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# Optic nerve sheath schwannoma of the orbit: A case report



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## ABSTRACT

**Introduction:** From the anatomical perspective Schwannoma of the optic nerve is impossible, as Schwann cells are absent in the optic nerve. However, there were five cases of optic nerve Schwannoma had been reported. Among the five cases, three were located in orbit.

**Case:** A 26-years-old female presented with gradual decreasing of vision followed by proptosis of the right eye over four years duration. Three weeks before admission the patient was noted blind. The globe was straightly proptosed with ocular motility was restricted to all direction. There was no perception of light despite clear cornea and intact sensation. There were no birthmarks and family history suggesting neurofibromatosis. CT scan showed an ovoid homogenous enhanced tumor that filled most of the orbit and extended into cavernous sinus. Frontotemporal craniotomy with orbital osteotomy

was performed to unroof the optic canal and the superior orbital fissure. The tumor was well encapsulated, and intratumoral decompression with capsule resection were conducted. The optic nerve just behind the globe was fused with the tumor and posteriorly it could not be identified as it was engulfed by the tumor. Postoperatively, the patient experienced temporary ptosis and ophthalmoplegia. The pathological report confirms the diagnosis of optic nerve Schwannoma.

**Conclusion:** This is the fourth case of primary orbital optic nerve Schwannoma that has ever been reported. Since this tumor originates from the optic sheath, decreasing of vision would appear first. The authors suggest to include orbital optic nerve Schwannoma in differential diagnosis of perioptic tumors despite its exceedingly rare occurrence.

**Keywords:** optic nerve sheath, Schwannoma, orbit

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## INTRODUCTION

Schwannomas are benign tumors that originate from Schwann cells in the peripheral and sympathetic nerves.<sup>1-3</sup> Therefore, all cranial nerves that have Schwann cells are potential sites to develop Schwannomas.<sup>4,5</sup> However, it is an exception for the optic nerve.<sup>3,4</sup> Earlier, an anatomical study suggested that Schwannoma originating from the optic nerve sheath would be an anatomically impossible, as there are no Schwann cells in the optic nerve. Therefore, the early studies have suggested that the term of Schwannoma should not be applied for optic nerve.<sup>6</sup> However to the best of our knowledge, there were only five cases of Schwannoma of optic nerve had been reported.<sup>1,4,5,7</sup> Among the five cases, three were located in the orbit.<sup>1,5,7</sup>

In this article, the author further report one case of this extremely rare entity, the primary orbital optic nerve sheath Schwannoma.

## CASE REPORT

**History and examination.** A 26-years-old female presented with gradual decreasing of vision and followed with gradual proptosis of the right eye over the previous four years (**Figure 1A**). The vision was noted totally blind since three weeks before

admission. Physical examination showed the globe was straightly proptosed about 7 mm with the ocular motility restricted to all direction. The visual acuity examination shows no light perception. The cornea was clear with intact sensations and reflex. The pupil was dilated and fixed. There was increased resistance to retropulsion, but no palpable mass. The left eye was essentially normal. The patients have no family history as well as no birthmarks that support the presence of neurofibromatosis. Computed Tomography (CT) scan showed an ovoid homogenous enhanced tumor that filled most of the orbit with extension into the cavernous sinus through the enlarged superior orbital fissure and into the anterior portion of the optic canal, producing an enlargement of that structure (**Figure 2A, 2B**). Furthermore, the medial wall of the orbit was thinned by the mass, and the optic nerve just behind the globe was displaced laterally. The preoperative diagnosis of this tumor included cavernous hemangioma, optic nerve glioma, and meningioma.

**Operation.** Frontotemporal craniotomy with orbital osteotomy was performed. The optic canal and superior orbital fissure were unroofed. After retracting the muscles, a rubbery, pale, hard mass with well encapsulation was identified. The exposed capsule was coagulated, followed by intra-tumoral decompression and piecemeal capsule resection. As

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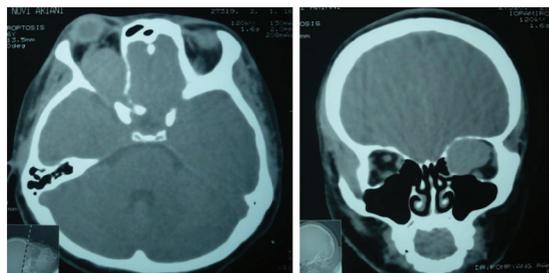
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the tumor size was decreased, the optic nerve just behind the globe was identified. Few millimeters behind the globe, the medial side of optic nerve fused with the tumor (Figure 3). Posteriorly, the optic nerve could not be distinguished from the tumor as it was engulfed by the tumor. A cut was made to the optic nerve just behind the globe. Intratumoral decompression and piecemeal capsule excision were continued. Extension of the tumor into the superior orbital fissure and cavernous sinus limit the surgical resection thus were left unresected (Figure 4). Based on these surgical findings, Schwannoma of the optic nerve was highly suspected.

*Histopathological Findings.* Microscopically, the tumor mass composed of moderately packed elongated spindle cells in interlocking fascicles (Antoni A), intermingled with loose meshwork textured tissue with myxoid stroma (Antoni B) (Figure 5A). Immunohistochemical study showed that the tumor cell was strongly positive for S-100 protein reactivity (Figure 5B). These histopathological findings confirmed a diagnosis of Schwannoma.

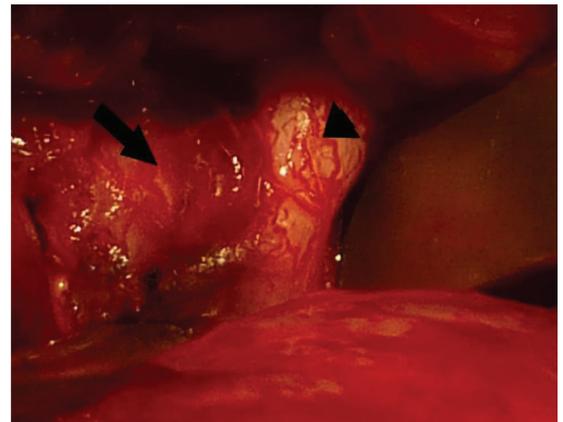


**Figure 1** (A) Preoperative clinical photograph of the patient demonstrating proptosis of the right eye (with permission from the patient). (B) Postoperative clinical photograph of the patient showing that the proptosis of the right eye was completely disappeared and the ptosis was completely improved (with permission from the patient)

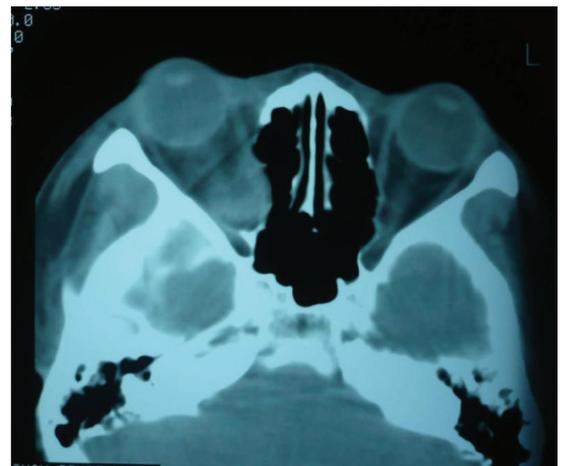


**Figure 2** (A) (axial view) and (B) (coronal view) orbital CT scan of the patient showed an ovoid tumor filling almost all orbit with the posterior side of the tumor completely attached to the apex

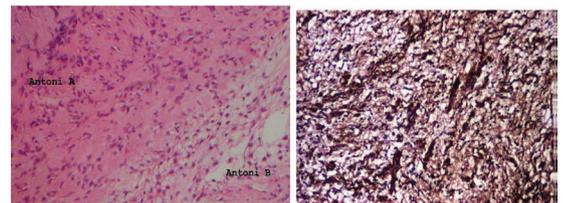
*Postoperative Course.* The patient experienced ptosis and ophthalmoplegia postoperatively. The ptosis was gradually improved and completely disappeared after two months, while the ophthalmoplegia was completely recovered three months after surgery (Figure 1B).



**Figure 3** Intraoperative photograph of the orbital tumor showed the optic nerve (arrowhead) with its medial side fused with the tumor (black arrow)



**Figure 4** Postoperative orbital CT scan of the patient showed a remnant tumor at the apex of the orbit



**Figure 5** A Light microscopy revealed a biphasic pattern of dense cellular Antoni A and hypocellular Antoni B, which consistent with Schwannoma (H & E, x100). Fig 5B Immunostaining of the tumor cell showed diffuse staining of S 100 (x400)

## DISCUSSION

Optic nerve has been considered as an impossible site for Schwannoma since there are no Schwann cells in the optic nerve.<sup>3,4,6</sup> Therefore Schwannoma presumably could not occur in the optic nerve.<sup>3,4</sup> However to the best of our knowledge, there were five cases of optic nerve Schwannoma had been reported.<sup>1,4,5,7</sup> Among the five cases, two were intracranial Schwannoma, and another three were presented as an intraorbital Schwannoma. The authors report further one case of orbital Schwannoma, which is believed to have originated from the optic nerve.

Many histopathogenetic hypotheses of unusual location of this Schwannoma had been proposed.<sup>4</sup> The origin of Schwann cells in this unusual Schwannoma could be from ectopic Schwann cell from neural crest, conversion of mesenchymal cells capable of multipotential differentiation in the pia mater of meninges or Schwann cells ensheathing the small nerve twigs innervating the dura.<sup>4-6</sup> Another possible origin of Schwann cells in this Schwannoma could be from perivascular nerves plexus innervating central retinal artery.<sup>7</sup> Kim et al. believed that the origin of the Schwann cell in their two cases with optic sheath Schwannoma was from perivascular nerve plexus in the central retinal artery.<sup>4</sup> Although there is no conclusive evidence to support the hypothesis, the authors presume that orbital optic nerve sheath Schwannoma reported here, originated from perivascular nerve plexus innervating central retinal artery, as reported by Kim et al.<sup>4</sup>

Schwannoma of the orbit usually presents with slowly progressive proptosis associated with limited ocular movement.<sup>3,8,9</sup> This slowly growing tumor causes late visual impairment.<sup>3,9,10</sup> However, in Schwannoma originating from the optic nerve, decreasing of vision would be the first presenting feature as in our case.<sup>5,7</sup> Kulkarni et al. reported one case of optic nerve Schwannoma presented with the blurring of vision before the appearance of proptosis.<sup>1</sup>

The exceedingly rare occurrence of this tumor makes it difficult to diagnose by imaging findings alone.<sup>4,5,8,9,11,12</sup> In fact, preoperatively the authors diagnosed this patient with cavernous hemangioma, meningioma, and optic nerve glioma. Imaging studies such as CT scan is only helpful to delineate the extent of the tumor and to plan surgical strategies.<sup>5,8,9,12</sup> Therefore, histopathological examination is essential to confirm the diagnosis of Schwannoma, otherwise, it would be clinically confusing.<sup>4,5,9,11</sup> The histological distinction between

Schwannoma and other neurogenic tumors is not difficult.<sup>8</sup> The histopathology of the tumor, in this case, was characteristic of Schwannoma, which was shown by the presence of both Antoni-A and Antoni-B patterns.<sup>3,4,8,11</sup> These microscopic findings were further confirmed by immunohistochemical studies.

Origin of the tumor from the specific nerve could not always be identified even at the time of surgery.<sup>7-9,12</sup> In this case, the authors observed that the medial part of optic nerve fused with the mass just a few millimeters behind the globe and the continuity of nerve was obscured as it was engulfed by the mass. These surgical findings make the authors believe that this presenting Schwannoma originates from the optic nerve.

Since Schwannomas are well-encapsulated tumors, complete surgical removal is usually recommended.<sup>1,3-5,7,8,13</sup> In the author's case, the extension of tumor into the superior orbital fissure and cavernous sinus limited the extent of surgical resection as reported by Rose et al.<sup>14</sup> and Butt et al.<sup>8</sup> Since Schwannomas are benign tumors with a slow growth, therefore they rarely reoccur after a local excision even if the capsule is left behind.<sup>1,8,12</sup>

## CONCLUSION

To the best of author's knowledge, there were only five cases of optic nerve Schwannoma that so far have been published in the English-language literature. Among the five cases reported, only three were located in orbit. In this article, we are reporting another one case. As the visual loss appeared first, the possibility of the optic nerve as the origin of the tumor was very likely. The histopathological diagnosis is essential in this tumor since imaging studies alone would not be able to confirm the diagnosis. Although the occurrence is exceedingly rare, it is essential to include this tumor in the differential diagnosis of the periop-tic tumor.

## CONFLICT OF INTEREST

The authors declare that they don't have any competing interest regarding manuscript

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