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Case Report

A rare case of pediatric medulloblastoma presented with radiographic characteristics of pontine glioma

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ABSTRACT

Medulloblastoma is a common malignant brain tumor of childhood which characterized as an infratentorial mass in the roof of the 4th ventricle, causing mass effect and non-communicating hydrocephalus. A 3 years old boy with history of chronic progressive weakness one-month prior admission, followed by difficulty in walking and ataxia. His MRI examination showed an intraaxial well defined mass, partial irregular border, size 4.2x4.0x4.1 cm at pons, hypointense in T1W1, hyperintense in T2W1, iso-hypointense in flair, and showing inhomogen contras enhancement after gadolinium administration. It pressed the 4th ventricle without dilatation of other ventricles, suggesting a pontine glioma. Patient underwent surgery to remove mass and the pathological examination revealed small round cells, with differential diagnose of medulloblastoma and anaplastic astrocytoma. This case presented as there was a discrepancy in the clinicoradiological diagnosis with the pathological diagnose of medulloblastoma. The mass was located at the pontine without any sign of hydrocephalus suggested a pontine glioma. Both medulloblastoma and pontine glioma are common for infratentorial mass of pediatric population, but difficult to diagnose due to its heterogeneous presentation. Good comprehension of both radiological characteristics is noteworthy.

Keywords: Medulloblastoma, Magnetic resonance imaging, Pontine glioma, Radiographic

INTRODUCTION

Medulloblastoma (MBs) is a common malignant brain tumor of childhood which characterized as an infratentorial mass in the roof of the 4th ventricle, causing mass effect and non-communicating hydrocephalus.[1,2] However, this tumor can be rarely found outside its predominant location, some in cerebellopontine angle and one case founded in cerebellar hemisphere mimicking other tumor presentation.[3] Adding to this point, the clinical manifestation is closely related to the location of mass, with most common findings are ataxia accompanied by spasticity and gait instability, making it hard for diagnosis.[1] Medulloblastoma is mainly a pediatric brain tumor that account for 10% of all intracranial neoplasms and 29% of all pediatric fossa tumors in children, whereas they represent less than 1% of CNS adult neoplasms. Most MBs are located in the vermis in children and in the cerebellar hemispheres in adults.[2] Typical and atypical MRI features have been reported in MBs: most of them present with well-defined margins, iso- and hyperintensity signal respectively on T1 and T2-images and marked enhancement on T1-postcontrast images. In rare cases they present with ill-defined margins, hypo intensity signal on T2, mild or nodular enhancement.[3]

CASE REPORT

A 3-years old boy presented with chronic progressive weakness one month prior admission, followed by difficulty in walking and ataxia. His vital sign and

physical examination are within normal range. No developmental delay or familial history.

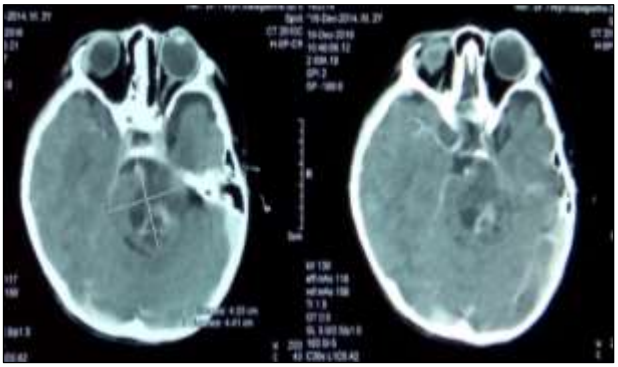

Figure 1: Head CT scan with contrast administration suggesting a solid mass in the pontine.

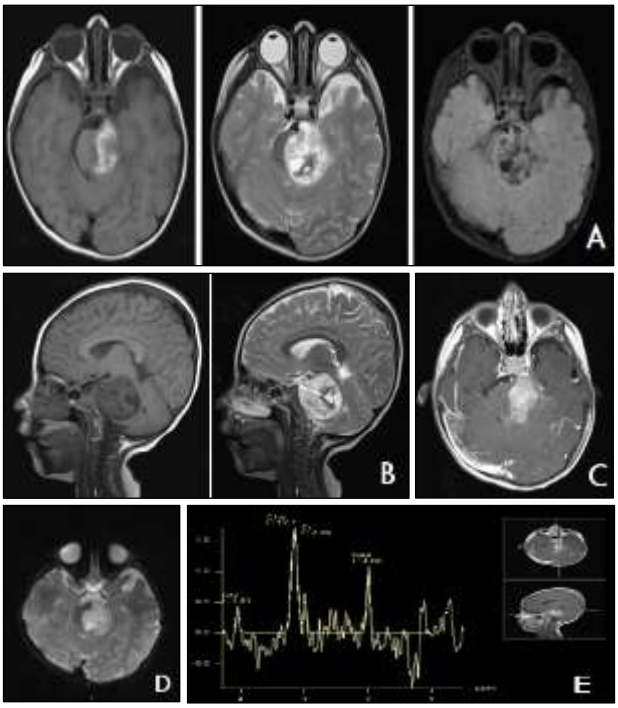

Figure 2: Brain MRI examination. (A) Axial MRI showing well defined mass, partial irregular border at pons, hypointense in T1W1, hyperintense in T2W1, iso-hypointense in flair, pressing the 4th ventricle. (B) Sagittal MRI showing well defined mass, partial irregular border at midbrain until pons, hypointense in T1W1, hyperintense in T2W1. Please note there are no dilatation of ventricles system. (C) Gadolinium injection shows inhomogeneity enhancement of solid component. (D) DWI demonstrates restricted diffusion in the solid portions of this tumor. (E) MR spectroscopy of tumor. Arrows point to the resonance doublet of lipid lactate, reflecting the anaerobic metabolism characteristic of this aggressive tumor.

No abnormality in laboratory studies. A head Computed Tomography (CT scan) was performed in revealing an isodense and hypodense well-defined solid mass, on pons, size 4.3x4.4 cm with heterogenous contrasts enhancement after contrast administration (Figure 1).

He underwent a brain Magnetic Resonance Imaging (MRI) examination. It showed an intraaxial well defined mass, partial irregular border, size 4.2x4.0x4.1 cm at pons, hypointense in T1W1, hyperintense in T2W1, iso-hypointense in flair, and showing inhomogen contras enhancement after gadolinium administration. It pressed the 4th ventricle without dilatation of other ventricles, suggesting a pontine glioma (Figure 2).

Diffusion-Weighted Imaging (DWI) characteristically demonstrates restricted diffusion within tumor (Figure 2D). MR spectroscopy is frequently employed in the preoperative evaluation of tumor. These tumors show a characteristically aggressive metabolite spectrum, with marked elevation of choline and suppression/absence of N-Acetyl-Aspartate (NAA) (Figure 2E). Lipid and lactate elevation can be seen as a manifestation of anaerobic metabolism in the tumor.

Patient then underwent surgery to remove mass. A well-defined mass locating at pons is seen and total surgical excision was performed. Histological examination of tumor consisted of small round cells (Figure 3), with differential diagnose are medulloblastoma and anaplastic astrocytoma. After further discussion and confirmation, the final histological result is confirming MBs. The patient is still under the surgeon care for further treatment.

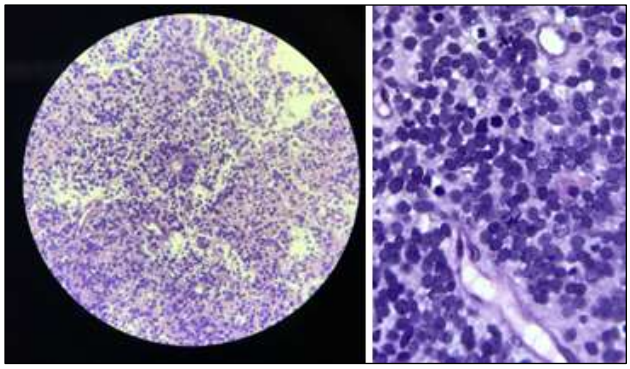

Figure 3: Small round cells of histopathology slide (H and E staining) a striking feature of medulloblastoma.

DISCUSSION

Medulloblastoma is a predominantly pediatric tumor which arises from vermis commonly occurring in the in the roof of the 4th ventricle as the most common site of origin.[4-6] It has variation in location that may be explained by the different cellular origin, however MBs is dominated by symptoms of raised intracranial pressure as a result of obstructive hydrocephalus.[7] This case presented as there was a discrepancy in the clinic-radiological diagnosis with the pathological diagnose of MBs.

The mass was located at the pontine without any sign of hydrocephalus suggesting a pontine glioma instead of MBs, because of their radiographic features that are nearly the same, such as hypo intensity in T1W1, hyperintensity in T2W2, and heterogenous enhance in T1W1+contrast.[2]

From literatures, author know that MBs arise from germinal cell (or their remnants) anywhere along their migratory path.[3] It may grow to occupy CPA by lateral extension from the 4th ventricle through foramen Lushka, or it may do direct exophytic growth from the site of the surface of cerebellum or pons.[8]

In this report, author present a rare case of primary MBs occurring in the pons mimicking a pontine glioma, with confirmation from histological finding of small round cells a striking feature of MBs. Medulloblastoma located outside its predominant location in the midline is rare, and only 33 cases have been previously reported.[2] Most of the reported cases occurred at the CPA and only 1 case reported in the cerebellar hemisphere.[3,8] Typical and atypical MRI features also have been reported in MBs, which in rare cases may present with ill-defined margins, hypo intensity signal on T2, mild or nodular enhancement.[3]

As described above, MBs can occur far from the roof of 4th ventricle, and clinically it is very difficult to distinguish these neoplasms from another lesion. Although it is extremely rare to be found outside its predominant location, author should always consider a MBs in the differential diagnosis of pediatric brain tumor.

Table 1: Differential diagnosis of posterior fossa tumor in children.

	Medulloblastoma	Diffuse pontine glioma	Ependymoma	Juvenile pilocytic astrocytoma
Signal intensity characteristics (on T2-Wi)	Homogenous; low to moderate intensity	Ill defined; high, commonly cystic	Markedly heterogenous intensity	Sharply demarcated; solid portion; high signal
Contrast enhancement	Common; dense	Variable	Common; irregular	Common in solid portion (mural nodule)
Calcification	Uncommon	Rare	Common	Uncommon
Hemorrhage	Uncommon	Uncommon	Common	Rare
CSF pathway	High	Low	Low to moderate	Extremely low

Table 1 shows radiographical differential diagnosis of posterior fossa tumor in children. Posterior fossa tumor in pediatric population can be differentiated by its signal intensity characteristic, contrast enhancement pattern, calcification, incidence of hemorrhage, and CSF pathway. Great variability in imaging characteristics makes it difficult to distinguish each type of tumor.

CONCLUSION

Medulloblastoma can occur far from the roof of 4th ventricle, and clinically it is very difficult to distinguish this neoplasm from another lesion. Radiologist must understand that brain tumor may not follow the normal pattern as written in the existing literature. Both MBs and pontine glioma are common for infratentorial mass of pediatric population, but difficult to diagnose due to its heterogeneous presentation. Good comprehension of radiological characteristics is noteworthy, and author should always consider MBs in the differential diagnosis of pediatric brain tumor. Definitive diagnosis requires histologic confirmation in all cases.

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