Pre-operative misdiagnosis of pediatric schwannomas as hemangioblastomas

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To the Editor: Schwannomas, which are benign tumors derived from Schwann cells, are common soft-tissue tumors. However, schwannomas in children are rare. Furthermore, the imaging findings of some cases of pediatric schwannomas can be similar to those of hemangioblastomas, which makes them easily misdiagnosed. We reported a case of a pediatric schwannoma in the foramen magnum which was misdiagnosed as a hemangioblastoma before surgery.

A Chinese girl, 8 years old, underwent an examination because precocious puberty was suspected by the family members. The patient’s head computed tomography accidentally revealed “an occupying lesion in the right foramen magnum region.” A pre-operative head magnetic resonance imaging (MRI) showed a lumpy and slightly prolonged T1/T2 signal in the right foramen magnum with a clear border, with an enhanced scan showing significantly less homogeneous enhancement of 24 mm × 24 mm × 28 mm and diffusion-weighted imaging showing no significant diffusion limitation [Figure 1A–1F]. The physical examination and laboratory tests were negative.

The pre-operative diagnosis of hemangioblastoma in the right foramen magnum region was established. With no obvious surgical contraindications shown by pre-operative examinations, a craniotomy at the posterior cranial fossa for tumor resection was performed. During operation, a gray-red tumor closely associated with the nerve root was observed in the right C1 nerve root, which was resected.

The pathologic results showed that the fusiform tumor cells were arranged in bundles and the nuclei were organized in a fence-like pattern, which indicated that the tumor was a schwannoma [Figure 1G]. Enhanced head MRI at 3 months [Figure 1H] showed post-operative changes in the posterior fossa, with the tumor completely resected.

Schwannoma usually develops slowly and shows no specific symptoms in the early stage. Thus, the condition is likely to be misdiagnosed. Besides, schwannomas in children are rare, and the patient described in this report was 8 years old, which is a factor that led to the misdiagnosis. Also, the clinical imaging characteristics of schwannomas generally include a lesion with low or medium signal intensity on the MRI T1-weighted images, a high heterogeneous T2-weighted signal, and mild heterogeneous enhancements. However, in the case, the enhanced MRI showed that the lesion had an irregular and slightly prolonged T1/T2 signal, with a clear border, and the enhanced scanning of the lesion revealed significantly less homogeneous enhancement. The imaging characteristics of this case mimicked a hemangioblastoma.[2] Furthermore, because the tumor was derived from the C1 nerve root and extended into the cerebellum but did not grow outside of the spinal canal, it lacked the typical imaging feature of schwannoma, the dumbbell-like shape.[3]

As schwannomas can be completely treated by complete excision of the tumor without damaging the adjacent nerve in most patients, a clear pre-operative diagnosis is critical. A previous study discovered that the recurrence rate of the patients in the subtotal resection group was significantly higher than that of the total resection group.[4] Fortunately, although this case was pre-operatively misdiagnosed as a hemangioblastoma, the neurosurgeon intra-operatively determined that the case might be misdiagnosed and performed a total resection without causing damage to the peripheral nerve.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient or her guardian has given their consent for her images and other clinical information to be reported in the journal. The patient or her guardian understand that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Conflicts of interest

None.

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