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Extensive supratentorial and leptomeningeal dissemination in a child with large cell/anaplastic medulloblastoma

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ABSTRACT

Of the posterior fossa tumors in children, 30–40% are due to medulloblastomas. We report a 4-year-old boy, who presented with vomiting and headache, and was subsequently diagnosed with anaplastic medulloblastoma. Neuroimaging revealed a posterior fossa mass, with unusual extensive supratentorial and leptomeningeal dissemination, illustrating the aggressive nature of this tumor.

1. Introduction

Medulloblastomas often occurs between the ages of 4 and 10 years. It presents as a relatively discrete mass usually found in the cerebellar midline or hemisphere, generally hyperdense on CT, with low signals on T1 weighted MR images, with almost always an enhancement pattern [1]. Tumor seeding of the subarachnoid space occurs commonly in children and infants [2].

Large cell/anaplastic medulloblastoma, which represents about 4% of medulloblastomas, is a distinct histological variant that is characterized by highly aggressive behavior, with early cerebrospinal fluid dissemination and extra-CNS metastases. It is associated with a poorer prognosis, with a 4-year event-free (EFS) and overall survival (OS) being reported as 58% and 67%, respectively, with metastases being associated with a poorer outcome [3]. We present a case of a child with anaplastic medulloblastoma, with unusual extensive supratentorial and leptomeningeal dissemination.

2. Case report

A 4-year-boy was transferred to our hospital from India with an initial presentation of vomiting and intermittent headache for 6 months. A CT performed in India had shown a posterior fossa tumor with significant hydrocephalus. The child presented to us with a VP shunt in situ. Physical examination revealed ataxia and bilateral lower limb weakness, but was otherwise unremarkable.

An MRI was performed in our hospital and multiplanar pre- and post-contrast images of the brain and spine were obtained. The child was not sedated and coped well with the procedure. The study was performed on a 1.5 T Siemens Magnetom Symphony MRI scanner and sequences done included axial T2-weighted, gradient-recalled echo (GRE), diffusion-weighted, coronal FLAIR, and axial, coronal and sagittal T1-weighted post-contrast images after administration of intravenous Magnevist (gadopentetate dimeglumine).

A large solid mass was seen occupying almost the whole of the fourth ventricle, causing expansion and displacement of the adjacent cerebellar hemispheres. It measured about 5.0 cm (SI) × 2.2 cm (LR) × 2.9 cm (AP) in size, and appeared heterogeneously low in signal intensity on T1 weighted FLAIR images (Fig. 1a) and was of intermediate to high signal intensity on T2 weighted images (Fig. 1b). There was no evidence of hemorrhage within or around the mass (Fig. 1c), no infarcts and no MRI evidence of diffusion restriction. The primary mass showed significant post-contrast enhancement. There was an expansion of the fourth ventricle, with an unusual supratentorial spread involving the leptomeninges and parenchyma. Focal masses were seen involving the right occipital (Fig. 1d), bilateral temporal (Fig. 2a) and bilateral frontal lobes (Fig. 2b and c), with areas of subcortical white matter edema. Extensive CSF spread was demonstrated with nodular leptomeningeal enhancement (Figs. 2d, and 3a and b), as well as small masses seen particularly in the dependent CSF spaces. MRI spine revealed extensive continuous drop metastases involving nearly the entire neuraxis (Fig. 3c and d).

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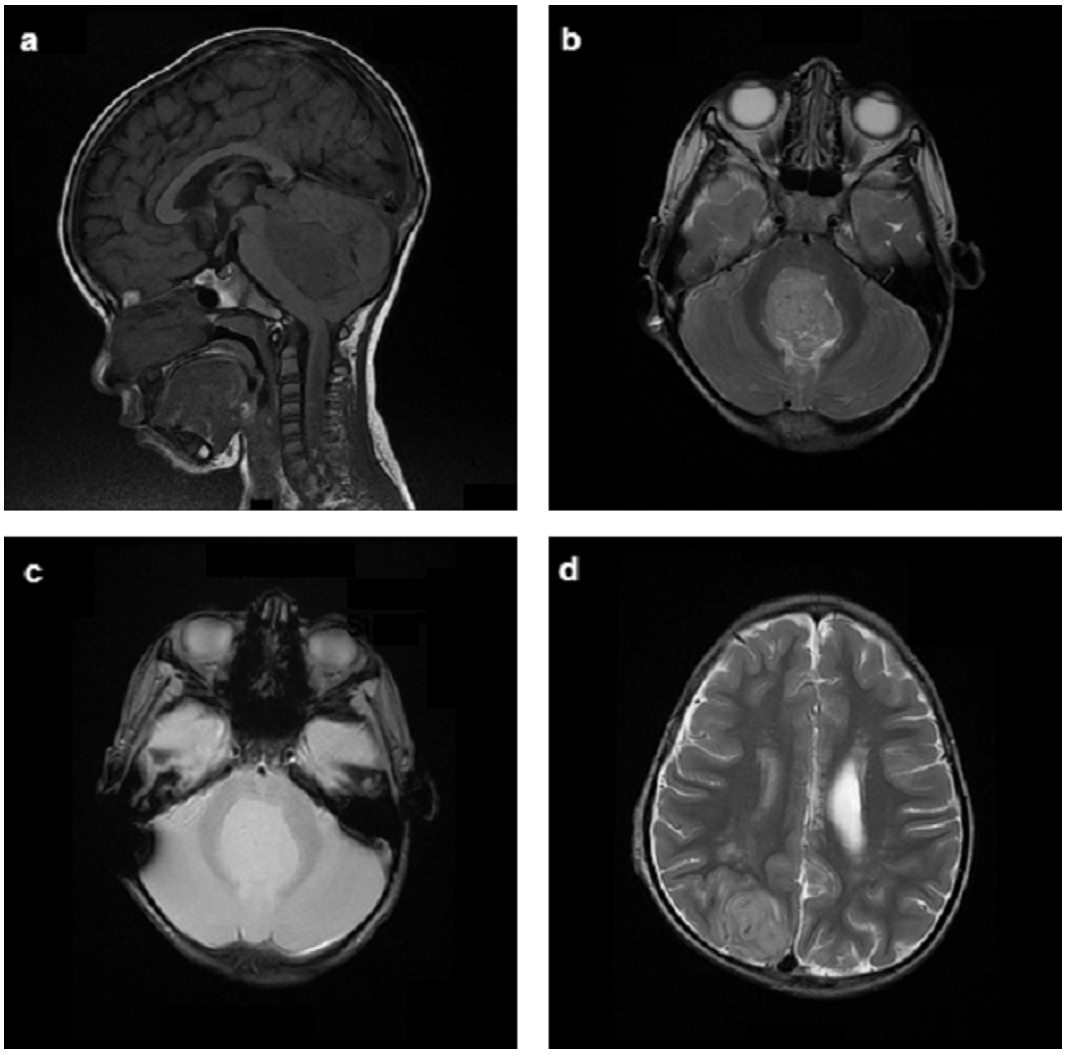

Fig. 1. (a) Sagittal T1-weighted FLAIR image shows a posterior fossa tumour of low intensity with mass effect on the brainstem. (b) Axial T2-weighted image shows a midline tumour of intermediate to high intensity in the 4th ventricle, with bitemporal spread. (c) Axial GRE image did not reveal any hemorrhage. (d) Axial T2-weighted image shows a discrete nodular mass in the right occipital lobe.

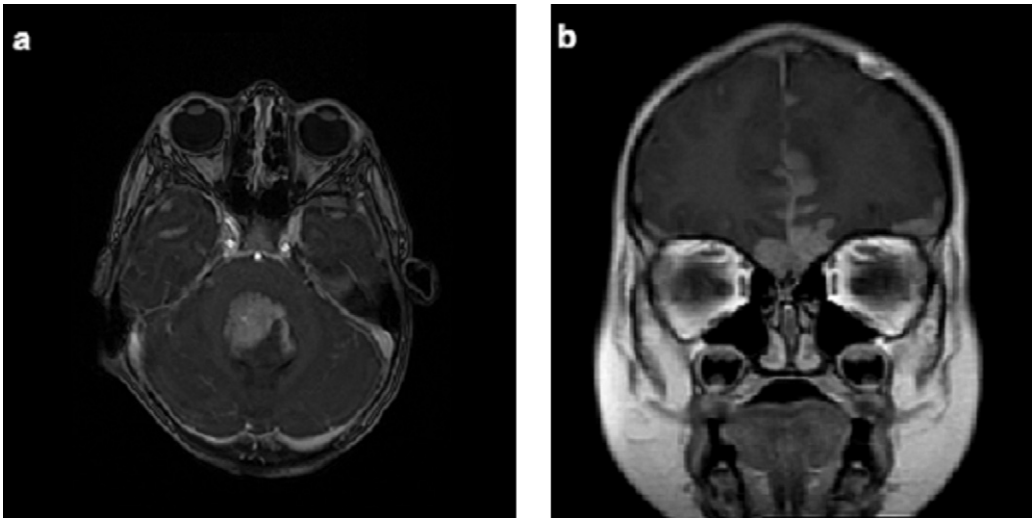

Fig. 2. (a) Axial T1-weighted image after gadolinium enhancement shows intensely enhancing lesions in both temporal lobes, likely to be metastatic deposits. (b) Coronal T1 weighted contrast enhanced image shows enhancement in both frontal lobes. (c) Sagital T1-weighted post-contrast image shows an enhancing left frontal nodule. (d) Coronal T1-weighted contrast enhanced image shows enhancing nodular leptomeningeal spread of the tumour.