Child's Nervous System (2019) 35:1041–1044
https://doi.org/10.1007/s00381-018-04045-4

CASE REPORT

Pediatric intracranial lower cranial nerve schwannoma unassociated with neurofibromatosis type 2: case report and review of the literature

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Received: 3 August 2018 / Accepted: 28 December 2018 / Published online: 13 January 2019

Abstract

Introduction Pediatric schwannomas are rare, and most of them are associated with neurofibromatosis type 2 (NF2) and usually located in the vestibular nerve. Herein, we present the first pediatric case of intracranial schwannoma derived from the IX/X nerve complex unrelated to NF2.

Case report The patient was a 9-year-old boy who presented with a 3-month history of headache and nausea. There was no family history of NF2. Imaging studies revealed a cystic lesion with enhanced wall in the left cerebellomedullary fissure. During the operation, the IX/X nerve complex was strongly adhered to the tumor at the jugular foramen. The tumor was totally excised, and the postoperative MRI demonstrated no residual tumor. Histopathological diagnosis was schwannoma. Genetic analysis revealed no mutation associated with NF2 and schwannomatosis.

Conclusion We reported the first case of pediatric lower cranial nerve schwannoma which was not associated with NF2. The schwannoma should be included as differential diagnosis of pediatric posterior fossa tumors.

Keywords Pediatric intracranial schwannoma · Lower cranial nerves · Neurofibromatosis type 2

Case report

History and examination

The patient was a 9-year-old boy who presented with a 3-month history of headache and nausea. There was no family history of NF2.

On examination, he had slightly weak gag reflex. No obvious skin lesions were observed.

Computed tomography (CT) scan revealed a cystic lesion in the left cerebellomedullary fissure (Fig. 1a). Magnetic resonance (MR) imaging showed an enhancement of cyst wall (Fig. 1b). The apparent diffusion coefficient (ADC) was relatively high (Fig. 1c). Low-grade glioma was initially suspected. We proceeded to surgical resection of the tumor.

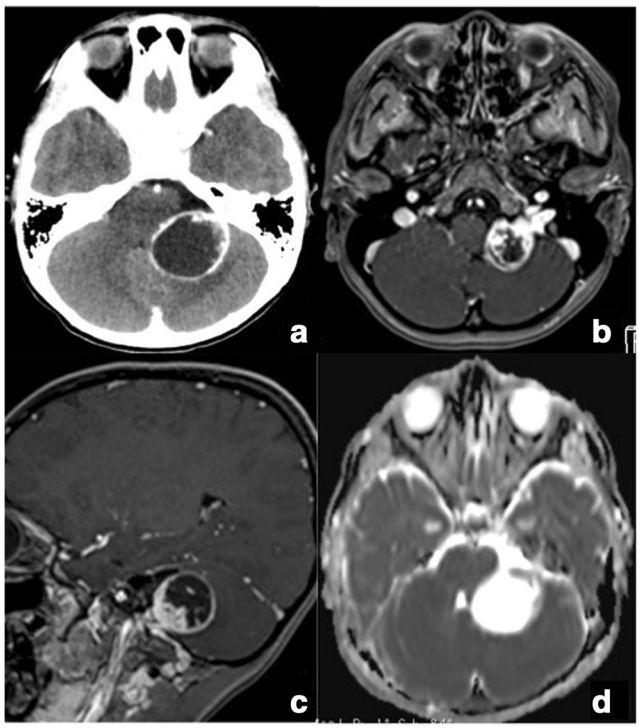

Fig. 1 Preoperative CT **a** showed a cystic mass lesion in the left cerebellar inner part and MRI (**b**, **c**) showed an enhancement effect of cyst wall by gadolinium contrast agent. Since the decrease in tumor ADC (**d**) was not clear, we initially suspected low grade glioma from image findings

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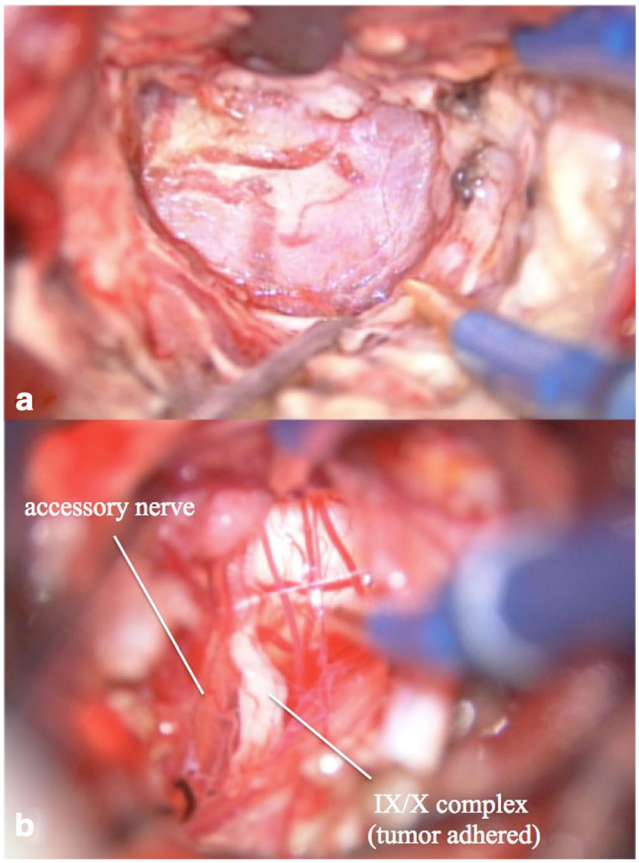

Fig. 2 Intraoperative imaging showed that the tumor was grayish white, hard and vascularity was moderate and tumor was particularly strongly adhered to the IX/X nerve complex

Operation

The patient was placed in the prone position. A left lateral suboccipital craniotomy was performed. After 1.5 cm-uncapping of lateral cerebellar hemisphere, the tumor was exposed (Fig. 2a). The VII/VIII cranial nerve complex was identified on the rostral side, and the lower cranial nerves were observed on the anterior caudal surface. Those nerves were dissected from the tumor. The IX/X nerve complex were strongly adhered to the tumor at the jugular foramen (Fig. 2b). The tumor was totally excised. The postoperative MRI demonstrated no residual tumor (Fig. 3).

Histological examination

The histological examination demonstrated tumor cells with short spindle-shaped nuclei growing in palisade or bundles, mild hyalinization of the interstitium, and the hyalinized vitreous wall (Fig. 4a). In immunostaining, S-100 was positive in the tumor cells, and GFAP and Oligo2 were negative (Fig. 4b). The Ki-47 was 4–5%. Those findings were compatible with schwannoma (WHO grade I). The tumor was considered as schwannoma originated from the nerve sheath of the left lower cranial nerves (Fig. 5).

Postoperative course

Postoperatively, decreased pharyngeal reflex, left vocal cord paralysis, dysphasia and dysarthria were observed. Those symptoms improved gradually and resolved 7 months after the surgery. Whole-exome sequencing revealed no mutation in the NF2 tumor suppressor gene on 22q12, which encodes *Merlin*. Mutations in *LZTR1* and *SMARCB* gene, which were found in schwannomatosis, were not observed, either. He had been stable without recurrence at a year follow-up.

Discussion

The common age of schwannomas ranges from 40 to 50 years old. They consist of 10.4% of brain tumors in all ages, and 0.8% of brain tumors in childhood [1]. The proportion of pediatric cases in schwannomas is approximately 2% [2]. Most of them develop in vestibular nerve. Schwannomas occurring outside the vestibular nerve comprise 2.9–4% [3]. Most of schwannomas in children are associated with NF 2,

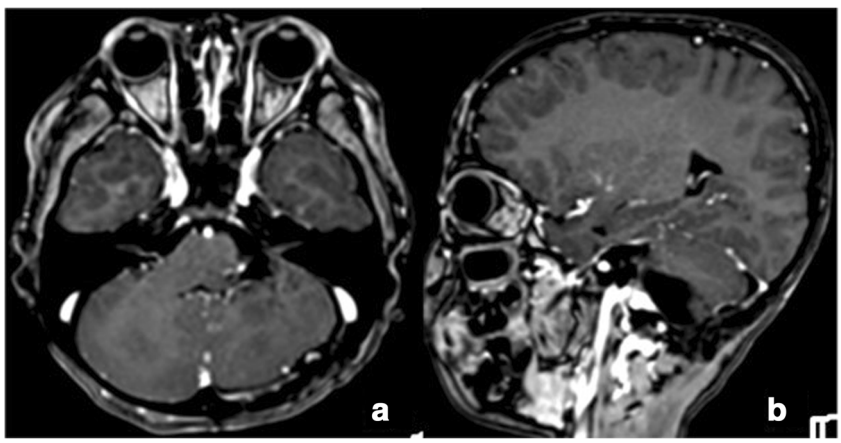

Fig. 3 The tumor was completely excised and the postoperative MRI

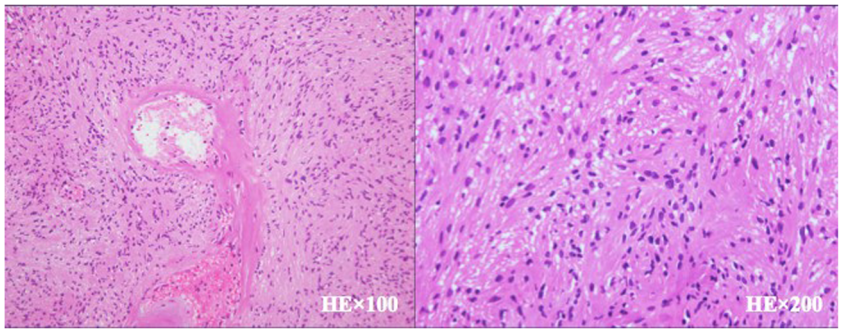

Fig. 4 Histological findings of cell with short spindle-shaped nuclei growing in palisade or bundle were confirmed

and usually located in the vestibular nerve [4]. The locations of nonvestibular cranial nerve schwannomas in NF2 included the lower cranial nerves complex (36.4%), trigeminal nerve (27.3%), hypoglossal nerve (27.3%), and oculomotor nerve (9.1%) [5].

The present case did not meet the diagnostic criteria of NF2. Genetic analysis revealed no mutation associated with NF2 and schwannomatosis.

There have been 24 pediatric cases of schwannoma which were not related to NF2. Of those, 13 cases originated in the oculomotor nerve, 10 in the vestibular nerve, and 1 in hypoglossal nerve. The age of patients with oculomotor nerve schwannomas ranged from 2 to 15 years old [6]. The patient with a hypoglossal nerve schwannoma was a 9-year-old girl [4]. In a report of 10 cases of unilateral vestibular schwannomas of children unrelated to neurofibromatosis, the mean age was 13.9 years and ranged from 11 to 15 years. In this series, genetic search has not been done, and not relating to neurofibromatosis was decided from physical findings and family history [7].

To date, there have been no reports of intracranial schwannoma derived from the IX/X nerve complex which is not associated with NF2. The present case is the first pediatric report of intracranial schwannoma of derived from the IX/X nerve complex unrelated to NF2.

Jugular foramen schwannomas are classified into the following four types by location, Type A: posterior cranial fossa; type B: jugular foramen; type C: extracranial portion; or type D: a dumbbell tumor extending in and out of the skull [8]. For type A tumor, resection is usually performed with a retrosigmoid approach. The infralabyrinthine approach for type B and C, and the combined approach for type D, are generally used for resection. It is reported that injury of lower cranial nerve occurs in 15–50% of surgical cases [9]. Postoperative neurological deficit of the glossopharyngeal and vagus nerve is reported to be 30.1% for transient and

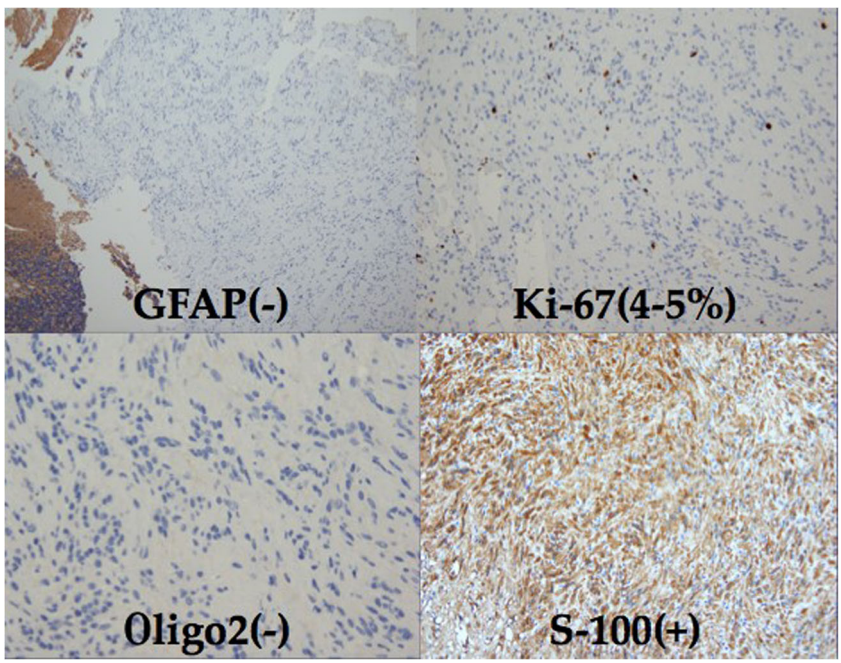

Fig. 5 In immunostaining, S-100 was positive in the cells to be increased, and GFAP and Oligo2 were negative

26.4% for persistent. In the accessory nerve, transient deficit is 17%, permanent one is 11.3% [10]. The present case was the type A tumor, and we have been able to achieve total removal of the tumor using the retrosigmoid approach as reported, although adhesion was relatively strong between tumor and the lower cranial nerves. The patient had dysphagia due to the glossopharyngeal nerve and vagal nerve paralysis after surgery, which eventually resolved in 7 months. There have been no recurrence for 1 year since the surgery. Considering the relatively long life expectancy and better chance of recovery of neurological deficit in pediatric cases, total removal of the tumor may better be attempted as the first strategy for pediatric lower cranial nerve schwannomas.

Conclusion

We reported the first case of pediatric lower cranial nerve schwannoma which was not associated with NF2. The schwannoma should be included as differential diagnosis of pediatric posterior fossa tumors.

Compliance with ethical standards

Conflict of interest On behalf of all authors, the corresponding author states that there is no conflict of interest.

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