

## Case Report

# Orbital schwannoma of abducens nerve: a case report and literature review

Guihong Li<sup>1</sup>, Yaonan Ding<sup>1</sup>, Chaochao Zhang<sup>1</sup>, Jing Shen<sup>2</sup>, Haiyan Huang<sup>1</sup>

<sup>1</sup>Department of Neurosurgery, The First Hospital of Jilin University, Changchun, P. R. China; <sup>2</sup>Department of Cardiology, Shengze Hospital of Jiangsu Province, Suzhou, P. R. China

Received December 1, 2017; Accepted May 14, 2018; Epub July 15, 2018; Published July 30, 2018

**Abstract:** Intraorbital schwannoma arising from abducens nerve is an extremely rare entity, with only six previous case reports identified in the literature. We describe a case of intraorbital abducens nerve schwannoma in a 54-year old male, who presented with proptosis of the right eye and diplopia. Computed tomography (CT) scan and magnetic resonance imaging (MRI) showed an intraconal well-defined mass in the right orbital apex. The tumor was totally excised with the nerve intact, and the outcome was favorable. The relevant literature regarding intraorbital abducens nerve schwannoma was reviewed, and the clinical manifestations, radiological characteristics, surgical strategies, histopathological findings, and prognosis are discussed. A surgical approach should be elaborately designed for the functional preservation of the abducens nerve, in which MRI is a preferred method for providing valuable guidance. Furthermore, a strong understanding of the complicated anatomical structures surrounding the abducens nerve and lateral rectus is crucial for the complete resection of the tumor and nerve preservation.

**Keywords:** Schwannoma, intraorbital tumor, abducens nerve, orbital

### Introduction

Schwannomas are benign neurogenic tumors arising from Schwann cells of the myelin sheath wrapped around the peripheral nerves. The tumors are usually solitary, slow-growing, and pseudo-encapsulated, which can be seen in almost all the intracranial and spinal compartments [1-3]. The intracranial schwannoma can arise from all cranial nerves except optic and olfactory nerves. Sometimes, the true origin of specific tumor may remain unidentified. Intraorbital abducens nerve schwannoma is an extremely rare entity arising from the terminal branches of the abducens nerve to the lateral rectus muscle, with only six previous case reports identified in the literature [4-9]. Herein, we describe a case of intraorbital abducens nerve schwannoma in a 54-year-old male, who presented with proptosis of the right eye and diplopia. The clinical profile, treatment and prognosis are presented. Furthermore, relevant literature regarding intraorbital abducens nerve schwannoma are reviewed.

### Case report

A 54-year-old man presented with progressive proptosis of the right eye and diplopia. Binocular vision examination showed that the right eye (OD) had 0.4 diopters of myopia with -1.25 diopters of astigmatism, diopter sphere (DS) = 0.8; the left eye (OS) had 0.5 diopters of myopia with -0.75 diopters of astigmatism, DS = 1.0. Intraocular pressure in the OD was 13 mmHg; and in OS 12 mmHg. The anterior segment of eyes and the fundus were normal. The patient lacked any dermatological abnormalities of neurofibromatosis. A further ophthalmologic examination showed a binocular ametropia, while the binocular visual field was roughly normal. Right eye proptosis and right VI nerve palsy was clearly identified. No other neurological abnormalities were found. An intraconal oval mass in the right orbital apex, running along the right optic nerve was observed by computed tomography (CT) scan. The maximum diameter of the tumor reported was approximately 4.5 cm. The involved optic nerve was displaced



**Figure 1.** Computed tomography (CT) scan showed an intraconal oval mass in the right orbital apex, extending along the right optic nerve. The maximum diameter of tumor was approximately 4.5 cm. The involved optic nerve was displaced medially with significant enlargement.

medially with significant enlargement (**Figure 1**). Magnetic resonance imaging (MRI) revealed a 5.4 cm×4.3 cm×5.4 cm well-defined intraconal mass in the posterior right orbit. The intraconal oval mass was medial to the lateral rectus muscle and extended to the superior orbital fissure. The tumor was homogeneously hypointense on T1-weighted images (T1WI), homogeneously hyperintense on T2-weighted images (T2WI), and contrast enhancement tend to be remarkable and heterogeneous (**Figure 2**). Based on above clinical and radiological findings, a preoperative diagnosis of benign intra-orbital neoplasm was made, and surgical excision was planned.

A lateral orbitotomy was performed via an orbitozygomatic approach. An arcuate scalp incision was made at the base of the zygomatic arch, 0.5 cm anterior to the tragus, and was extended to the opposite mid-pupillary line. Orbitozygomatic craniotomy was then performed by removing the orbital rim and the zygomatic arch, followed by dissection of the Tenon capsule through a sharp incision. Intraoperatively, the frontal branch of the facial nerve and upper branch of the ophthalmic artery were properly preserved. A vascularized, pseudo-encapsulated yellowish-brown mass was encountered beneath the lateral rectus muscle. The tumor was confirmed to have originated from the terminal branches of the abducens nerve to the lateral rectus muscle. The tumor was totally excised along the enveloping tissue with an en-

bloc fashion, and the abducens nerve and the lateral rectus muscle were preserved intact.

Histopathological examination of the specimen revealed a pseudo-encapsulated tumor with clear boundaries. The tumor cells were spindle shaped and arranged in fascicles along with Antoni A-pattern structures and loose paucicellular Antoni B-pattern structures, the former occupying the majority of areas. Based on these observations, a diagnosis of schwannoma was made. Positive expression of S-100 protein supported the diagnosis (**Figure 3**).

The right abducens nerve palsy and diplopia remained unchanged after the surgery. The postoperative CT and MRI confirmed the complete resection of the tumor. However, there was no improvement in the thickening of right optic nerve (**Figure 4**).

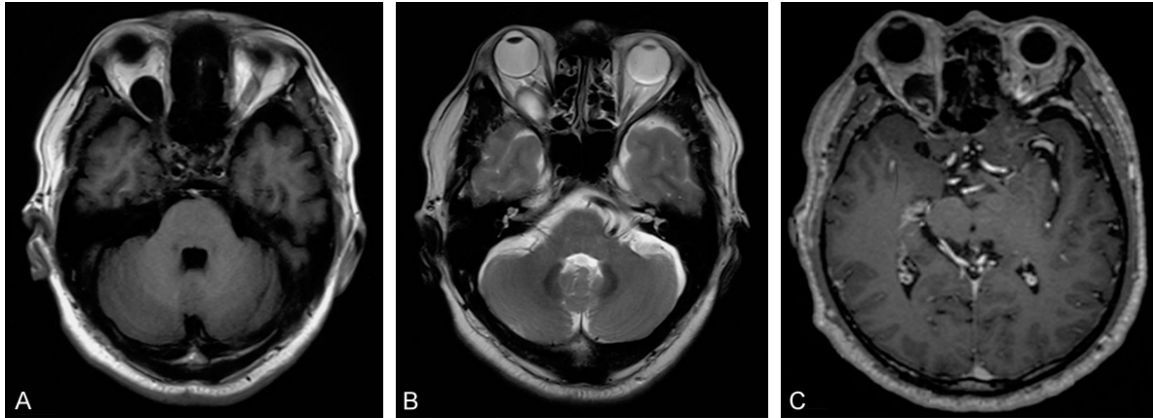
At follow-up two years after surgery, the patient had no complaints of diplopia and there was no detectable abducens nerve paresis. The visual acuity was also significantly improved.

### Discussion

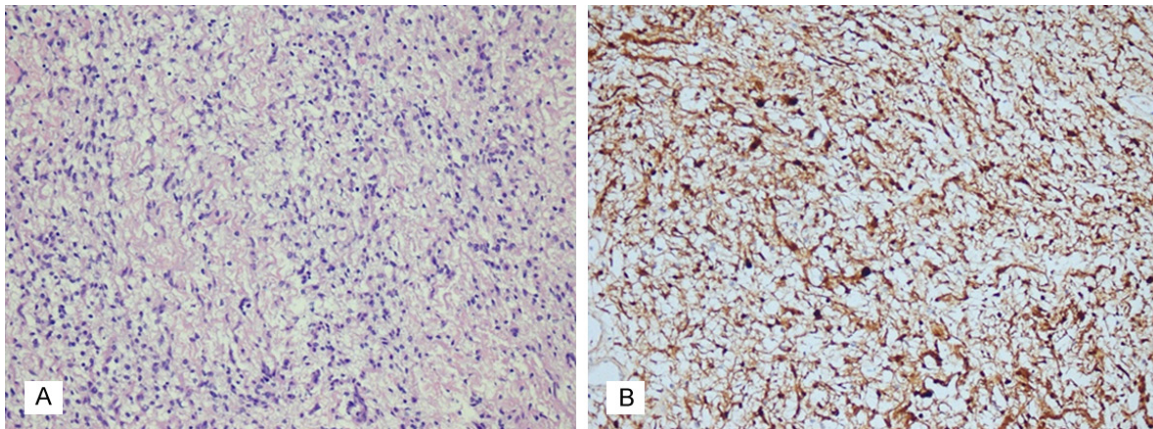
We searched the MEDLINE database for articles related to orbital schwannoma of the abducens nerve published between 1945 and 2017. A total of six publications reporting on three patients were retrieved [4-9]. The clinic radiological profiles are summarized in **Table 1**.

Schwannoma, also known as neurinoma or neurilemmoma, is a benign pseudo-encapsulated tumor originating from the Schwann cells of the peripheral nerves. Verocay, in 1910, described the first case of neurinoma, and the term “schwannoma” was first proposed by Masson in 1932. Both classified this entity as a benign tumor [10]. The tumor is usually solitary, slow-growing and pseudo-encapsulated, in which calcification, hemorrhage, necrosis, and cystic degeneration can also be seen [6, 11]. Although the benign nature of schwannoma has been widely accepted, a few cases with malignant transformation have also been reported [12, 13]. Intraorbital schwannomas account for approximately 1 to 4% of all orbital tumors, and they can occur at any age, without any significant gender predominance [14, 15]. Most intraorbital schwannomas originate from the branches of oculomotor, trochlear, trigemi-

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**Figure 2.** Magnetic resonance imaging (MRI) revealed a well-defined intraconal mass in the posterior right orbit. The intraconal oval mass was medial to the lateral rectus muscle and extended to the superior orbital fissure. The tumor was seen as homogeneously hypo-intense on T1-weighted images (T1WI, A), homogeneously hyper-intense on T2-weighted images (T2WI, B), and contrast enhancement tend to be remarkable and heterogeneous (C).

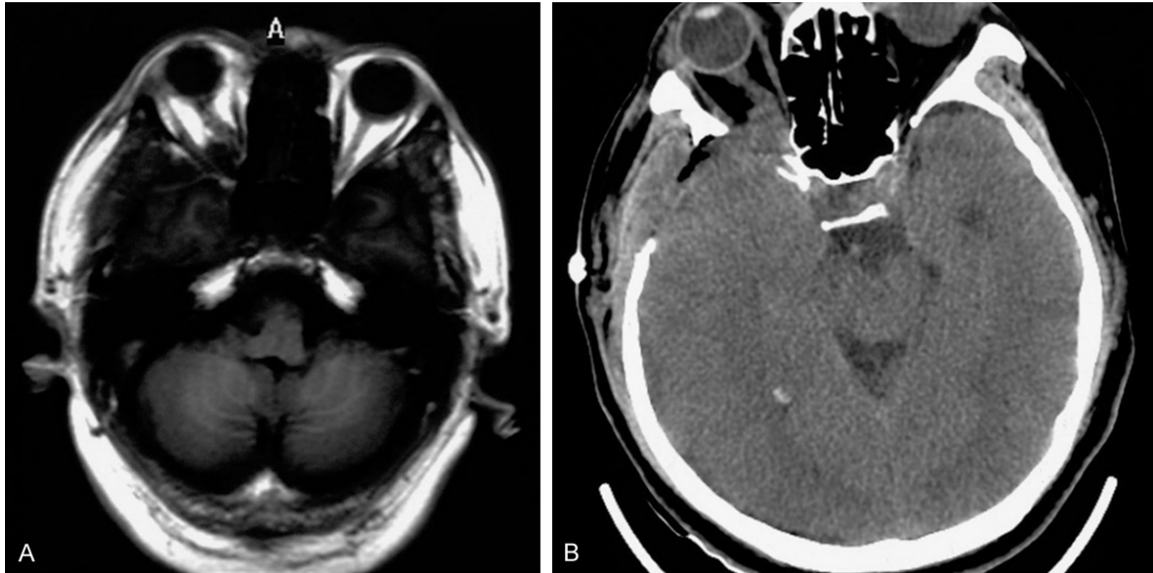


**Figure 3.** Histopathological examination of the specimen demonstrated a pseudo-encapsulated tumor with clear boundary. The tumor cells were arranged in a fascicled growth pattern, and the spindle nucleus arranged in fence. Micrograph of the Schwannoma showed both cellular Antoni A-pattern structures and loose paucicellular Antoni B-pattern structures, the former occupying the majority of areas (A, H&E staining, 100 $\times$ ). The positive expression of S-100 protein also supports the diagnosis (B).

nal, and abducens nerves and from sympathetic and parasympathetic fibers [16-18]. In the majority of cases reported earlier, identifying the true origin of specific tumor was difficult. Housepian et al. hypothesized that intraorbital schwannomas are most frequently originated from the nasociliary nerve [19]. Meanwhile, other studies indicated that the original site may lie on the terminals of supraorbital nerve and other sensory nerves.

A comprehensive understanding of the tumor location, size and infiltrated structures should be emphasized for an effective surgical resection. Patients with intraorbital abducens

schwannoma usually present with diplopia, VI nerve paresis, painful or painless unilateral ptosis, and/or impaired visual acuity. Additionally, some patients with smaller lesions may remain asymptomatic. On MRI, intraorbital abducens schwannomas are generally hypo-intense to isointense on T1WI, and hyper-intense on T2WI, while contrast-enhanced T1WI can show homogeneous, heterogeneous, or annular enhancement. The MRI signal variations may relate to the histopathological differences. Intraorbital abducens schwannomas are mostly located in the temporal quadrant of orbital cavity, and medially to the lateral rectus muscle. In the present case, a mass in the



**Figure 4.** Postoperative MRI (A) and CT scan (B) confirmed a complete resection of the tumor.

orbital apex, extending to the superior orbital fissure was observed, and these findings were consistent with the previous case reported by Feichtinger et al. [7]. The ipsilateral optic nerve can be displaced medially due to a significant enlargement, similar to the radiological manifestations of the optic nerve edema found in the optic neuritis. Differential diagnosis should include the intraorbital neurofibroma, cavernous hemangioma, and MRI as the preferred diagnostic modality.

Prompt diagnosis and early surgical treatment should be implemented to avoid the severe ocular consequences. The resectability of an intraorbital tumor is mainly determined by the surgical approach. Krönlein et al. suggested that lateral orbitotomy was an effective approach for removing intraorbital lateral tumors in 1889 [3, 6]. In subsequent reports, Irace et al. and Feichtinger et al. performed partial resection of solid masses [4, 7], and as Miguel et al. pointed, only cystic tumors were unable to gross total resection via the lateral orbital approach [6]. Therefore, Bhaganagare et al. modified the surgical strategy, and proposed a fronto-orbital approach [5]. The superior frontal approach involves a frontal craniotomy followed by superior orbitotomy, which is effective for removing the tumor from the posterior orbital cavity. With the goal of effective resection of tumors and functional preservation of surrounding structures, we suggest that the indi-

vidual surgical strategy should be based on the location, size, and morphology of the tumors. A lateral orbital approach can be applied for resecting the tumors in the temporal quadrant of orbit. A fronto-orbital approach can be suitable for the tumors in the superior nasal quadrant of orbit and an orbital-zygomatic approach can facilitate a complete resection of the large-size tumors in the posterior orbital cavity or orbital apex. Since most abducens nerve schwannomas are solid, an orbital-zygomatic approach should be superior to a lateral orbitotomy for achieving complete tumor resection.

According to some follow-up studies, schwannoma has been confirmed to be a slow-growing neoplasm with a benign nature, and aggressive total resection may not be necessary, as recurrence was rather infrequent. However, there were also sporadic intraorbital schwannomas, in which the extension to superior orbital fissure hindered a complete resection, progressed several years after the partial removal [20]. In the present case, the tumor was located in the orbital apex and extended to the superior orbital fissure, therefore an orbital-zygomatic approach was scheduled. The tumor was totally resected, and the abducens nerve was functionally preserved.

Identification of the accurate origin of intraorbital schwannoma is difficult. However, Erdog-

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**Table 1.** Reported cases of intracranial abducens nerve schwannoma

No.	Author	Age/sex	Clinical presentation	Preoperative imaging finding	Tumor size (cm)	Surgical approach	Extent of resection	Tumor type	VI nerve function	Follow-up
1	Jack Rootman et al./1982	60/M	Abducens nerve palsy; painful proptosis	CT: A smooth, well-defined oval tumor on the right side of the orbit, with a weak enhancement and a calcified focus in the middle.	Not available	Lateral orbitotomy	Total	Solid Antoni A and B	Total recovery	Not available
2	Irace et al./2008	55/M	Abducens nerve palsy; painless proptosis	MRI: The images of the tumors in the left orbit extended beyond the external rectus, the optic nerve compressed to the medial, and the T1WI showed a mixed signal.	Not available	Lateral orbitotomy	Partial	Solid	Partial recovery	Not available
3	Rui Miguel et al./2012	42/M	Abducens nerve palsy; painful proptosis	MRI: A smooth oval tumor was on the left side of the orbit. The optic nerve was displaced medially without enhancement or atrophy, T1WI showed slightly equal signal, and T2WI showed double signal cystic lesions with liquid and liquid planes, which showed homogeneous ring enhancement after enhancement.	2.2	Lateral orbitotomy	Total	Cystic	Total recovery	6 months
4	Feicbtinger et al./2013	53/F	Abducens nerve paresis, optic atrophy	MRI: Tumor imaging was in the right orbit, the medial rectus muscle and the dorsal aspect of the orbit were extended to the superior orbital fissure. Optic nerve compression was displaced medially and optic atrophy was seen.	4.5×2×1.5	Lateral orbitotomy	Partial	Solid Antoni A and B	Partial recovery	1 year
5	Bhaganagare et al./2015	32/F	Pain in right eye	MRI: Phylloides tumor image in the right eye, close to the lateral rectus, occupying orbital apex, and the optic nerve was inward shift. T1WI was even slightly signal, T2WI showed homogeneous high signal, enhanced homogeneously.	2.1×1.8×1.7	Superior orbitotomy	Total	Solid Antoni A and B	Preserved	8 months
6	Iida, Y., et al./2016	51/M	No abducens nerve palsy or proptosis	MRI: Coronal MRI showed a cinctroconal mass medial to the lateral rectus muscle. The lesion was homogeneously hypointense on T1WI. Preoperative axial T2WI showed smooth contours and the oval shape of the hyperintense lesion. Preoperative sagittal gadolinium-enhanced T1WI showed central enhancement in the lesion.	1	Lateral orbitotomy	Total	Solid Antoni A and B	Preserved	6 months
7	Present case	54/M	Abducens nerve palsy; painless proptosis	MRI: The tumor image in right side of the orbital and close to medial rectus muscle tumor, occupy the orbital apex, extending into the superior orbital fissure, optic nerve inward side shift, and T1WI showed low signal, T2WI showed high signal, enhanced heterogeneous enhancement.	2×1×0.6	Lateral orbitotomy	Total	Solid Antoni A	Total recovery	2 years

mus et al. proposed that the intraorbital schwannoma may arise from the neuromuscular junction where terminal branches of the cranial nerve enter the muscle [21]. In the current case, we observed the tumor arising from the terminal branches of the abducens nerve to the lateral rectus muscle, and an abducens nerve originated tumor could be diagnosed. Moreover, in the literature, the nerve of origin was unidentifiable in nearly half of the patients [22, 23]. Postoperative outcomes may facilitate a definite diagnosis, for instance, the anesthesia in the area of innervations or the extraocular muscle paralysis can help in finding out which is the nerve of origin.

Intraorbital schwannomas are usually solid and completely encapsulated. However, Rato et al. reported a case of a cystic schwannoma in the orbit [6]. The accurate diagnosis of schwannoma depends on pathological criteria.

Histopathologically, schwannomas can be divided into two types: Antoni A pattern, with fascicles of spindle tumor cells with palisaded nuclei; and Antoni B pattern, revealing a loose, vacuolated stroma. Intraorbital schwannomas are usually blending-type, Antoni A pattern occupying the majority of areas, which is in accordance with the present case.

Three out of the six previously reported cases, experienced a complete remission of clinical symptoms. Two had partial recovery of the abducens nerve function, and one remained unchanged. During the follow-up period, the abducens nerve dysfunction completely recovered in the current patient.

We reported an extremely rare case on intraorbital schwannoma. Diagnosis of common tumors at an unusual site requires a high index of suspicion. The surgical approach should be elaborately designed for functional preservation of the abducens nerve, and MRI could provide valuable guidance. Moreover, a strong understanding of the complicated anatomical structures surrounding the abducens nerve and lateral rectus is crucial for complete resection of the tumor and nerve preservation.

### Disclosure of conflict of interest

None.

**Address correspondence to:** Dr. Haiyan Huang, Department of Neurosurgery, The First Hospital of

Jilin University, 71 Xinmin Avenue, Changchun 130021, Jilin, P. R. China. Tel: +86-13578967878; E-mail: huanghy@jlu.edu.cn; Dr. Jing Shen, Department of Cardiology, Shengze Hospital of Jiangsu Province, Suzhou 215000, P. R. China. Tel: +86-13732651876; E-mail: 809998432@qq.com

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