

LETTER TO EDITOR

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Medulloblastoma with extreme nodularity

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Full Text

Sir,

We report a case of medulloblastoma (MB) with extreme nodularity (MBEN), which was diagnosed preoperatively based on imaging finding.

This 8-month-old girl presented with irritability and progressive increase in head size for 1 month and vomiting for 20 days. Her developmental milestones were delayed. A non-contrast computed tomography (CT) scan of brain showed a well-defined hyperdense vermian mass lesion compressing the fourth ventricle with hydrocephalus [Figure 1]a. The mass was showing striking nodularity in the peripheral part. Post ventriculo-peritoneal shunt placement, contrast-enhanced magnetic resonance (MR) imaging showed a midline vermian mass lesion (59×46×48mm), which had peripheral nodular and central cystic component. The nodular component of the lesion was iso to hypointense on T1-weighted [Figure 1]b and T2-weighted images (WI) [Figure 1]c and d, with restricted diffusion on diffusion-weighted images (DWI) [Figure 1]e, and had dense enhancement on post-gadolinium T1-WI [Figure 1]f. The central cystic part was isointense to CSF and was not enhancing. Based on typical age and neuroimaging finding, a preoperative diagnosis of MBEN was considered. The patient underwent craniotomy with total excision of the tumor. The tumor was nodular and vascular with solid and cystic components. On histopathological examination, the tumor was completely composed of large "reticulin free" nodules with very little internodular component [Figure 2]a and b. Within the nodules, small neurocytic cells were seen "streaming" in a background of neuropil [Figure 2]a. These findings confirmed the diagnosis of MBEN. {Figure 1} {Figure 2}

MBs are the commonest malignant brain tumors in pediatric age group, and despite multimodality treatment, they carry grave prognosis. [1] The MBEN is the newly recognized variant of the MB group included in the WHO classification of the brain tumors in 2007. [2] MBEN is a relatively uncommon variant with an incidence of 4.2%

in one large series. [3] However, in children less than 14 years of age, the incidence increases to as high as 15%, being more than even the desmoplastic variant (12%). [4] MBEN presents at a relatively earlier age (typically <3 years) and carries better prognosis. [5] Distinctive neuroimaging findings of MBEN are nodular appearance of the solid component, classically described as being "grape like". [2] These nodules are composed of compactly packed cells and are reticulin-free zones that represent zones of neuronal maturation, with reduced nuclear: cytoplasmic ratio, fibrillary matrix and uniform cells with a neurocytic appearance. These nodules show restriction on DWI with low ADC values due to densely packed cells. [6] The frequency of metastasis in these patients is also much less (along with desmoplastic variant) as compared to other variants. [4] This factor along with the presence of relatively "mature" neuronal components in the tumor appears to be responsible for a better prognosis. [5] Better response to chemotherapy and radiotherapy and lesser incidence of tumor relapse has also been noted. As a result of these factors, patients with MBEN have better survival rates.

The report highlights the preoperative diagnosis of MBEN based on imaging findings. This may help to better prognosticate and decide further adjuvant radiotherapy and chemotherapy.

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