Medulloblastoma Mimicking an Extraaxial Tumor on Radiological Examination

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The extraaxial presentation of medulloblastoma is a rare phenomenon. This article describes the case of a 19-year-old woman who presented with mild headache and nausea and was diagnosed with medulloblastoma. The tumor arose from the right cerebellar cortex, and it was misdiagnosed meningioma on the basis of radiological examination. We review the literature and discuss the such atypical presentation of medulloblastoma.

KEY WORDS: Medulloblastoma · Meningioma · Atypical

Introduction

Medulloblastoma is the most common childhood intracranial tumor, accounting for 25% of all pediatric intracranial tumors and 33% of all posterior fossa neoplasm in children. The common radiological appearance of a medulloblastoma is that of a hyperdense, enhancing, well-demarcated mass arising from the vermis, but variations from this ‘typical’ presentation are not uncommon. There have also been reports of cystic changes, calcifications, and metastases in frontal and suprasellar areas and of a primary tumor arising in the cerebellopontine angle. In adults this tumor may differ significantly from the childhood variety, and mimic a wide variety of posterior cranial fossa neoplasm. Here we report a case of medulloblastoma, which seemed like an extraaxial tumor on radiological diagnosis. The lesion was initially considered to be meningioma on the basis of clinical and neuro-imaging examination and surgery was performed. On histopathological examinations, the lesion was diagnosed as medulloblastoma. This case underline the difficulties encountered when relying on an imaging diagnosis of medulloblastoma.

Case Report

History

This 19-year-old woman presented with mild headache, nausea and dyspepsia. She had been visited a private clinic and was noted cerebellar mass on brain magnetic resonance image, and referred to our hospital. She had worsened during the preceding few weeks.

Examination

Neurological examination yielded normal findings. There were no central vestibular signs on the videonystagmotic study. Magnetic resonance imaging revealed a lesion which was slightly low signal intensity on T1-weighted image on right cerebellum, and enhanced inhomogeneously on T1-weighted MR images after injection of gadolinium. On T2-weighted images, the lesion appeared slightly high signal intensity and mixed signal intensity. Computerized tomography scanning revealed a slight enlargement of the ventricle. Consequently, radiological diagnosis was extraxial tumor such as meningioma most likely.

Operation

In May 2002 the patient underwent surgery performed by the neurosurgical team through the suboccipital approach. After right suboccipital craniotomy a moderately swollen cerebellum was noted. The tumor had a gray colored slightly firm character and was adhered to the right tentorium. The tumor was removed gross totally. The frozen biopsy study was not undergone.

Histopathologic findings

The tumor was composed of densely packed cells with round-to-oval or carrot-shaped highly hyperchromic nuclei surrounded by scanty cytoplasm and these cells were formed Homer-Wright rosette. On the immuno-histochemical study S-100 protein and neuron specific enolase were positive but GFAP, neurofilament, and cytokeratin were negative.
Postoperative course and additional treatment
Postoperative period was uneventful. Complementary investigation (magnetic resonance image of the brain) was conducted to evaluate the residual tumor mass, but there was no definite residual or recurrent mass. After biopsy result was reported, magnetic resonance imaging of the spinal cord and lumbar puncture were conducted to evaluate the metastasis to the spine. But those studies did not reveal any evidence of metastasis to the spinal cord. The patient underwent craniospinal radiotherapy. Magnetic resonance images obtained 3 months postoperatively demonstrated no residual or recurrent mass. After discharge, she admitted our hospital for adjuvant chemotherapy.

Discussion
The radiological features of medulloblastoma are classic: they display an iso or hypointense signal on T1-weighted images, are heterogeneous on T2-weighted images, and exhibit homogeneous enhancement after addition of gadolinium, sometimes demonstrating a central hemorrhagic Zone2,5). Most medulloblastoma are located in the midline of the cerebellum, especially in children10). The typical CT appearance of medulloblastoma is that of a well-defined, hyperdense enhancing soft tissue mass with surrounding edema arising in the cerebellar vermis, associated with obstructive hydrocephalus10). The SIOP II trial reviewed 233 cases and showed ‘typical’ features in only 30% of cases9). The primary lesions were cystic in 59% of the patients, calcified in 22%, cystic and calcified in 15%, ill defined in 7%, not associated with edema in 5% and not accompanied by hydrocephalus in 4%. Eighteen percent of tumors were nonvermal, while 15% were already metastatic at presentation9).

There are currently two general hypotheses regarding the origin of medulloblastoma. In the first, medulloblastomas are proposed to arise from primitive multipotential cells in the external granular cell layer of the medullary velum, which are therefore unique to the cerebellum. The alternative view is that medulloblastoma arise from multipotential cells in the subependymal region and within the fetal pineal region and give rise to all PNETs, regardless of location.7 In the other words, medulloblastoma can be occurred on everywhere germ cell tumor’s normal migration course to lateral side6).

We reports a case that there was a extraaxial localization of tumor on magnetic resonance images with no clinical alteration in brain stem so that can be misdiagnosed as extraaxial tumor such as meningioma. On the basis of radiological examination we thought that it is medulloblastoma. On operation the mass was slightly soft and grey colored, and it didn’t seem like medulloblastoma. As we mentioned above, medulloblastoma presented variously on radiological examination. Besides the typical radiological finding the tumor also presented hemispheric and cerebellopontine angle, so can be misdiagnosed extraaxial tumor such as meningioma. So biopsy result was must be confirmed for prevent misdiagnosis.

Conclusion
Medulloblastoma is a tumor that varies in its presentation and its biological behavior. This being so, it is a moot point as to what should be regarded as atypical for these tumors. Further prospective clinicoradiological studies are required to establish the atypical profile of this not uncommon posterior fossa tumor.

References
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