PICTORIAL MEDICINE

Medulloblastoma with leptomeningeal metastases

Case history

A 27-month-old previously healthy boy was brought to the hospital presenting with signs of raised intracranial pressure including vomiting, irritability, adducted right eye, and fever. Urgent magnetic resonance imaging (MRI) of the brain showed a small T1- and T2-weighted isointense nodule (Fig 1), which enhanced avidly over the right lateral aspect of the fourth ventricle after intravenous contrast (Fig 2). Features on diffusion-weighted image (DWI) of bright signal intensity and apparent diffusion coefficient (ADC) map of low values (0.67-0.78 x 10^3 mm^2/s) in the abnormal area suggested restricted diffusion of the nodule at the fourth ventricle (Fig 3). In addition, diffuse nodular supratentorial and infratentorial leptomeningeal enhancement was noted (Fig 4). Considering the age of the patient, and the location and characteristic imaging features of the lesion, a diagnosis of medulloblastoma with leptomeningeal metastases was proposed by the radiologists. The diagnosis was later confirmed by excisional biopsy.

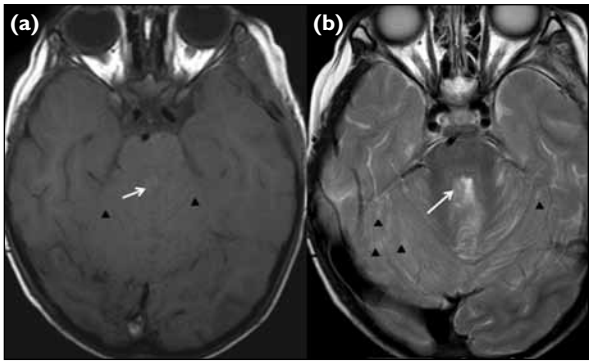

FIG 1. (a) T1-weighted (T1W) and (b) T2-weighted (T2W) magnetic resonance axial images showing a tiny T1W and T2W isointense nodule (arrow) over the lateral wall of the fourth ventricle. There are also T1W isointense and T2W hyperintense nodularities along the cerebellar foliae (arrowheads)

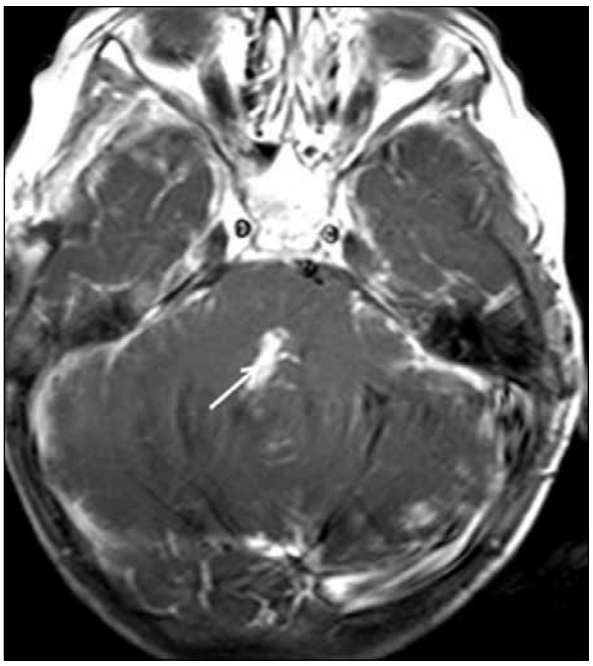

FIG 2. A T1-weighted postcontrast axial image of the nodule over the lateral wall of the fourth ventricle shows avid enhancement after intravenous contrast (arrow)

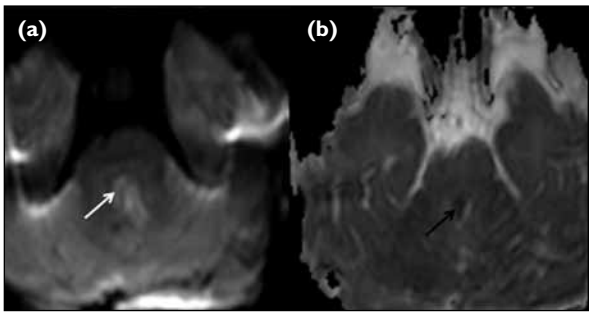

FIG 3. (a) A diffusion-weighted image showing bright signal intensity (arrow). (b) Apparent diffusion coefficient map showing low values (0.67-0.78 x 103 mm^2/s) in the abnormal area (arrow)

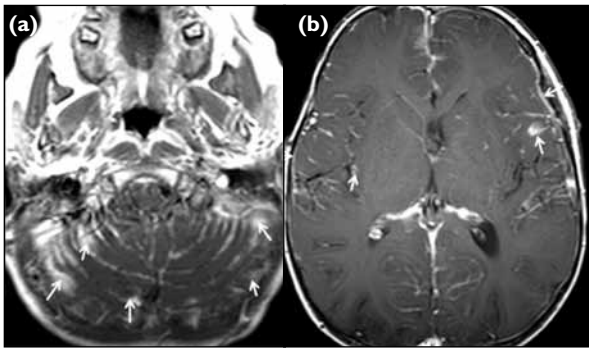

FIG 4. A T1-weighted postcontrast axial images: (a) thick avidly enhancing nodularities are present along the infratentorial meninges (arrows) and (b) supratentorial meninges (arrows) compatible with diffuse leptomeningeal metastases

Discussion

Medulloblastoma accounts for 15 to 20% of all intracranial neoplasms in children. Medulloblastoma is one of the most common malignant central nervous system neoplasms in the paediatric population. Medulloblastoma accounts for up to 38% of posterior fossa tumours.[1] Medulloblastoma typically occurs in the midline and involves the cerebellar vermis in 75 to 93% of patients. Other sites include the cerebellar hemispheres, and the ventricular and supratentorial regions.[2]

Owing to the tightly packed small round cells with a high nuclear-to-cytoplasmic ratio and relatively low interstitial water content, medulloblastoma has a characteristic radiological appearance. Typically, medulloblastomas are hyperdense or isodense on computed tomography, T1- and T2-weighted hypointense or isointense on MRI, and show bright signal intensity on DWI.

Depending on the amount of the cystic or necrotic component, which has been reported in up to 50% of patients with medulloblastoma,[1] the tumour might appear to be completely cystic or solid-cystic. Enhancement might be variably patchy or negligible on postcontrast scans. Leptomeningeal metastasis is not uncommon, and occurs in approximately 33% of medulloblastomas at the time of diagnosis.[3] Diffuse leptomeningeal enhancement is a known feature of the initial MRI presentation of medulloblastoma[4] while, infrequently, isolated leptomeningeal enhancement without a 'primary' tumour mass lesion has been reported.[5]

In children presenting with acute neurological symptoms and diffuse leptomeningeal enhancement on MRI, meningitis or encephalitis are the other important differential diagnoses. However, if the clinical history is atypical of sepsis, such as in this patient, especially when the imaging feature of leptomeningeal enhancement is nodular in appearance, intracranial metastases should be considered. Commonly, intracranial metastases in children might arise from primary posterior cranial fossa neoplasms such as medulloblastoma, ependymoma, or stage IV neuroblastoma.

This report describes the imaging findings of diffuse intracranial metastatic medulloblastomas at presentation. The utility of DWI, which aids characterisation of the primary lesion, hence diagnosis, is also highlighted.

In conclusion, nodular leptomeningeal enhancement in aseptic children presenting with neurological symptoms can be caused by metastases from the primary brain neoplasm in the posterior fossa such as medulloblastoma. Restricted diffusion on DWI and ADC is typical for medulloblastomas due to the intrinsic high cellularity of the tumour.

Darshana D Rasalkar, FRCR
Winnie CW Chu, FRCR, MD
Email: winnie@med.cuhk.edu.hk
Department of Imaging and Interventional Radiology
MK Shing, FRCP
CK Li, MD, FRCPCH
Department of Paediatrics
Prince of Wales Hospital
The Chinese University of Hong Kong
Shatin, Hong Kong

References

1. Koeller KK, Rushing EJ. From the archives of the AFIP: medulloblastoma: a comprehensive review with radiologic-pathologic correlation. Radiographics 2003;23:1613-37.
2. Roberts RO, Lynch CF, Jones MP, Hart MN. Medulloblastoma: a population-based study of 532 cases. J Neuropathol Exp Neurol 1991;50:134-44.
3. David KM, Casey AT, Hayward RD, Harkness WF, Phipps K, Wade AM. Medulloblastoma: is the 5-year survival rate improving? A review of 80 cases from a single institution. J Neurosurg 1997;86:13-21.
4. Meyers SP, Kemp SS, Tarr RW. MR imaging features of medulloblastomas. AJR Am J Roentgenol 1992;158:859-65.
5. Mehta RI, Cutler AR, Lasky JL 3rd, et al. "Primary" leptomeningeal medulloblastoma. Hum Pathol 2009;40:1661-5.