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Medulloblastoma with atypical dynamic imaging changes: A case report with literature review

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Objective We analyzed a case of medulloblastoma with atypical dynamic imaging changes retrospectively to summarize the atypical MRI features of medulloblastoma by reviewing the literature.

Methods One atypical case of medulloblastoma in the cerebellar hemisphere confirmed by pathology was analyzed retrospectively, and the literature about it was reviewed.

Results The radiologic findings of the patient were based on three examinations. The first examination showed that the cortex of the bilateral cerebellar hemisphere had diffuse nodular thickening, with a high signal on DWI imaging and significant enhancement. Contrast enhancement MRI 1 year later showed the signal of cerebellar hemisphere returned to normal, but revealed an enhanced nodule. A re-examination 6 months later showed an irregular mass with a high-density shadow in the cerebellar vermis on CT scan, the T2-weighted image revealed multiple degenerative cysts and the mass was significant enhancement.

Conclusion The radiologic characteristics of atypical medulloblastomas vary in adults and children. Understanding the radiologic characteristics of medulloblastomas, such as MRI features, age of onset, and location of atypical medulloblastomas, can help improve the diagnosis of medulloblastomas.

Key words Medulloblastoma, Magnetic resonance imaging, Atypical

Introduction Medulloblastomas are the most common malignant tumors in children. The posterior fossa, such as the cerebellar vermis and 4th ventricle, are typical locations for medulloblastomas⁴. The classic imaging of medulloblastomas has been widely reported⁵. Indeed, a correct diagnosis of medulloblastoma can be made based on the age of onset, location, and typical imaging on MRI, but some atypical manifestations, such as irregular enhancement, locations other than the cerebellar vermis, and cyst formation, may initially lead to a misdiagnosis. In our study, we report and analyze an atypical medulloblastoma with dynamic imaging changes and summarize the atypical MRI features of medulloblastomas to improve the diagnostic accuracy.

Case Report A 4-month-old boy was admitted to our institution for evaluation of an inability to gaze upward. He underwent three examinations with time. The first examination showed the cortex of the bilateral cerebellar hemisphere had diffuse nodular thickening with a high density on CT scan (the image of it was lost for some reason) and high signal on DWI imaging (Fig. 1A). The MRI enhancement scan showed that the cortex of the cerebellar hemisphere enhanced significantly and homogeneously (Fig. 1B). Considering that the lesion was high density on CT scan, we made a presumptive diagnosis of neoplastic lesion, and in combination with the age of the patient, we thought the greatest possibility was medulloblastoma. Because of the lesion was...
too diffuse to operated on by surgeon, the family decided to follow up without treatment. Contrast enhancement MRI 1 year later showed that the cerebellar hemisphere signal returned to normal, with the exception of an enhanced nodule (Fig. 2). The development process was beyond our imagination, but we insisted diagnosis as neoplastic lesion. Meanwhile we could not help thinking why and how the lesion developed like this without any treatment. He was rechecked 6 months later and there was an irregular mass with a high-density shadow in the cerebellar vermis on CT scan (Fig. 3A). The CT scan showed isointensity on the T2-weighted image with multiple degenerative cysts (Fig. 3B) and the mass was significant enhanced (Fig. 3C).

The pathologic examination showed that the tumor cells were composed of a large number of nodules with an abundance of neutrophils. The cells were consistent with the characteristics of nerve cells and mitotic figures were rare (Fig. 4). A medulloblastoma with extensive nodularity was diagnosed on the basis of these findings.

**Discussion** Medulloblastomas account for 1.5% of brain tumors and appear mostly in children born before 15 years of age. The origin of the tumor is controversial\(^{[3,4]}\). Most researchers believe that typical medulloblastomas originate from the external granular layer at the top of 4th ventricle, which is the main reason why the cerebellar vermis and 4th ventricle are the typical locations for medulloblastomas occurred in children. With the development of genomic approaches and molecular detection technology, researchers divided medulloblastoma into at least 4 subgroups (WNT, SHH, Group 3, and Group 4)\(^{[5]}\). We still used the old pathological classification in this case.

Medulloblastomas are generally high density on CT scans. T1WI on MRI is slightly low or an equal signal, T2WI is slightly high or an equal signal. The signals are always homogeneous and occasionally have multiple cystic changes. DWI and enhancement of medulloblastomas are significant to distinguish medulloblastoma from other tumors occurring in the posterior fossa, such as pilocytic astrocytomas. In this case, the imaging findings during the third follow-up were typical, but the first examination showed that the cortex of the bilateral cerebellar hemisphere was diffuse nodular thickening and the enhancement was significant. Follow-up 1 year later showed that the signal of the cerebellar hemisphere returned to normal, but an enhanced nodule was noted. The development process is extremely atypical, and it is difficult to make a correct diagnosis before surgery.

Based on a review of the English literature, there are no reports involving medulloblastoma development similar to the current case. We put forward the following three possibilities to explain this manifestation. The first one is medulloblastoma may have more than one origin, bilateral cerebellar hemispheres have embryonic cells of multi-centric origin in the external granular layer which could develop to medulloblastoma with diffuse lesions. Second, researchers have shown that the postnatal cerebellum contains multipotent neural stem cells\(^{[6]}\), their normal development and the failure of these progenitor cells to undergo program cell death, may potentially result in neoplastic cell genesis and medulloblastoma development. Furthermore, their expression of CD133 increases invasion\(^{[7]}\). Third, classic medulloblastomas were thought to originate from the ventricular zone, we hypothesize the origin of medulloblastomas with extensive nodularity may different from classic medulloblastomas. A Chinese doctor Liu\(^{[8]}\) reported a case of an atypical medulloblastoma with extensive nodularity in an adult. The MRI images showed diffuse abnormal signals in the cerebellar vermis, bilateral cerebellum, and left part of the brachium pontis, with a slightly long T1, long T2, and marginally high signal in DWI.
The diffuse lesions of the tumor is similar to our case, and the histopathological diagnosis was consistent with it. Medulloblastomas with diffuse lesions are surprisingly rare, we can only make assumptions from the limited information, the exact basis of pathology needs more samples with comparative observation and further analysis.

We herein review the literature and summarize the imaging manifestations of atypical medulloblastomas. (1) The cerebellar vermis and 4th ventricle are typical locations for medulloblastomas, but the location could deviates from the midline. Medulloblastomas occur more commonly in an off-midline location such as cerebellar hemisphere among older children and adult population\(^{[9,10]}\). Some scholars speculate that’s because medulloblastomas originate from granular layer of the cerebellar cortex\(^{[11,12]}\). (2) Desmoplastic medulloblastomas could extend to the meninges occasionally and lead to reactive hyperplasia of meninges\(^{[13]}\). If it shows "dural tail sign", we could more inclined to misdiagnose a medulloblastoma as a meningioma. Pinpointing is the key to distinguish they two. A mass of medulloblastoma which was located on the surface of the cerebellar hemisphere and protruding into the cerebellopontine angle seems to be located outside the brain, it is important to observe the relationship between the tumor, cerebellar hemisphere, and dura carefully, and then to determine the tumor is intra- or extra-cerebral. (3) Tumors could show multiple nodular enhancements in the shape of "a grape"\(^{[14]}\), which are most often seen in medulloblastomas with extensive nodularity. I just presented a case of medulloblastoma with extensive nodularity, the lesion of which resembled "a bunch of grapes" on the initial examination, but only a nodule with abnormal signal left after 1 year. It conforms to the performance of atypical medulloblastomas to some extent, but the development was unexpected. The case showed different characteristics from we had ever seen before, the different characteristics not only enriched our clinical experience, but also produced new challenges for the pre-operative diagnosis of the disease. (4) It is rare to have cystic degeneration or necrosis in medulloblastomas occurring in children because of the abundant blood supply, but they can always be seen in adults\(^{[15]}\). (5) The tumor is enhanced significantly in children, while it shows mild enhancement or no enhancement in adults, which makes it difficult to ensure there is no tumor residual after surgery.

In conclusion, atypical medulloblastomas are not uncommon in older children and adults. It is difficult to distinguish medulloblastomas from other tumors occurring in the posterior fossa only based on the imaging. When the imaging of tumors in the posterior fossa can not be explained by common tumors, the diagnosis should consider the possibility of medulloblastoma, but pathologic examination after surgery is the only effective method of diagnosis.

References


Fig. 1 Patient, female, 4 months old. A~B. The first examination showed that the cortex of the cerebellar hemisphere had diffuse nodular thickening, a high signal on DWI imaging, and significant enhancement bilaterally. Fig. 2. Contrast enhancement MRI 1 year later showed the signal of the cerebellar hemisphere returned to normal, but an enhanced nodule was demonstrated. Fig. 3. Re-examination 6 months later: A shows an irregular mass with a high-density shadow in the cerebellar vermis on CT scan, B~C shows the mass was isointensity on the T2-weighted image with multiple degeneration cysts and significant enhanced. Fig. 4. The pathologic findings showed that the tumor cells were composed of a large number of nodules with an abundance of neutrophils. The cells were consistent with the characteristics of nerve cells and rare mitotic figures. Medulloblastoma with extensive nodularity was diagnosed on the basis of these findings. H&E. Original magnification ×200
Highlights
We retrospectively analyzed a case involving a medulloblastoma with atypical dynamic imaging changes to evaluate the atypical MRI features of medulloblastomas, and reviewed and summarized the characteristics by reviewing the literature. The radiologic characteristics of atypical medulloblastomas varies in adults and children. Understanding the radiologic characteristics of medulloblastomas, such as MRI features, age, and location of atypical medulloblastomas, can help improve the diagnosis of medulloblastomas.
Abbreviations

MRI: Magnetic Resonance Imaging

CT: Computed Tomography