arrow) and irregular contrast enhancement (**d**) (arrow). Contrast-enhanced T1-weighted axial image (**e**) showing diffuse dural enhancement (arrow) including the falx cerebri and mild left-sided subdural collection. Coronal section (**f**) showing encasement of both cerebral (arrows) and cerebellar (arrow heads) hemispheres by thickened dura mater. Axial section (**g**) at the level of cavernous sinus showing nodular enhancement of anterior and lateral portion of the middle cranial fossa (arrow) and thickened tentorium (arrow head)

other immunomodulating agents may be considered in refractory cases [3]. Delay in treatment of cases with hypertrophic pachymeningitis may result in permanent ischemic damage and residual disability.

Compliance with ethical standards

Conflict of interest Nothing to declare.

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