Recurrent isolated oculomotor nerve palsy caused by schwannoma in a pediatric patient

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Isolated schwannomas of motor nerves to extraocular muscles are uncommon. In addition, most previous studies on oculomotor nerve schwannoma discuss adult patients, and pediatric cases are rare. We report a 10-year-old girl who developed recurrent oculomotor nerve palsy caused by schwannoma without any vascular malformation. Although the incidence is rare in pediatric patient, the recurrent isolated oculomotor nerve palsy due to schwannoma can develop, and it should be considered in the differential diagnosis of ocular motility disorders in pediatric patients. Careful imaging evaluation is needed to identify schwannoma due to its small size, deep location in the brain, and rarity.

Key words: Oculomotor nerve schwannoma, pediatric patient, recurrent isolated oculomotor nerve palsy

Isolated schwannoma arising from the oculomotor nerve can produce an acute or progressive oculomotor nerve palsy and it is rare in children without an associated neurofibromatosis. We report a recurrent isolated oculomotor nerve palsy caused by schwannoma in a pediatric patient. A 10-year-old girl visited our clinic due to recurrent binocular diplopia and right ptosis. On examination of eyes, she was diagnosed with a right oculomotor nerve palsy. Magnetic resonance imaging (MRI) with magnetic resonance angiography (MRA) of the brain revealed a right nodular schwannoma located within the cisternal segment of the oculomotor nerve without any vascular malformation. Isolated recurrent oculomotor nerve palsy caused by oculomotor nerve schwannoma can develop in pediatric patients. High-resolution MRI and thin-section cranial MRI can be helpful to avoid missing the lesion.

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Case Report

A 10-year-old girl presented to the ophthalmic department with right eye drooping and double vision for 1 day. She had headache for 1 week. She had a history of suffering headaches about four to five times a year since age 2 years; her headaches caused a dull pain sometimes accompanied by nausea and vomiting. Because of the recurring headaches, she had undergone MRI of the brain in the pediatric department 1 year ago, which had revealed no evidence of abnormality. Her mother reported that this was not the first time she was experiencing ocular movement limitation during headache. A similar ocular motility disorder had developed when she was 2 years old and lived in the United States at that time. She had been diagnosed with a typical cranial neuropathy in the right eye, which had improved after administration of a steroid. The visual acuity was 20/20 in both eyes. The right pupil was poorly reactive to direct and consensual stimulus. The ocular motility examination showed a limitation of ocular movement except abduction in the right eye [Fig. 1]. There was no ocular injection, chemosis, or pain on eye movement. The fundus showed normal appearance of the optic disc in both eyes. All serologic tests, including tests for thyroid function and anti-acetylcholine receptor antibodies, were within normal range. Thin-section cranial MRI with MRA of the brain was performed and it revealed a 6-mm nodular enhancement located within the cisternal segment of the right oculomotor nerve, suggesting oculomotor nerve schwannoma without

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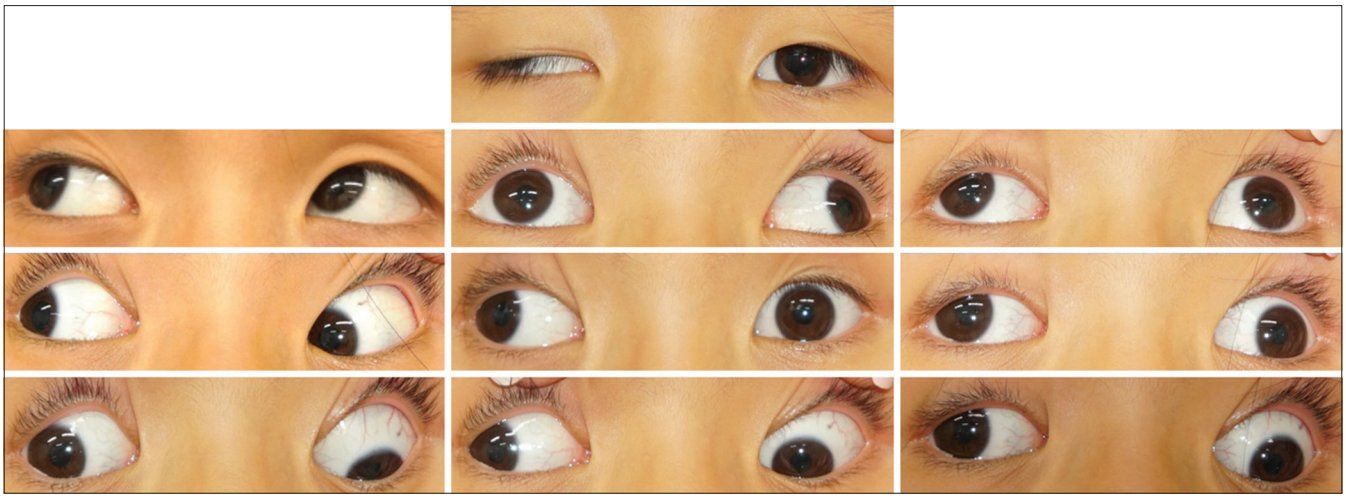

Figure 1: Images of the subject in nine diagnostic positions of gaze at first visit, demonstrating 50 prism diopters exotropia in the right eye in the primary position with limitation of ocular movement except abduction

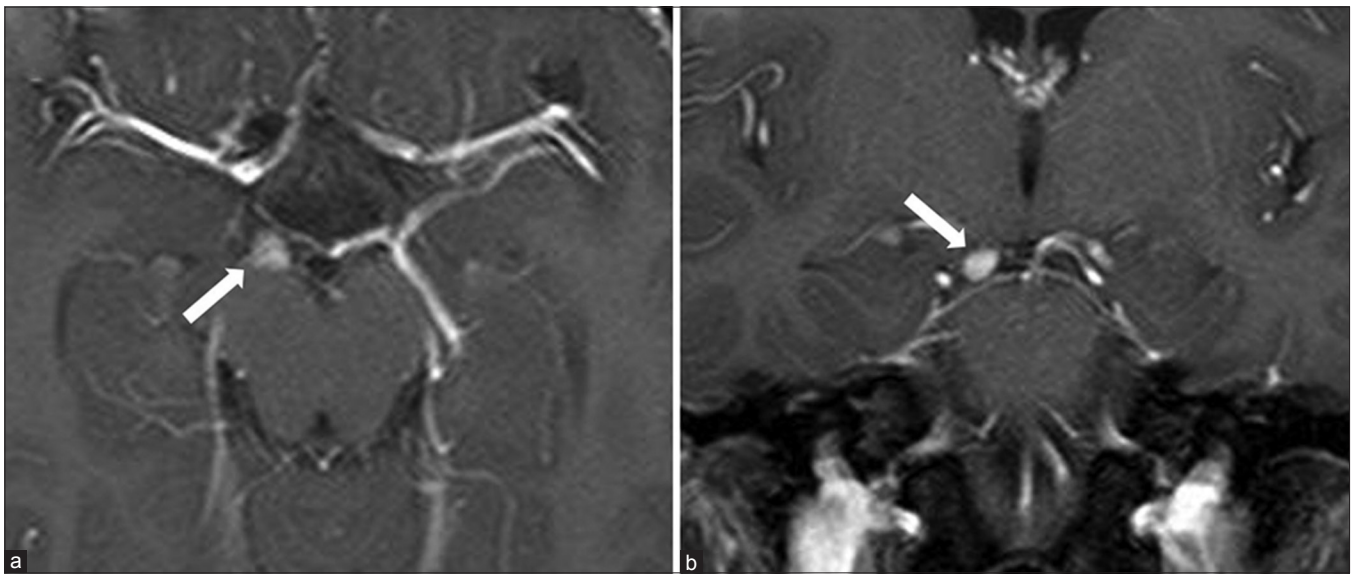

Figure 2: Contrast T1 MRI showing a 4.6-mm enhancing nodule (white arrow). The schwannoma is considered to be located within the cisternal segment of the oculomotor nerve. (a) Axial view. (b) Coronary view

any vascular malformation [Fig. 2a and b]. Although the MRI of the brain performed 1 year earlier had failed to reveal any abnormal lesion at the time, we reviewed the images and found a 3.7-mm thickening lesion of the cisternal segment of the affected oculomotor nerve on the constructive interference in steady-state axial T2 MRI [Fig. 3]. Therefore, we concluded that the schwannoma, which had been 3.7 mm in size 1 year earlier, had gradually increased in size and led to oculomotor nerve palsy. The neurosurgeon recommended gamma knife radiosurgery to remove the tumor; however, the patient's mother refused it due to the risk of permanent sequelae on the oculomotor nerve. She was empirically treated with oral prednisone and 1 week after treatment, her ptosis and headache improved; however, she was lost to follow-up. The patient presented to the ophthalmic department again after a year for checkups. On her follow-up brain MRI, the size of schwannoma had not changed and there was no newly developed lesion. Because her recurrent headache improved, her mother still did not want her to undergo brain surgery. However, exotropia with limitation of ocular movement remained, and therefore, her mother is considering strabismus surgery for her cosmetic reasons.

Discussion

Schwannoma, originating from Schwann cells, is the most common benign peripheral nerve sheath tumor; intracranial schwannoma is also known as neuroma and accounts for 8% of all intracranial tumors.[1] While intracranial schwannomas can affect almost any cranial nerve, the most commonly affected nerve is the vestibulocochlear nerve.[1] Many affected patients complain of tinnitus or hearing loss due to mass effect and dysfunction of the involved nerve. Isolated schwannomas of motor nerves to extraocular muscles are uncommon.[2] A previous study identified only 8 schwannoma cases among 647 patients imaged for strabismus to screen for presumed

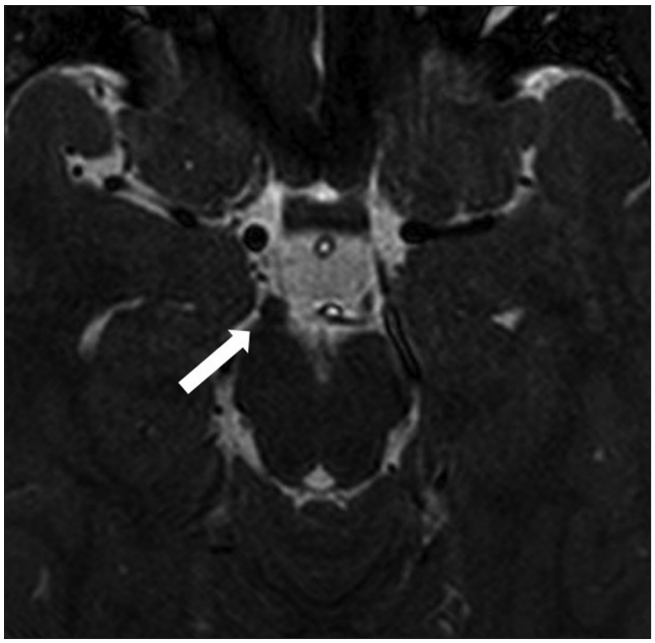

Figure 3: Constructive interference in steady-state (CISS) axial T2 MRI taken 1 year earlier and determined to show no abnormal lesion at the time; however, a 3.7-mm thickening lesion (white arrow) was found when reviewing the images

cranial nerve schwannomas.[3] In addition, most previous studies on oculomotor nerve schwannoma discuss adult patients,[4] and pediatric cases are rare.[5,6] The size of most intracranial schwannomas is less than 1 cm[3,7] and clinical presentations are unspecific and variable according to the location of the tumor; these characteristics can lead to misreading MRI images. Yulek and Demer reported that in six of their eight cases, cranial schwannoma had been previously missed on routine MRI.[3] Their results are consistent with the case of our patient whose lesion was missed in previous neuroimaging. The diagnosis of a schwannoma is often made by imaging evaluation and clinical symptoms that correlated with distribution of the involved nerve. In our case, the mass presents on MRI as a solid nodular mass and shows hyperintense signal on T2-weighted images. In addition, the location of the lesion is within the nerve pathway and the size of the mass has changed little when comparing series of MRIs. These characteristics of the mass on MRI are compatible with an oculomotor nerve schwannoma. Therefore, careful imaging evaluation is essential to avoid missing lesions; high-resolution MRI or thin-section cranial MRI can be helpful with this.[8]

Total resection of the tumor is the standard treatment modality[4] and radiotherapy is an alternative to surgery.[9] As a conservative treatment, steroid administration can be tried based on a "wait-and-see" policy.[4] The prognosis of pediatric oculomotor nerve schwannoma is unknown. Most adult patients can remain stable and not develop additional symptoms. Malignant change or recurrence is unusual.[1] However, in our case, the ocular motility disorder had disappeared after steroid treatment when she was 2 years old, but recurred 8 years later. Therefore, clinicians should be aware of the possibility of the presence of schwannoma in patients with recurrent ocular motility disorders.

Conclusion

In summary, the recurrent isolated oculomotor nerve palsy due to schwannoma can develop in pediatric patient. Careful imaging evaluation is needed to identify schwannoma due to its small size, deep location in the brain, and rarity.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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