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Case Report

Orbital Abducens Nerve Schwannoma: A Case Report and Review of the Literature

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Intraorbital schwannoma is a rare tumor which accounts for about 1–2% of all neoplasms of the orbit. Orbital schwannomas most commonly arise from the sensory branches of the trigeminal nerve. On the other hand, intraorbital abducens nerve schwannomas are extremely rare, with a search of the English literature identifying only four cases of intraorbital abducens nerve schwannoma. This is the 5th reported case of an orbital schwannoma arising from the terminal branch of the abducens nerve to the lateral rectus muscle. We report a case of an intraorbital abducens nerve schwannoma in a 51-year-old man with no signs of neurofibromatosis. The tumor was totally excised with functional preservation of the nerve by a zygomatic approach with lateral orbitotomy. With knowledge of these anatomic features, total removal of the tumor with preservation of the abducens nerve function might be possible.

Keywords: abducens nerve, shwannoma, intraorbital, lateral rectus muscle

Introduction

Schwannomas are tumors arising from Schwann cells of the neural sheaths of motor and sensory nerves. They arise focally from the sheath of the fascicle and present as well-defined, eccentrically placed masses. Three clinical forms may present: localized schwannoma; in association with neurofibroma (as part of von Recklinghausen syndrome); or as schwannomatosis.[1)] Schwannoma is seen predominantly in cranial nerve VIII.

Intraorbital schwannoma accounts for about 1–2% of all neoplasms of the orbit.[2)] Orbital schwannomas most commonly arise from the sensory branches of the trigeminal nerve.[3)] Intraorbital abducens nerve schwannomas are extremely rare, with a search of the English literature identifying only four cases of intraorbital abducens nerve schwannoma.[1,3–5)] We report a case of orbital schwannoma arising from the terminal branch of the abducens nerve.

Case Presentation

History and examination

A 51-year-old man with a history of hypertension, hyperlipidemia, gout, and retinal detachment was found to have a left intraorbital tumor when he was hospitalized for a medical checkup of the brain. He showed no visual loss or diplopia. Physical examination revealed no left abducens nerve palsy. Ophthalmologic examination revealed normal visual acuity. No stigmata of neurofibromatosis type 1 or 2 were identified, and no other neurological abnormalities were found.

Magnetic resonance imaging (MRI) (Fig. 1) revealed a 1.0 × 1.2 × 1.2-cm intraconal mass behind the lateral rectus muscle (LRM). The lesion was homogeneously hypointense on T_1-weighted imaging (T_1WI) and hyperintense on T_2-weighted imaging. On gadolinium-enhanced T_1WI, the lesion showed central enhancement. Preoperative diagnosis of a benign intraorbital neoplasm was made by these MRI findings.

Six months later, the tumor had slightly increased in size and he wanted surgical treatment, therefore, radical treatment was performed.

Operation

The patient underwent total removal with a zygomatic approach and lateral orbitotomy. The tumor was found just behind to the LRM displacing the muscle (Fig. 2A), and we were able to identify the origin of the tumor from the terminal branch of cranial nerve VI (Fig. 2B, 2D). The tumor was totally removed with transaction from the origin of the abducens nerve (Fig. 2C).

Pathological findings

Pathological examination revealed typical findings of schwannoma. Antoni A and Antoni B were identified (Fig. 3). The tumor cells showed diffuse nuclear positivity for S100 protein.

Postoperative course

In the immediate postoperative period, the patient had no complaints of diplopia and no paresis of cranial nerve VI was detected. Postoperative MRI suggested complete tumor removal (Fig. 4).

Discussion

Schwannomas are well-defined, encapsulated, slowly progressive benign tumors that develop as eccentric growths from the sheaths of peripheral nerves. They have a predilection for the head and neck region. In the orbit, they account for 1–2% of all tumors.[2)] Schwannomas of the abducens nerve are extremely rare.[6)] The tumor may be located within the cavernous sinus or, more often, at the prepontine region.[6)]

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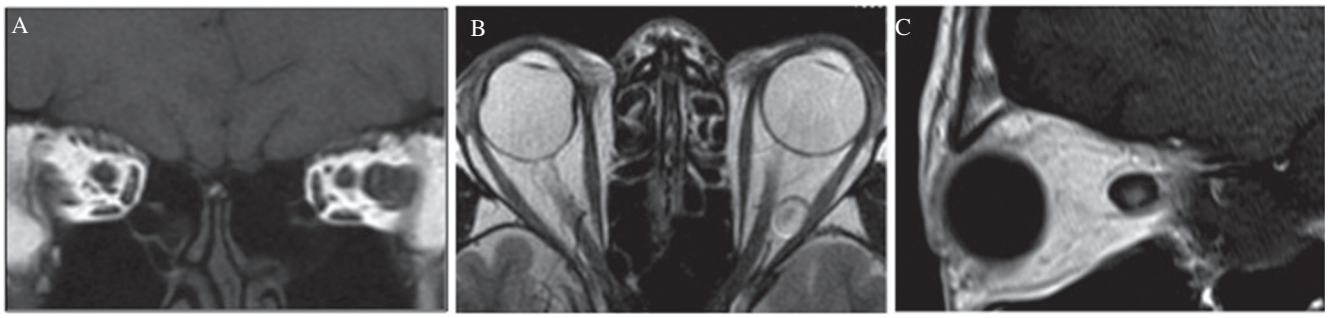

Fig. 1 (A) Radiological findings. Preoperative coronal T_1-weighted magnetic resonance imaging (MRI) showing a 1.0 × 1.2 × 1.2-cm intraconal mass medial to the lateral rectus muscle. The lesion is homogeneously hypointense on T_1-weighted imaging (T_1WI). (B) Preoperative axial T_2-weighted MRI shows smooth contours and the oval shape of the hyperintense lesion. (C) Preoperative sagittal gadolinium-enhanced T_1WI shows central enhancement in the lesion.

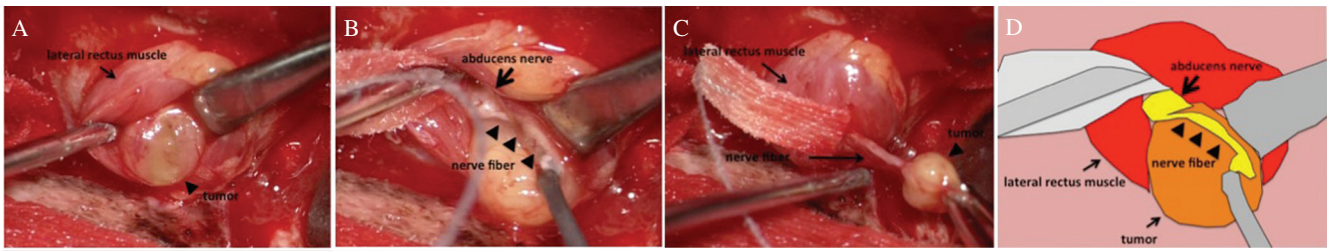

Fig. 2 (A) Intraoperative photograph. The tumor was found at the medial surface of the lateral rectus muscle (LRM) by displacing the LRM. (B) The tumor originated from the terminal branch of cranial nerve VI. (C) The tumor was totally removed with transaction from the origin of the abducens nerve. (D) The illustrative drawing of Fig 2B.

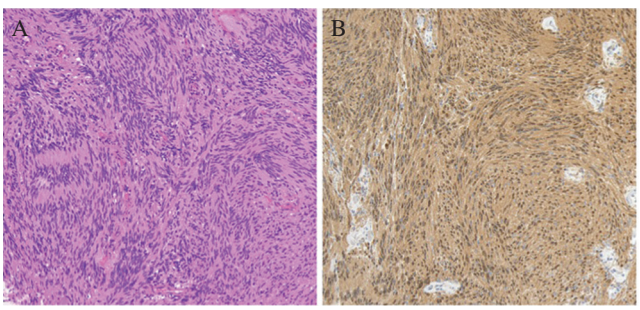

Fig. 3 Pathological findings. (A) Microphotograph showing a histology characteristic of schwannoma. Antoni A and Antoni B patterns are identified (hematoxylin-eosin stain). (B) Tumor cells show diffuse nuclear positivity for S100 protein.

Our search of the English medical literature identified only four other cases of isolated schwannoma of the orbit arising from terminal branches of the abducens nerve to the LRM.[1,3–5)]

Orbital schwannoma on MRI usually appears as hypointense on T_1WI and hyperintense on T_2WI, with homogeneous or heterogeneous enhancement. The histology of these lesions underlies the variation in their appearance on MRI.[2)] Intraorbital abducens schwannoma is situated in the lateral intraorbital quadrant, medial to the LRM.[1,3–5)]

We summarized the main features by comparing our case with those described by Irace et al.,[5)] Rato et al.,[1)] Feichtinger et al.,[4)] and Bhaganagare et al.[3)] (Table 1).

Similar to the other descriptions,[1,3,4,5)] we were able to demonstrate the origin of the schwannoma from the abducens nerve preoperatively on MRI (Fig. 1) and intraoperatively by identifying the area of attachment of the tumor (Fig. 2B).

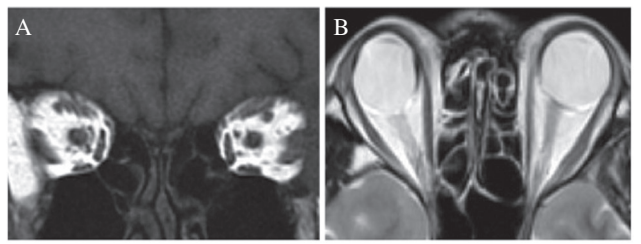

Fig. 4 (A) Postoperative coronal T_1-weighted magnetic resonance imaging (MRI). (B) Postoperative axial T_2-weighted MRI, showing complete tumor removal.

Based on the work of Erdogmus et al.,[7)] Irace et al.[5)] proposed that the entry point of nerve fibers in the muscle (e.g., area nervosa or myoneural junction)[8)] is the zone where intraorbital schwannomas truly arise. When performing intraorbital surgery, anatomic knowledge of the innervation of the extraocular muscles allows the minimization of damage to the neural structures. In particular, when approaching the superolateral compartment of the orbit through a lateral orbitotomy, knowledge of the course and branching of the abducens nerve is essential. This cranial nerve runs on the medial surface of the LRM and innervates the middle third of the muscle, with an average of three small branches entering the muscle.[7)] This knowledge was essential to preserve nerve integrity and function. In our case, we were able to identify the tumor originating from one of these branches. Although we excised one branch of the abducens nerve with the tumor, the other branches of

Table 1 Reported cases of intracranial abducens nerve schwannoma

Case	Author	Age/sex	Clinical presentation	Tumor size (cm)	Tumor type	Surgical approach	Extent of resection	VI nerve function
1	Irace et al., 2008	55/M	Abducens nerve palsy; painless proptosis	?	Solid	Lateral orbitotomy	Total	Partial recovery
2	Rato et al., 2012	42/M	Abducens nerve palsy; painful proptosis	2.2	Cystic	Lateral orbitotomy	Total	Total recovery
3	Feichtinger et al., 2013	53/M	Abducens nerve paresis, optic atrophy	4.5 × 2 × 1.5	Solid	Lateral orbitotomy	Partial	Partial recovery
4	Bhaganagare et al., 2015	32/M	Pain in right eye	2.1	Solid	Superior orbitotomy	Total	Preserved
5	Present case	51/M	-	1	Solid	Lateral orbitotomy	Total	Preserved

the abducens nerve were preserved. We were therefore able to completely excise the tumor while preserving the function of the abducens nerve.

According to the past fourcases,[1,3–5] when the tumor was less than 2.2-cm, the abducens nerve palsy was total recovery after an operation. On the other hand, when it was more than about 4.0-cm, it is difficult to preserve the function of the abducebs nerve (Table 1). Therefore, from a point of view of functional preservation, it had better to remove the tumor in early stage.

Conflicts of Interest Disclosure

The authors have no personal financial or institutional interest in any of the drugs, materials or devices in the article. All authors who are members of The Japan Neurosurgical Society (JNS) have registered online Self-reported COI Disclosure Statement Forms though the website for JNS members.

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