Case Report

Schwannoma of the oculomotor nerve

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A 63-year-old woman presented with an extremely rare oculomotor schwannoma not associated with neurofibromatosis, manifesting as a transient diplopia and ptosis. Magnetic resonance images showed a well-enhanced mass extending from the cavernous sinus to the intraorbital region. Surgical exposure confirmed the tumor originating from the oculomotor nerve in the cavernous sinus. The intraorbital cystic part of the tumor was partially resected to preserve the oculomotor function. This is the first case of oculomotor schwannoma in the cavernous sinus with intraorbital component.

Key words: oculomotor nerve, schwannoma, surgery

Introduction

Schwannomas arising from the oculomotor nerve without neurofibromatosis are extremely rare. To our knowledge, only 38 cases have been reported in the literature. All these tumors were located in the cistern or in the cavernous sinus. We report the first case of oculomotor schwannoma extending from the cavernous sinus to the intraorbital region, mimicking trigeminal schwannoma. Clinical manifestation as well as the surgical indication is reviewed.

Case Report

A 63-year-old woman had complained of diplopia, ptosis, chemosis and dull pain on her right eye over the one month prior to admission. The symptom disappeared three days later. On neurological findings, pupils were isocoric and light reflex was prompt bilaterally. There was no deficiency in extraocular movement and any other abnormal signs were not present. There was no evidence of neurocutaneous markers. Magnetic resonance study showed a tumor in the lateral compartment of the cavernous sinus (lateral to C4 segment of the internal carotid artery) to the orbit through the superior orbital fissure. Tumor was partially cystic in the orbit [Figure 1].

Under the possible diagnosis of trigeminal schwannoma, combined orbitozygomatic infratemporal epidural and subdural approach was performed. The tumor in the cavernous sinus was exposed via a combined epidural and subdural approach after optic canal unroofing and anterior clinoidectomy. The oculomotor nerve was dilated in the funnel-shaped at the porus oculomotorius and continued to the tumor in the cavernous sinus [Figure 2]. Intraorbital tumor was accessed between the lateral rectus muscle and superior rectus muscle and found to be located beneath the optic nerve. Thin nerve fibers, intervened between the tumor and optic nerve sheath, were tightly adherent to the tumor. Those nerve fibers were thought to be a part of short ciliary ganglion. We removed the cystic part of the tumor in the orbit, although some branch of short ciliary nerve was sacrificed. The tumor in the cavernous sinus was totally left behind. Histopathological examination of the tumor revealed schwannoma WHO Grade I.

Figure 1: Preoperative axial T1-weighted precontrast (A) and postcontrast (B) images showing a well-enhanced mass extending from the cavernous sinus to the intraorbital region. T2-weighted image (C) revealing the cyst formation inside the retrobulbar tumor part. Coronal postcontrast image (D) demonstrating the total occupation of the lateral compartment of the cavernous sinus by the tumor. Note that these radiological features are similar to trigeminal schwannoma

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After operation, ocular movement except for levator function was slightly decreased. During six years’ follow-up period, the tumor in the cavernous sinus has slightly grown without neurological deterioration [Figure 3]. We will continue the follow-up of the patient with periodical examination of magnetic resonance image.

Discussion

Although it is reported that intraorbital schwannoma accounts for 1-6% of all intraorbital tumors,[12] the accurate diagnosis of the tumor origin may be difficult because of the complex orbital anatomy. Operative finding is the key to confirm the tumor origin. Thirty-eight cases of solitary oculomotor schwannoma reported in the literature included 15 male and 23 female patients, whose age ranged from 8 to 74 years. These age and sex distributions were similar to vestibular schwannoma. It is noteworthy that preoperative oculomotor dysfunction was manifested in 29 cases out of 38. The tumor was located in the orbit in four cases (solitary orbital type), in the subarachnoid space in 17 cases (cisternal type), in the cavernous sinus in 12 cases (cavernous type), extending from the cavernous sinus to the cistern in five cases (cisterno-cavernous type).[11-10] Conceivably, two clues leading to the diagnosis are an oculomotor paresis and tumor location along the course of the oculomotor nerve, especially in its cisternal portion. In the present case, differential diagnosis from trigeminal schwannoma was very difficult as the tumor extended from the cavernous sinus to the intraorbital area like trigeminal schwannoma and the oculomotor paresis was transient. Considering the onset of third nerve deficit being subacute and having disappeared in three days, the possibility of intratumoral hemorrhage may be considered.

Surgical intervention was conducted in 34 cases and most of them underwent total or subtotal resection of the tumor. However, oculomotor function was not improved or recovered postoperatively in the majority of those cases. In the present case, only the cystic intraorbital part of the tumor was removed as the relation between the oculomotor nerve and the tumor in the cavernous sinus would be more complex. Undoubtedly, worsening of oculomotor function might happen even after partial resection from the cavernous portion.

In the management of the intraorbital tumor, a logical sequential workup should follow the usual clinical examination. Under the condition in which clinical parameters have not firmly established the surgical indications, the precise ophthalmological workup of visual acuity or intraorbital pressure may be helpful. We recommend that removal of oculomotor schwannoma should be indicated only for large tumors with intractable symptoms and, if surgery is conducted in the case of not large or less symptomatic tumors involving the cavernous sinus, the surgical strategy should be changed based on the operative findings to prevent further neurological damage. It would be possible to observe the natural course of the tumor in the cavernous sinus using MR image until the tumor becomes large or stereotactic radiosurgery may be an alternative to radical resection.

References