Orbital Schwannoma Extending to the Lateral Wall of the Cavernous Sinus Through the Superior Orbital Fissure

—Case Report—

Yasuo HIRONAKA, Hiroyuki NAKASE, Yasushi MOTOYAMA, Hideaki MISHIMA, Young-Su PARK, Hidehiro HIRABAYASHI, and Toshisuke SAKAKI

Department of Neurosurgery, Nara Medical University, Kashihara, Nara

Abstract

A 58-year-old man presented with a rare orbitocavernous sinus schwannoma that originated from the orbital ophthalmic nerve, and manifested as slowly progressive hypesthesia of the right side of the forehead, proptosis, and ocular pain with rapidly worsening visual acuity. Magnetic resonance imaging revealed a huge orbital tumor extending to the lateral wall of the cavernous sinus through the superior orbital fissure. Microsurgical total resection of the tumor was achieved using an epidural orbitofrontal approach with orbito-fronto-zygomatic craniotomy. The histological diagnosis was schwannoma with Antoni type A formation. The postoperative course was uneventful except for the hypesthesia on the right side of the forehead and transient oculomotor paralysis. Surgery was effective to relieve the symptoms and improve the activities of daily living.

Key words: cavernous sinus, fronto-orbito-zygomatic epidural approach, ophthalmic nerve, orbital schwannoma, superior orbital fissure

Introduction

Orbital schwannoma accounts for 1–4% of all orbital tumors, although orbital schwannoma tends to be reported without definite identification of the origin. The majority of orbital schwannomas are benign, although some show malignant changes. Most originate from branches of the oculomotor, trochlear, trigeminal, and abducens nerves, and from sympathetic and parasympathetic fibers. However, orbital schwannoma originating from the first branch of the trigeminal (ophthalmic) nerve with extension to the lateral wall of the cavernous sinus from the orbit through the superior orbital fissure (SOF) is uncommon. Tumors of the cavernous sinus except for schwannomas tend to be treated by surgical resection rather than stereotactic radiosurgery due to concerns regarding removal, hemostasis, and the appearance of new postoperative deficits. Gamma-knife surgery for trigeminal schwannoma provides a high rate of tumor control and functional improvement, and surgical treatment leads to good outcomes. Therefore, surgical intervention is appropriate for schwannoma extending into the cavernous sinus.

Here, we report the successful surgical treatment of a case of orbital schwannoma that originated from the ophthalmic nerve and extended to the lateral wall of the cavernous sinus.

Case Report

A 58-year-old man presented with a 2-year history of slowly progressive numbness of the right forehead, proptosis, and blurred vision. He had experienced rapid deterioration of visual acuity in the right eye, and had suffered retrobulbar pain during the previous few months. He had no clinical or family history of neurofibromatosis. Neurological examination on admission revealed severe exophthalmos with corneal ulceration, blindness in the right eye, hypesthesia in the V1 area, and disappearance of corneal reflex. His ocular motility was normal, with no abnormal nystagmus. Magnetic resonance (MR) imaging demonstrated a huge smooth-bordered mass in the orbit that extended to the lateral wall of the cavernous sinus. The mass appeared as a homogeneous hypointense lesion with enhancement on the T1-weighted image, and as an isointense lesion with a hyperintense rim on the T2-weighted image (Fig. 1). Radiography and three-dimensional computed tomography (CT) revealed enlargement of the SOF on the tumoral side (Fig. 2), and extension of the tumor through the gap to the cavernous sinus. Angiography showed some tumor staining in the ophthalmic artery, but the cavernous sinus was not occluded. Direct surgery was performed to treat the severe symptoms, with the expectation of total tumor removal through the enlarged SOF. The patient was placed in the supine position with his head rotated 30° to the opposite side. Orbito-zygomatic craniotomy combining frontotemporal craniotomy with en bloc removal of the orbital su-
Fig. 1  Axial (A), coronal (B), and sagittal (C) T₁-weighted magnetic resonance images with gadolinium showing an orbital tumor extending to the lateral wall of the cavernous sinus through the superior orbital fissure.

Fig. 2 Three-dimensional computed tomography scan revealing enlargement of the right superior orbital fissure due to tumor extension (arrows).

Fig. 3 Photomicrograph of the specimen showing schwannoma with Antoni A formation. Hematoxylin-eosin stain, ×100.

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periorbital wall and the zygomatic arch was performed, with full rongeuring of the sphenoid wing and opening of the SOF. Sufficient dissection of the meningo-orbital band was performed to permit retraction of the temporal lobe posteriorly from the epidural space, and to decrease the cerebrospinal fluid by spinal drainage. The enlarged SOF was opened following the orbital supero-lateral wall, so that the huge tumor was exposed directly by opening of the periorbita, removal of orbital fat, and approach through the interval between the levator and superior rectus muscle medially and the lateral rectus muscle laterally. The tumor was located in the right orbital intraconal space and extended to the lateral wall of the cavernous sinus through the SOF. Initially, the tumor was exfoliated from the normal orbital tissue. The optic nerve was adherent to the tumor with severe inferior compression, and was detached carefully. The other peripheral nerves and vessels were impossible to distinguish from peritumoral tissue, as the tumor had severely compressed the intraorbital contents. The orbital tumor was removed completely by debulking of the mass. The extension of the tumor into the lateral wall of the cavernous sinus was then extracted as a single mass by careful dissection and was not adherent to the orbital tumor. Gross total removal of the tumor was achieved without damage to the surrounding tissue or sinus bleeding. The optic nerve and oculomotor nerve running through the intradural space were transparent from the cavernous tumoral cavity of Dolenc’s triangle, and reacted to pulsation of the cavernous internal carotid artery. Finally, the bone defect of the SOF and orbital lateral wall were reconstructed using the bone flap to prevent enophthalmos.

Histological examination showed spindle-shaped cells without mitosis and nuclear fission, along with elongated cells with occasional nuclear pulsation (Antoni type A). The final diagnosis was schwannoma World Health Organization grade 1 (Fig. 3).

The patient recovered promptly from the operation. The retro-orbital pain improved rapidly, but the blindness and hypesthesia in the V1 area were not resolved, and de novo oculomotor paralysis occurred without decreasing the quality of life. Postoperative MR imaging showed that the orbito-cavernous sinus tumor had been removed without creating a new intracranial abnormality (Fig. 4). The patient had no neurological deficits except for blindness and hypesthesia of the right side of the forehead, numbness, and mild oculomotor paralysis at discharge. After 6 months, the oculomotor paralysis was minimal, and almost complete recovery had occurred compared with the immediate postoperative state.
Fig. 4 Postoperative axial (A), coronal (B), and sagittal (C) T₁-weighted magnetic resonance images with gadolinium showing elimination of the tumor.

Discussion

The neural components of the orbit might lead to neurogenic tumors originating from branches of the oculomotor, trochlear, trigeminal, and abducens nerves, and from sympathetic and parasympathetic fibers. Most peripheral nerve tumors originate from the first branch of the trigeminal nerve. Patients with this type of tumor present with slowly progressive proptosis or displacement of the globe, and seldom develop diplopia, which is a defect of visual function. Our patient presented with numbness of the right forehead, proptosis, orbital pain, and blurred vision in the early stage, and the visual disorder worsened rapidly, resulting in blindness due to compression by the huge tumor.

Orbital schwannomas, and particularly trigeminal schwannomas, are rarely extensions of intracranial schwannomas. The present tumor became gigantic due to absence of treatment over a long period, which allowed extension of the tumor to the cavernous sinus through the SOF. MR imaging indicated that the orbital tumor was larger than the cavernous sinus. This is extremely rare, as the increased use of MR imaging and CT usually leads to the identification of an asymptomatic lesion at an earlier stage. Identification of the tumoral nerve in orbital tumors is difficult, but the origin could be established based on the clinical symptoms and postoperative indications. The tumor was adherent to the optic nerve and other orbital components, so we were unable to find the attachment of the original nerve. The tumor in the cavernous sinus part was not adherent to the wall, unlike the orbital tumor. The growing part of the tumor might have been located in the cavernous sinus wall, because a Schwann cell is present in the perivascular nerve sheath of the internal carotid artery, and in each cranial nerve sheath.

Our patient presented with forehead numbness and hypesthesia which persisted throughout the course. Trigeminal nerve dysfunctions were observed in more than 60% of patients in one series, and facial hypesthesia is the most common abnormal finding resulting from trigeminal neurinomas. The oculomotor paralysis was a new postoperative symptom, which was transient, and might have been caused by the cavernous or orbital surgical invasiveness. If the origin of the tumor was the oculomotor nerve, then diplopia would be observed over a long period. We identified the tumor origin as the orbital first branch of the trigeminal nerve based on this evidence.

The optimum treatment for orbital schwannoma is complete surgical excision which can be achieved if limited to the orbital region. However, total resection can be difficult if the tumor extends to regions such as the cavernous sinus. In such cases, selection of an appropriate surgical approach is important. The choice of the surgical route is determined by the location, size, and extension of the tumor. Various methods can be used, including a cranio-orbital approach with or without skull-base exposure, and transconjunctival, supraorbital, and lateral orbital approaches. If the tumor is located only in the orbit, the approach that allows the easiest access to the tumor is chosen. For tumors extending to the cavernous sinus and middle fossa, a cranio-orbital-zygomatic approach is an excellent choice, which allows wide access and should be used for large intraorbital tumors. Total resection of all intracavernous trigeminal neurinomas in one series was achieved without major surgical complications using a fronto-temporal epidural approach. The large trigeminal neurinoma extending into multiple fossa could be removed during a one-stage operation using a skull-base approach, and the risk of morbidity or mortality related to this treatment was not high.

The cavernous sinus consists of lateral, medial, posterior, superior, and inferior walls. The lateral wall consists of two layers: the outer dural layer (dura propria) and the inner membranous layer. The cranial nerves, including the oculomotor, trochlear, trigeminal, and abducens nerves, course between the two layers. The inner layer separates the lateral wall and its contents from the venous channels of the cavernous sinus. Therefore, a tumor in the cavernous lateral wall must be separated from the cranial nerves without causing direct damage or hemorrhaging of the inner membranous layer. Experience with an intracavernous oculomotor schwannoma extending to the orbit suggested that removal of this cavernous...
tumor should be indicated only for large and symptomatic tumors.\textsuperscript{12} Similarly for orbito-cavernous schwannomas, surgical removal should be a carefully considered decision of surgical strategy and tumor localization.

Schwannomas in the cavernous sinus might be treated effectively by radiation therapy and radiosurgery. Stereotactic radiosurgery has achieved high tumor control rates (>80%).\textsuperscript{4,10} Predictors of a better radiosurgery response include smaller tumor volume (<8 cm\textsuperscript{3}), and root or ganglion type for trigeminal schwannomas.\textsuperscript{11} Schwannomas with a volume of 15 cm\textsuperscript{3} or greater should initially be removed surgically, due to the possibility of malignant transformation, and because little is known about the long-term outcomes of radiosurgery.\textsuperscript{19} We also believe that radiosurgery and radiotherapy might be unsuitable for huge schwannomas, because of the cystic component, and is suitable for tiny schwannomas and other cavernous tumors. Nonetheless, resection is a common treatment for patients harboring schwannomas due to its safety and the low risk in terms of morbidity or mortality related to the surgery.

Direct surgery is effective in most cases, provided that postoperative morbidity and tumor localization are considered carefully when selecting the strategy. We were able to perform total tumor resection using an epidural fronto-orbital-zygomatic approach. The wide surgical field improves the outcome by limiting the damage to the surrounding structures and preventing too much traction of the fronto-temporal lobe over the dura. The absence of severe postoperative neurological deficits in the current case might have been due to careful preoperative planning, including the selection of the surgical approach and the determination of the extent of resection. Using this strategy, we were able to obtain good results for a cavernous sinus tumor that originated from the orbital first branch of the trigeminal nerve and extended into the lateral wall of the cavernous sinus through the SOF.

References


Address reprint requests to: Yasuo Hironaka, M.D., Department of Neurosurgery, Nara Medical University, 840 Shizyo-tyou, Kashihara, Nara 634–8522, Japan.

E-mail: hironaka@naramed-u.ac.jp