Neurology Asia 2017; 22(2) : 173 – 176

Medulloblastoblastoma with excessive nodularity: Typical imaging appearance

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Medulloblastoma is the most common form of childhood primary brain tumour arising from the cerebellar vermis. It is classified as WHO grade IV embryonal tumours and currently at least four histological variants have been established. Only few case reports been published on the imaging features of the medulloblastoma with excessive nodularity variant. We report the MRI features of a rare case of medulloblastoma with excessive nodularity in a child which is confirmed by histopathology.

CASE REPORT

A previously healthy 9-month-old child presented with irritability, frequent vomiting not associated with meals and developmental regression. Motor assessment showed that the child could only sit with unsteadiness despite adequate support and demonstrated head lag when laid on its back. There was no history of seizure or fever, or any family history of cancer. Physical examination showed normal extremity muscle power, increased reflex in both lower limbs and Babinski reflex was extensor on the right side. No sign of cranial nerve palsy was detected and vital signs were stable.

Contrast enhanced magnetic resonance imaging (MRI) brain (Figures 1 & 2) revealed a large midline posterior fossa tumour that was occupying the right cerebellum. The tumour showed extensive grape-like nodularity in the T2 weighted image, with avid nodular enhancement on post-gadolinium T1 sequence and restricted

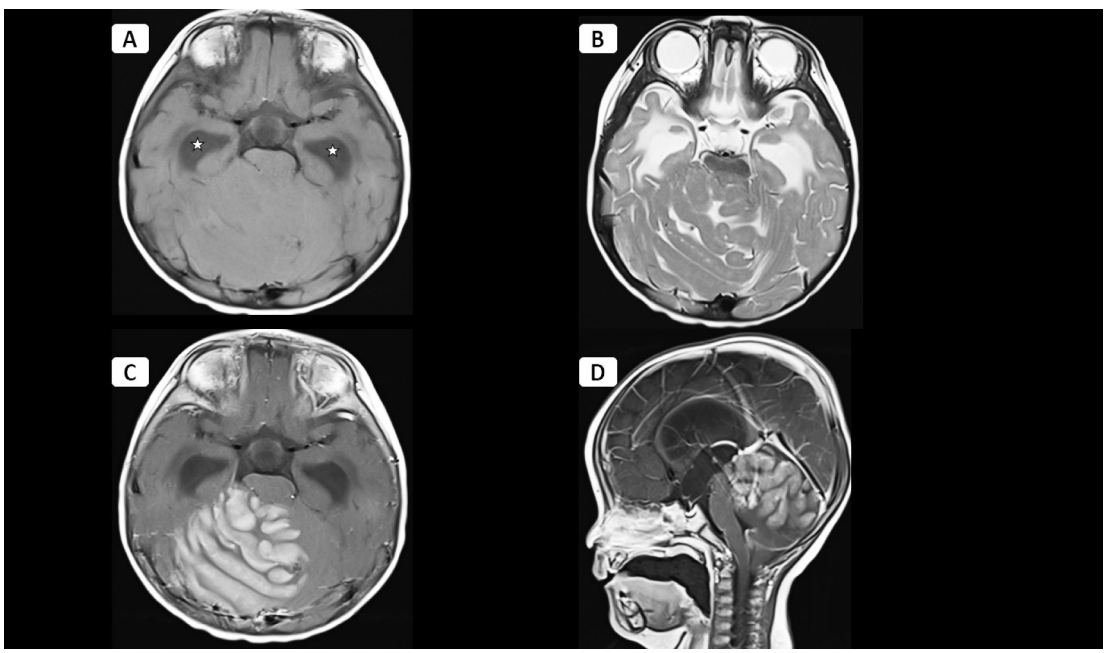

Figure 1: MRI brain images in pre-contrast axial T1W (A), axial T2W (B), post-contrast axial (C) and sagittal (D) T1W sequences showed a mass at midline posterior fossa occupying the right cerebellum. The mass is isointense on pre-contrast T1W (A), hypointense with grape-like nodular appearance on T2W (B) and avid nodular enhancement on post-contrast axial (C) and sagittal (D) T1W images. There was mass effect and bilateral lateral ventricles (star) hydrocephalus due to outlet obstruction at the level of fourth ventricle.

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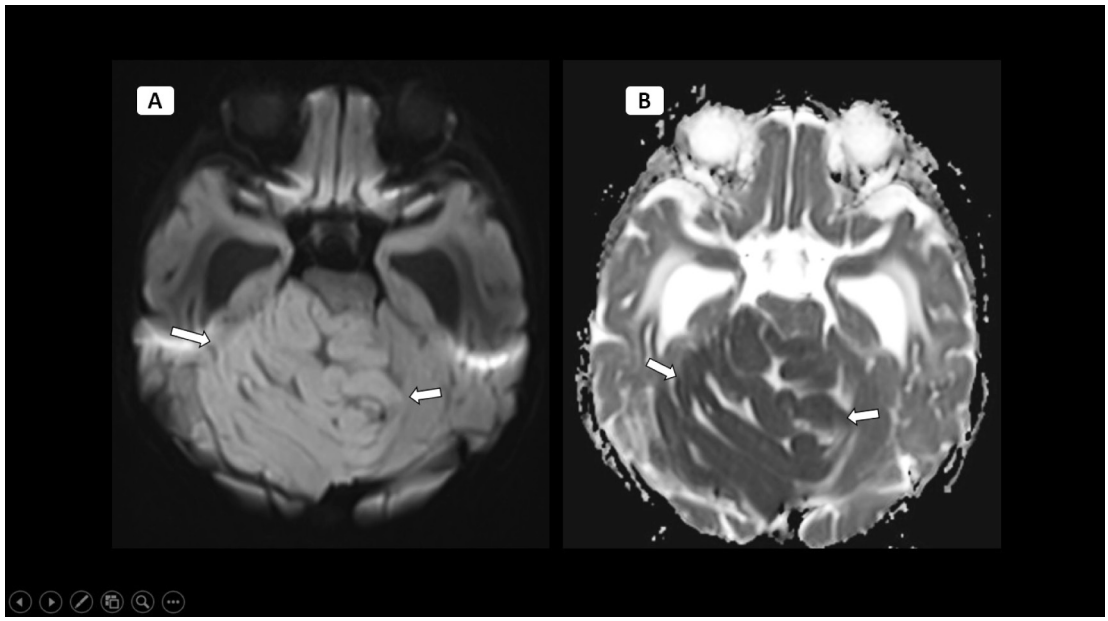

Figure 2: MRI brain images in axial DWI (A) and ADC (B), of the mass showing high signal on DWI and low signal on ADC map, compatible with restricted diffusion indicating high cellularity (white arrows).

diffusion as evident by high signal intensity on diffusion weighted imaging (DWI) and low signal on apparent diffusion coefficient (ADC) (Figure 2). Tumoral mass effect onto the adjacent structures, i.e. right cerebellum, fourth ventricle and brain stem, was noted with congestion of posterior fossa and obliteration of cerebellar vermis. Acute hydrocephalus was also seen due to outlet obstruction at the level of fourth ventricle, for which a ventriculo-peritoneal shunt was urgently inserted by neurosurgeon the next day after the MRI.

Patient underwent elective surgery for tumour resection one week later. Intra-operatively, only about half of the tumour mass could be resected as it was not feasible for complete resection due to high risk of haemorrhage.

Histopathological examination showed tumour compose of primitive-looking cells with round to oval vesicular nucleus and scanty cytoplasm. There is marked nodular appearance with hypocellular pale islands of loosely disposed tumour cells within fibrillary matrix surrounded by more compact tracts of tumour cells in cord (Figure 3). Special histochemistry at the region of hypocellular nodules showed loss of reticulin fibres (Figure 4) and all these findings are consistent with WHO grade IV medulloblastoma extensive nodularity type.

These histology features correlated well with the distinct neuroimaging findings for this subtype of tumour, especially the extensive nodular appearance on T2 and post gadolinium T1 weighted images.

The child remained stable with reduced irritability and vomiting after surgery. Patient was planned for MRI whole spine examination to exclude drop metastasis and referred to Oncology for chemotherapy which has been proven to improve survival for this tumour subtype.

DISCUSSION

Medulloblastoma is classified as a WHO grade IV embryonal tumours and at least four histological variants have been established: classical, desmoplastic/nodular, medulloblastoma with extensive nodularity, large cell, and anaplastic. There are four genetic (molecular) groups of medulloblastoma: WNT (wingless/integrated) activated, sonic hedgehog (SHH)-activated, and the numerically designated "group 3" and "group 4" medulloblastomas.[1] It is important to identify the tumour histological variants as each are associated with dramatic prognostic and therapeutic differences.[2]

Clinical presentation is acute and progression of symptoms is often rapid over a few weeks due to fast growth of these cellular tumours, with dominant symptom of raised intracranial pressure. 95% of cases on non-contrast CT appear as hyperdense midline vermian masses

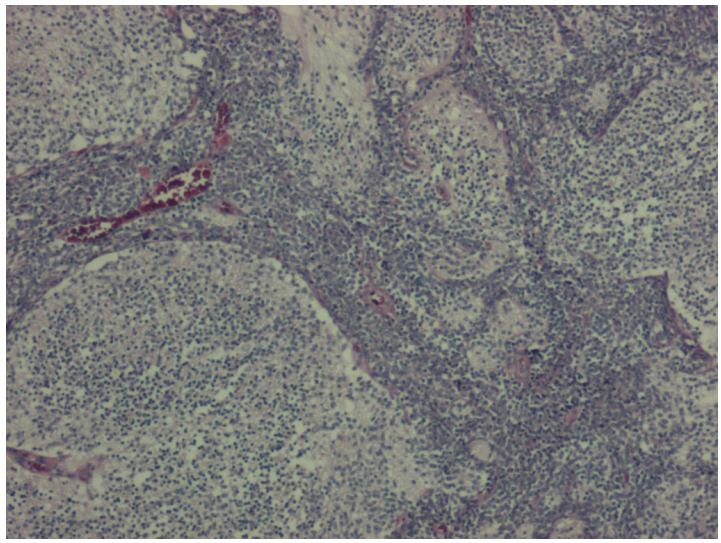

Figure 3: Tumour is composed of primitive-looking cells with round to oval vesicular nucleus and scanty cytoplasm. There is marked nodular appearance with hypocellular pale islands of loosely disposed tumour cells within fibrillary matrix surrounded by more compact tracts of tumour cells (Hematoxylin & eosin stain, x40 magnification)

surrounded by vasogenic oedema with the mass enhancing homogeneously after contrast injection. Hydrocephalus is present more than 90% cases at the time of presentation.

On MRI, a medulloblastoma appear iso- to hypointense relative to white matter on T1-weighted images. Variable T2 signal ranges from hyperintense to hypointense relative to grey matter. Restricted diffusion noted at the tumour with increased DWI signal and associated decreased signal on ADC reflect its consistency of tightly packed, small round cells with scarce cytoplasm and reduced free water. This feature helps in differentiating from other common benign posterior fossa tumours such as juvenile pilocystic astrocystoma and ependymoma.[3,4]

Presence of nodular grape-like appearance may predict a special histological subtype of medulloblastoma (i.e. excessive nodularity subtype) as described in our case. This subtype has a favourable prognosis. Differential diagnosis of this nodular pattern includes Lhermitte-Duclos

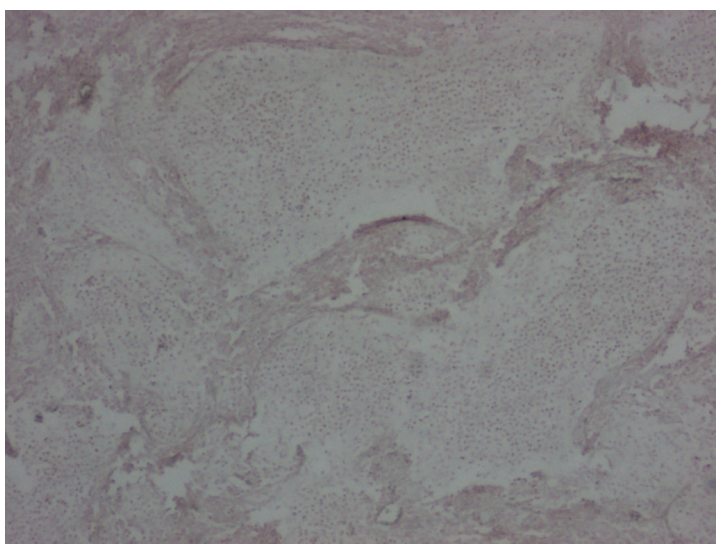

Figure 4: Special histochemistry with reticulin stain shows loss of reticulin fibres in the hypocellular nodules (Reticulin stain, x40 magnification)

disease (also known as dysplastic cerebellar gangliocystoma). The distinguishing feature for MBEN is the avid nodular gadolinium enhancement as oppose to Lhermitte-Duclos disease which rarely enhances.[4-6]

Current standard treatment for medulloblastoma includes surgery, radiotherapy and chemotherapy. The "excessive" nodularity neuro-imaging appearance of this medulloblastoma subtype is important to recognize because of its better prognosis. Giangaspero *et al*. showed that all her medulloblastoma with excessive nodularity patients were alive at a median follow-up of 66 months from the time of diagnosis and almost half of the patient have been cured by chemotherapy alone.[6] Craver *et al*.[7] review of 25-years' experience of diagnosing and treating medulloblastoma variants stated that the child with medulloblastoma with excessive nodularity has the best prognosis compared to other subtypes, with all patients alive, with maximum survival of 19 years (patient was diagnosed when 1-year-old).

In conclusion, medulloblastoma with excessive nodularity is a rare subtype of tumour with distinct imaging features. Our case study highlights this relatively uncommon neuro-radiographic appearance of this tumour. This imaging appearance is recognizable once seen, and may help the pathologist in diagnosis.

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