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CASE REPORT

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Medulloblastoma with extensive nodularity: US, CT and MRI findings

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Summary

Background: Medulloblastoma accounts for up to 25% of all paediatric CNS tumours. According to WHO classification (2007) medulloblastoma with extensive nodularity (MBEN) is a separate rare entity associated with younger age and better prognosis.

Case Reports: A 9-month-old girl was admitted and examined because of macrocephaly and disturbed psychomotor development. Transfontanel ultrasound revealed dilated ventricular system and hyperechoic mass in the posterior cranial fossa. Computed tomography showed hyperdense mass in the cerebellum. Magnetic resonance imaging revealed a mass with gyriform pattern and strong contrast enhancement after gadolinium administration. Differential diagnosis included dysplastic heterotopic cortex, Lhermitte-Duclos disease, atypical teratoid/rhabdoid tumour (AT/RT), and medulloblastoma (MB). The patient was operated on. Medulloblastoma with extensive nodularity (MBEN) was finally diagnosed.

The authors present and discuss three other cases of this rare entity.

Conclusions: Transfontanel sonografic examination is capable of detection of the posterior fossa tumour as a cause of hydrocephalus and macrocephaly. The mass in a child's posterior cranial fossa that is hyperdense on unenhanced CT and gyriform, nodular, and markedly enhancing on MRI may strongly suggest medulloblastoma with extensive nodularity (MBEN).

Key words: **medulloblastoma with extensive nodularity (MBEN) • ultrasound (US) • computed tomography (CT) • magnetic resonance imaging (MRI)**

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Background

Medulloblastoma belongs to the group of embryonal tumours and is the most common malignant tumour of the central nervous system (CNS) in children, accounting for up to 25% of all paediatric CNS neoplasms [1]. According to WHO classification (2007) medulloblastoma with extensive nodularity (MBEN) is a separate entity associated with younger age and better prognosis [2–6]. The imaging descriptions of this rare entity are not numerous. Textbooks and most papers in the literature show classic images of medulloblastoma: hyperdense mass on unenhanced CT scan and well-defined tumour, iso- to hypointense on T1-weighted images and with variable signal intensity on T2-weighted images, with surrounding oedema and contrast enhancement [1,7]. We present the sonografic, CT and MRI features of a histologically proven MBEN.

Case Report

A 9-month-old girl of the Caucasian race was born after unremarkable pregnancy as a second child to the healthy unrelated parents. Head circumference at birth was large

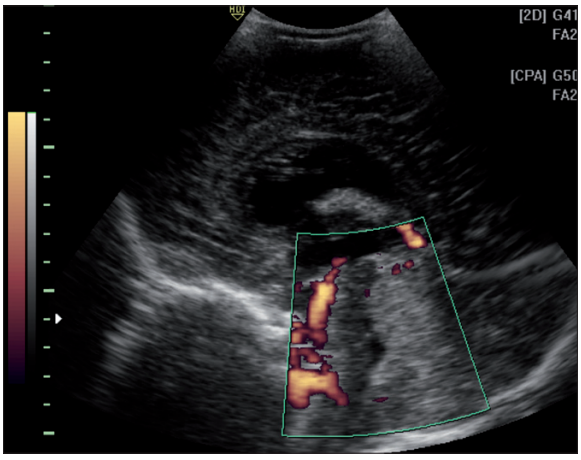


Figure 1. Transfontanel US. Ill-defined hyperechoic mass in the cerebellar vermis.

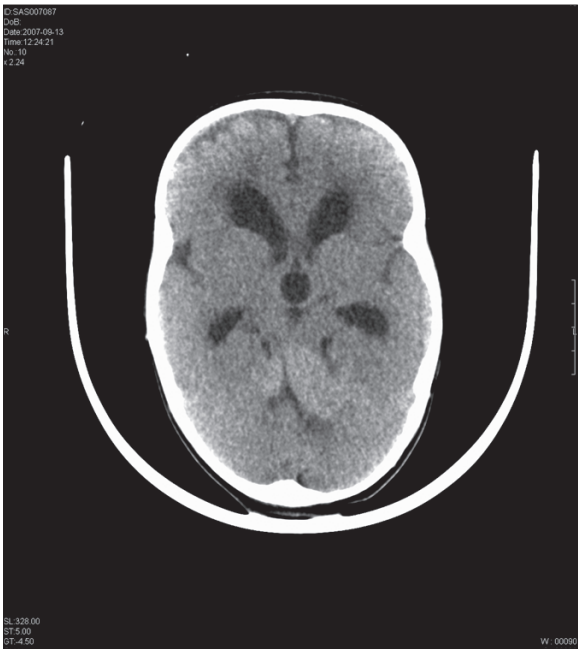


Figure 2. Non-contrast CT. Hyperdense lesion compressing the quadrigeminal plate. Dilated supratentorial ventricular system.

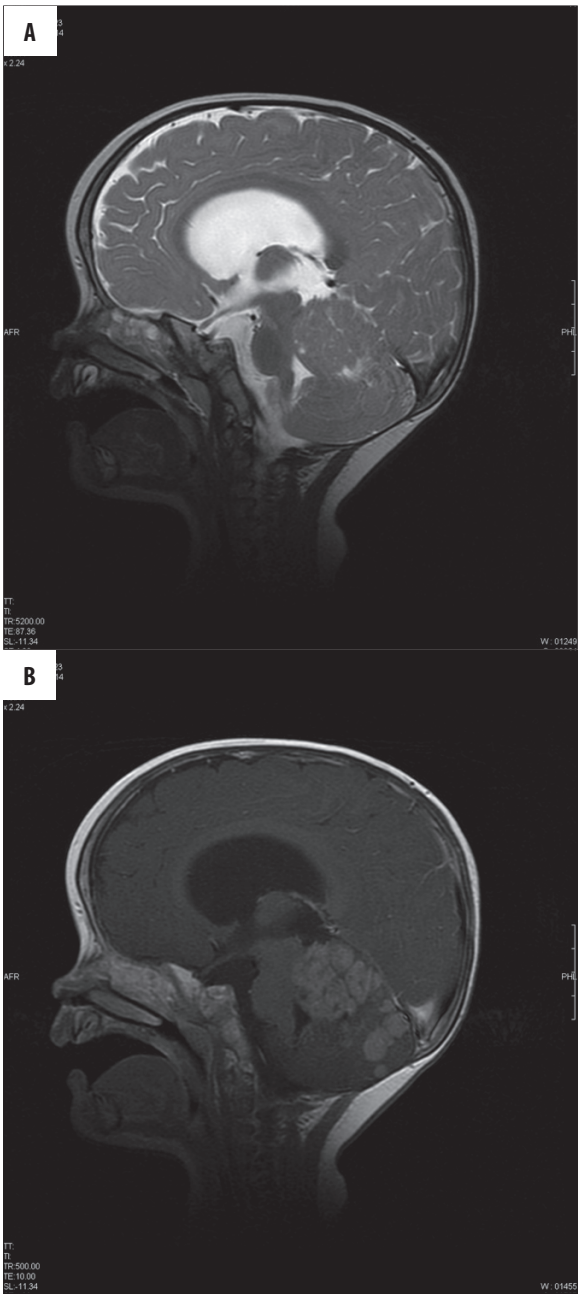


Figure 3. MRI. (**A**) Sagittal T2WI. Nodular lesions in the vermis, following signal intensity of the cortex, with gyral pattern. (**B**) Sagittal T1WI. Contrast enhancement of the mass after gadolinium administration.

and the rate of head growth was excessive. Transfontanel sonography was performed in the first, second and fourth month of life to exclude hydrocephalus. The size of the ventricles was normal. Psychomotor development was abnormal, especially as far as gross motor function was concerned. After 8th month of life she was admitted to the Department of Neurology of the Institute of Mother and Child in Warsaw because of excessive macrocephaly. Dilated ventricular system was found with the depth of the lateral ventricles of 18 mm on the right side and 17 mm on the left at the level of Monro foramen, 3rd ventricle of 13×16 mm and Evans index of 0.46. An ill-defined hyperechoic mass was visible in the posterior cranial fossa on the left, compressing the aqueduct and 4th ventricle (Figure 1). Further diagnostic process was necessary and CT scan was performed showing irregular hyperdense lesion in the vermis and both cerebellar hemispheres (Figure 2). Contrast medium was not administered because the decision was made to perform MRI. The girl underwent this examination under anaesthesia in a 1.5 T scanner. MRI revealed a lobulated mass that followed signal intensity of the cortex on all the sequences: SE/T1, volumetric 3D/T1/SPGR, T2flair and FSE/T2 (Figure 3A). Only a very narrow trace of oedema was visible in three slices on T2flair and FSE/T2WI in the surrounding cerebellum. After gadolinium administration the mass enhanced strongly and homogenously showing nodular pattern of the lesion (Figure 3B). The differential diagnosis included dysplastic heterotopic cortex, atypical teratoid/rhabdoid tumour, medulloblastoma. The patient

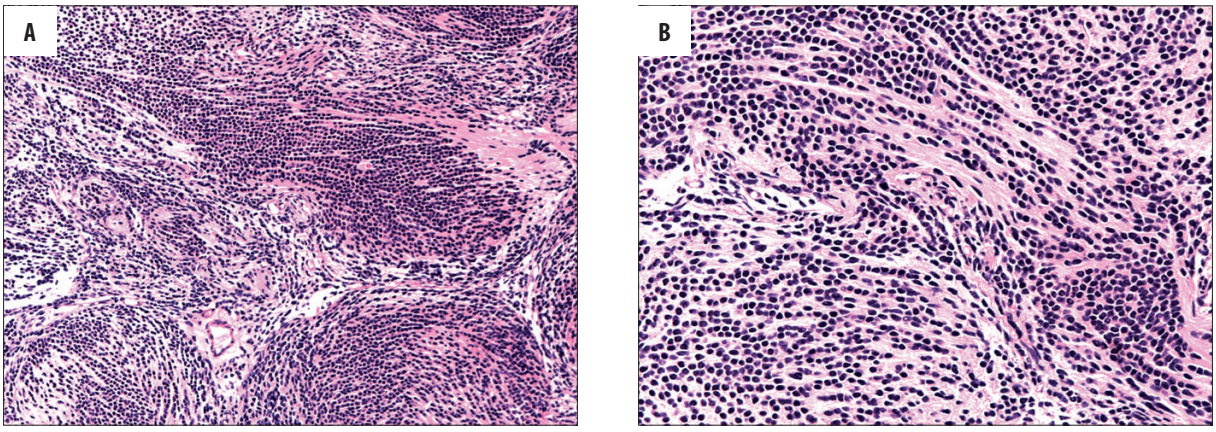


Figure 4. Pathological specimens. (**A**) Expression of nodular pattern rich in neuropil-like zones. (**B**) The streaming pattern of neurocytic-like cells in reticulin-free zones.

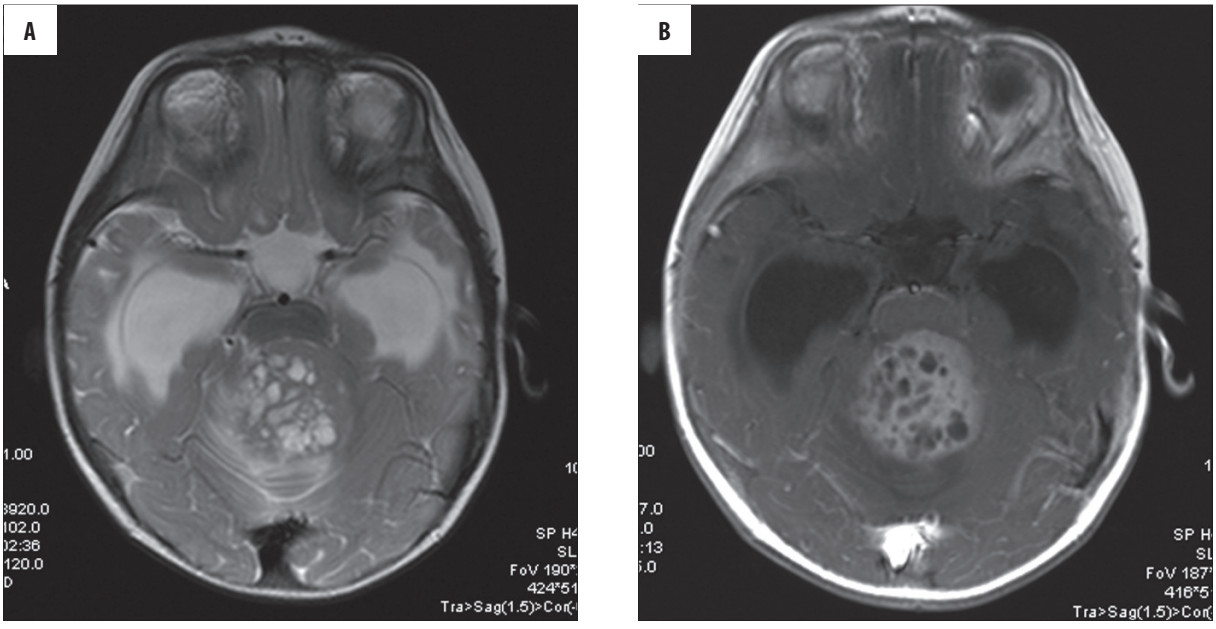


Figure 5. MRI of the retrospectively reviewed 9-month-old boy. (**A**) Axial T2WI. The lesion displays mixed signal intensity, with hypointensity similar to the cortex and hyperintensity representing tiny cysts. (**B**) Axial T1WI. Contrast enhancement of the tumour is inhomogenous due to its cystic parts.

was operated on at the Department of Neurosurgery, the Children's Memorial Health Institute in Warsaw, with partial resection of the tumour. Medulloblastoma with extensive nodularity (MBEN) was finally diagnosed (Figure 4A,B).

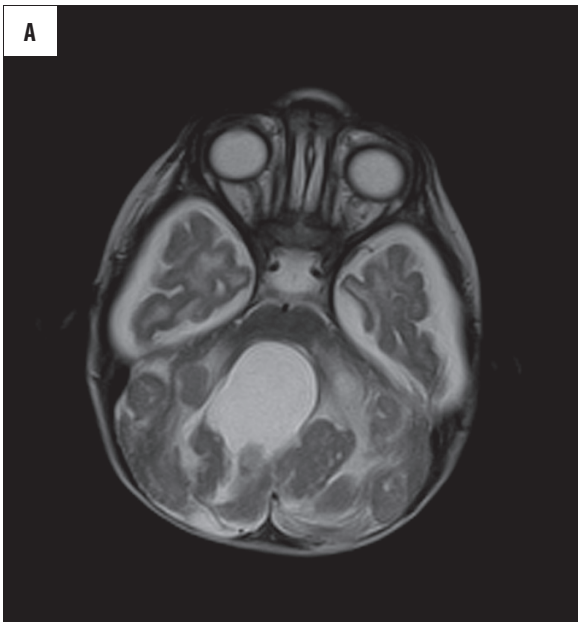
Discussion

Medulloblastoma with extensive nodularity is a rare type of medulloblastoma accounting for from less than 1% to more than 5% of all cases according to various authors [1,6] and its imaging features are not commonly known. Looking for similar images while preparing the list of differential diagnosis, we did not find such a tumour in textbooks. To our knowledge there are only very few descriptions of CT and MRI findings in cases of MBEN and no description of transfontanel sonographic features of this tumour.

On US the mass showed hyperechogeneity and was ill-defined. One has to keep in mind that visualisation of the lesion was difficult because of its location but its bigger part in the left cerebellar hemisphere was clearly depicted. Compression of the aqueduct and 4th ventricle was noted.

Hyperattenuation on unenhanced CT images and grape-like architecture on both CT and MR images are stressed in these few papers that deal with this rare entity [8–10]. It was observed in our case as well. But the most striking feature of the lesion on MR images was its gyriform pattern with signal intensity following that of the grey matter on all the sequences, including volumetric 3D/T1/SPGR, dedicated to the assessment of cortical anomalies. We found only two papers in the literature, stressing gyriform morphology of MBEN [11] and its isointensity to the cortex [8]. Otherwise similar case of MBEN described by Naitoh et al. [8] was different from ours in terms of the extent of the oedematous zone which was apparently bigger, while in our case only a trace of oedema around the tumour was observed.

We retrospectively analysed three previous cases of MBEN diagnosed at the Children's Memorial Health Institute in



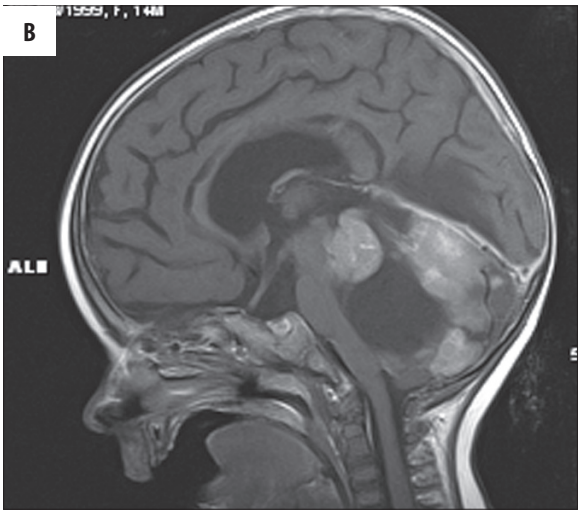


Figure 6. MRI of the retrospectively reviewed 14-month-old girl. (**A**) Axial T2WI. Big cystic portion of the mass. (**B**) Sagittal T1WI. The cyst within tumour does not enhance with gadolinium.

Warsaw in years 2000–2007. There were two boys and one girl in this group, aged 7, 9 and 14 months at the moment of MRI examination. These tumours were bigger at presentation and – although their signal intensity was in general similar to that of the grey matter – showed inhomogeneity with focal hypointensity on T1WI and hyperintensity on T2WI, corresponding to smaller or bigger cysts within the mass (Figures 5A,6A). As expected these foci did not enhance with gadolinium (Figures 5B,6B) while in the case presented above signal intensity and contrast enhancement of the tumour were homogenous. The zones of oedema in the reviewed cases were remarkably more extensive although still not very big, taking into account the diameter of tumours, the degree of compression of the IV ventricle and the aqueduct and degree of hydrocephalus. All four tumours shared grape-like architecture but the gyriform pattern was the most striking in our main case.

Less than 5% of MBs are very advanced in nodularity and reveal signs of neuronal differentiation. They usually present in young children and are associated with a good clinical outcome [5,12].

Histologically MBEN displays a lobular architecture with large elongated reticulin-free zones. These zones are rich in neuropil-like tissue and contain small round neurocytic-like cells (Figure 4A,B). The internodular components may be markedly reduced in some areas.

The favorable outcome is probably related to spontaneous neurocytic differentiation of this type of MB [10].

Conclusions

Transfontanel sonografic examination is capable of detection of the posterior fossa tumour as a cause of macrocephaly and hydrocephalus. The mass in a child's posterior cranial fossa that is hyperdense on unenhanced CT and gyriform, nodular, and markedly enhancing on MRI may strongly suggest medulloblastoma with extensive nodularity (MBEN).

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