

## Pure Intra-optic Canal Schwannoma: Report of Two Cases

### Abstract

We report two cases of “pure intra-optic-canal schwannoma.” The first patient was a 67-year-old female who presented with a visual field defect and visual impairment in the right eye, and the second patient was a 17-year-old female with progressive visual impairment. Both patients underwent tumor resection through frontotemporal craniotomy combined with extradural anterior clinoidectomy and unroofing of the optic canal. The tumors were not attached to the optic nerve (ON) and were located exclusively inside the optic canal. In both cases, the histological diagnosis was schwannoma. Although the origin of pure intra-optic-canal schwannoma is controversial, intra-operative findings suggested that in these cases, the tumors arose from the sympathetic nerve around the ON.

**Keywords:** *Optic canal, schwannoma, sympathetic nerve*

### Introduction

Schwannomas partially located in the optic canal are rare with only several cases reported, but those confined entirely within the optic canal are much rarer.<sup>[1-3]</sup> Their origin in the optic canal has not been elucidated to date. In this paper, we report two cases of “pure intra-optic-canal schwannoma” where entire tumor body was located inside the optic canal and review the literature regarding this very rare entity. The developmental mechanism and appropriate management will also be discussed.

### Case Reports

#### Case 1

A 67-year-old female was admitted for evaluation of visual field defect and visual impairment in the right eye. Despite steroid pulse therapy for suspected optic neuritis prescribed by an ophthalmologist, her visual acuity worsened over the course of 2 weeks to light perception. Imaging studies were done only after she consulted a neurologist. Magnetic resonance (MR) imaging with three-dimensional (3D) fast imaging employing steady-state acquisition (3D-FIESTA) showed an oval-shaped mass in her right optic canal with a maximal diameter of 3 mm. A T1-weighted MR image after gadolinium-diethylenetriamine pentaacetic acid (Gd-DTPA) revealed a

homogeneously enhanced mass located in the inferomedial side of the right optic canal [Figure 1a and b]. Reproduced with courtesy of the publisher.<sup>[3]</sup> Based on the radiological evidence, a provisional diagnosis of optic canal meningioma was made and was decided to operate upon. The lesion was removed through a right frontotemporal craniotomy combined with extradural anterior clinoidectomy and unroofing of the optic canal. Intraoperatively, the tumor was not directly attached to the optic nerve (ON) and was resected totally in piecemeal. The histological diagnosis was schwannoma with MIB-1 labeling index of 1% [Figure 1c]. The patient had a satisfactory postoperative course and visual symptoms in her operated eye almost normalized within 2 months of surgery.

#### Case 2

A 17-year-old female presented with a 2-week history of progressive visual impairment. Neurological examination revealed that her left visual acuity was restricted to light perception only. A 3D-FIESTA MR image after Gd-DTPA showed an oval-shaped and homogeneously enhanced mass. The tumor had a maximal diameter of 7 mm and was located medio-inferiorly to the ON in the optic canal [Figure 2a and b]. Based on our earlier experience with the similar case, we presumed that the lesion was an optic canal schwannoma. She underwent emergency surgery by frontotemporal craniotomy

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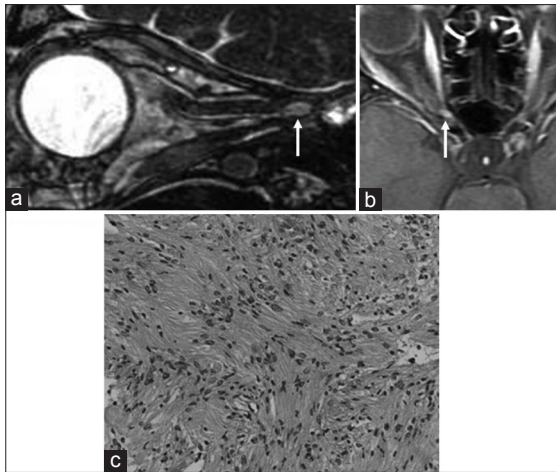
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**Figure 1:** (a) Sagittal section of magnetic resonance imaging with three-dimensional fast imaging employing steady-state acquisition showing an oval-shaped mass (arrow). (b) Axial section of the T1-weighted image after gadolinium-diethylenetriamine pentaacetic acid revealing a homogeneously enhanced mass localized at the inferomedial side of the right optic canal (arrow). (c) Hematoxylin and eosin staining showing schwannoma composed of spindle-shaped cells arranged in Antoni A and Antoni B pattern

combined with extradural anterior clinoidectomy and unroofing of the optic canal. The lesion was easily separable from the ON and was completely removed [Figure 2c]. The pathological diagnosis was schwannoma with MIB-1 labeling index of 3.5% [Figure 2d]. Postoperatively, her visual acuity improved to the level of finger recognition.

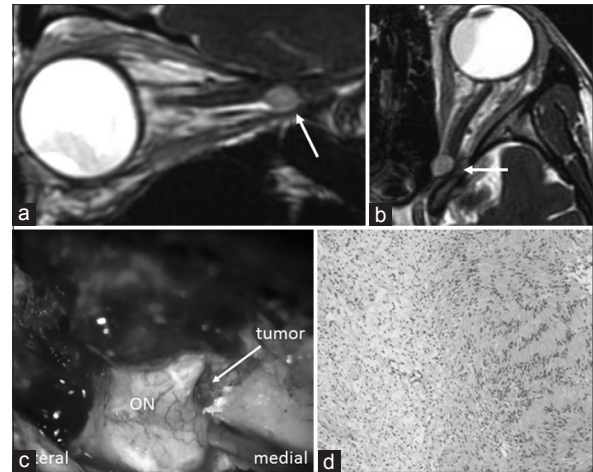
## Discussion

### Pure intra-optic-canal schwannoma as a distinct entity from intraorbital schwannoma

Intraorbital schwannomas are a particularly rare entity with only several case series reported, and they account for 1–2% of all tumors in the orbit.<sup>[1-9]</sup> These tumors usually arise from peripheral nerves in the orbit like the supratrochlear and supraorbital nerves.<sup>[10]</sup> Schwannomas arising in the optic canal are extremely rare because the optic canal has no visible nerve fibers except the ON.<sup>[1]</sup> Theoretically, schwannomas never arise from the ON itself because oligodendrocytes rather than schwann cells ensheath the ON.<sup>[1]</sup> However, intra-optic-canal schwannomas with a large orbital and/or cisternal component do exist, as have been reported in the literature.<sup>[1-4,7-9]</sup> Some of these tumors may have their origins outside of the optic canal.<sup>[4-9]</sup> Thus, “pure intra-optic-canal schwannoma,” defined as a tumor entirely confined within the optic canal, should be considered distinct from optic-canal schwannomas having combined components. As far as we know, only four cases of pure intra-optic-canal schwannoma including ours have been reported in English literature [Table 1].<sup>[1-3]</sup>

### Characteristics of pure intra-optic canal schwannomas

Schwannomas are usually seen in young to middle-aged adults. No significant gender predominance has been identified. Three



**Figure 2:** (a and b) Sagittal (a) and axial (b) section of three-dimensional fast imaging employing steady-state acquisition magnetic resonance images after gadolinium-diethylenetriamine pentaacetic acid showing an oval-shaped and homogeneously enhanced mass located medio-inferiorly to the optic nerve in the optic canal (arrow). (c) Intraoperative microscopic view showing the tumor located medio-inferiorly to the optic nerve in the optic canal. (d) Hematoxylin and eosin staining revealing schwannoma composed of spindle-shaped cells arranged in Antoni A and Antoni B pattern

out of the recorded four patients with pure intra-optic-canal schwannoma were younger than 30 years of age, and all were females. Symptoms were severe visual acuity disturbance and/or visual field defect [Table 1]. Since the optic canal is a narrow bony structure with a mean diameter of 4 mm, the ON can be compressed even with a tiny tumor.<sup>[11]</sup> In the present cases, the reason for the acute progression of visual loss within 2 weeks was uncertain because schwannomas are generally slowly progressive benign tumors. Neither hemorrhagic nor cystic changes were evident, and the proliferative indexes were low in both cases.

All four patients underwent surgical treatment with varying postoperative courses. In both our cases, we approached the tumor extradurally by drilling the anterior clinoid, unroofing the optic canal, and removing the tumor in piecemeal through a small dural incision on the medial side of the ON. The endoscopic approach might have been an alternative for these cases as it avoids external scars and manipulation of the cerebrum.<sup>[12]</sup> However, endoscopic removal is technically demanding, requires special equipment, and our cases required urgent attention, we chose the familiar transcranial approach. Visual symptoms of 2 months duration in Case 1 recovered well after the surgery. In our second case, the improvement in symptoms was only slight despite prompt diagnosis and surgical intervention. Acute progression in the symptom in Case 2 implies greater damage to the ON, which in turn may have contributed to the poor functional outcome.

Two hypotheses for the histopathogenesis of ON schwannomas have been proposed previously: (1) The tumor originates from ectopic schwann cells around the ON<sup>[13]</sup> and (2) the tumor is derived from the sympathetic nerves around the ON.<sup>[14]</sup> All four pure intra-optic-canal

**Table 1: Summary of the cases with pure intra-optic canal schwannoma**

Author	Age/sex	Duration of the symptom before the operation	Side/size of the tumor (mm)	Preoperative visual disturbance	Postoperative visual disturbance	Location of the tumor in the optic canal
Kim <i>et al.</i> <sup>[1]</sup>	10/female	2 months	Right/12 mm	Visual field defect and visual loss (20/30)	Normalized	Medio-inferior
Kinoshita <i>et al.</i> <sup>[2]</sup>	25/female	Not mentioned	Right/10 mm	Visual field defect	Slightly recovered	Medial
Present case	67/female	2 weeks	Right/3 mm	Light perception	Almost normalized	Medio-inferior
	17/female	2 weeks	Left/7 mm	Light perception	Finger counting	Medio-inferior

schwannomas described in the literature were located medial or medio-inferior to the ON. These locations are close to the cisternal portion of the internal carotid artery and distant from the frontal nerve in the superior orbital fissure. These anatomical features appear to support the theory that the origin of pure intra-optic-canal schwannoma is the sympathetic nerve derived from the carotid artery.

#### Differential diagnoses of the tumors in the optic canal

Representative tumors arising in the optic canal are ON glioma and nerve sheath meningioma. Gliomas are a most common primary tumor of the ON and 25–48% of them occur in the intraorbital ON. Imaging studies usually show fusiform enlargement of the ON and the nerve itself cannot be distinguished from the tumor. ON sheath meningiomas are the second commonest ON tumor. They usually have tram track configuration at axial contrast-enhanced MR imaging.<sup>[15]</sup> Because preoperative MR images in the present cases clearly showed oval shaped tumors compressing the ON, optic gliomas, and nerve sheath meningiomas were less likely. Presumptive diagnosis in the first case was meningioma arising within the optic canal. In the second case, we presumed the lesion was schwannoma based on the experience from the first case.

#### Imaging of intra-optic canal schwannomas

Intra-optic-canal schwannomas are difficult to appreciate on routine MR images because of the small size of the tumors and the artifacts caused by the surrounding bony structures and aeration in the paranasal sinuses. Difficulty in discovering the lesion on routine MR imaging can result in a delay of proper diagnosis.

In the present cases, multiplanar reconstructed images from 3D-MR images such as FIESTA and T1-weighted image after Gd-DTPA were effective to reveal the lesion in the optic canal. We should suspect intra-optic-canal schwannoma from the patient's symptoms and clinical history of a progressive visual disturbance when other disorders of the ON are excluded. Surgical removal of the tumor to decompress the ON should be the first choice for treatment.

#### Conclusion

To our knowledge, only 4 cases of pure intra-optic-canal schwannoma have been reported in the English-language

literature. Although the exact origin and developmental mechanism of the tumor have not been elucidated, intra-operative findings in the present cases support the theory that the tumor is derived from the sympathetic nerve in the optic canal.

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#### Conflicts of interest

There are no conflict of interest.

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