Optic Nerve Sheath Schwannoma of the Orbit: A case report
Nyoman Golden, M.D, PhD
Department of Neurosurgery, Sanglah General Hospital School of Medicine, Udayana University, Bali, Indonesia
*Corresponding Email: nyoman_golden@yahoo.co.id

Abstract

From the anatomical point of view schwannoma of optic nerve is anatomically 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 the orbit.

The authors present a case of orbital schwannoma arising from the optic nerve. A 26 year 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 totally blind. The globe was straight proptosed with ocular motility was restricted to all direction. There was no perception of light. The pupil was dilated and fixed. The cornea was clear and the sensation was present. 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 through the enlarged superior orbital fissure and into anterior portion of optic canal producing an enlargement of that structure. On the basis of CT scan findings, the preoperative diagnosis in this case included cavernous hemangioma, meningioma and optic nerve glioma.

Frontotemporal craniotomy with orbital osteotomy was performed. The optic canal and the superior orbital fissure were unroofed. The tumor was well encapsulated. Intratumoral decompression and piecemeal capsule resection were done. As the size of the tumor was decreased, the optic nerve just behind the globe was indentified and the medial side of this nerve fused with the mass. Posteriorly optic nerve could not be identified due to it was engulfed by tumor itself. Extension of some tumors into superior orbital fissure and cavernous sinus limited their surgical resection. Postoperatively, the patient experienced temporary ptosis and ophthalmoplegia. The pathological report was optic nerve schwannoma.

This present case is the fourth case of primary orbital optic nerve schwannoma has ever been reported. Since this tumor originates from optic sheath, decreasing of vision would appeared first. The authors suggest to include orbital optic nerve schwannoma in the differential diagnosis of perioptic tumors although its occurrence is exceedingly rare.

Key words: optic nerve sheath, schwannoma, orbit

Introduction

Schwannomas are benign tumors that originate from Schwann cells in the peripheral and sympathetic nerves. Therefore all cranial nerves that have Schwann cells are potential site to
develop schwannomas. However, it is an exception for the optic nerve. Earlier, anatomical study suggested that schwannoma originating from the optic nerve sheath would be an anatomical 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. However to the best of our knowledge there were five cases of schwannoma of optic nerve had been reported. Among the five cases, three were located in the orbit.

The authors report further one case of the extremelly rare entity of primary orbital optic nerve sheath schwannoma.

Case Report

*History and examination.* A 26 year old female presented with gradual decreasing of vision followed gradual proptosis of the right eye over four years duration. The vision was noted totally blind three weeks before admission. Physical examination showed the globe was straight proposted about 7 mm with ocular motility was restricted to all direction. There was no perception of light. Cornea was clear and sensations were present. The pupil was dilated and fixed. There was increased resistance to retropulsion, but no palpable mass. The left eye was essentially normal. There was no birthmarks suggesting diagnosis and the patient also has no family history of neurofibromatosis. CT scan showed an ovoid homogenous enhanced tumor that filled most of the orbit and extended into cavernous sinus through enlarged superior orbital fissure and into anterior portion of optic canal, producing an enlargement of that structure (Fig.1A). The medial wall of the orbit was thinned by the mass. The optic nerve just behind the globe was displaced to lateral side by the mass. 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 encasulated was identified. The exposed capsule was coagulated, followed by intratumoral decompression and piecemeal capsule resection. As the tumor size was decreased, the optic nerve just behind the globe was identified. Few millimetres behind the globe, the medial side of optic nerve fused with the tumor (Fig.2). Posteriorly the optic nerve could not be distinguished from the tumor due to it was engulfed by the tumor itself. Cut was made to the optic nerve just behind globe. Intratumoral decompression and piecemeal capsule excision were continued. Extension of some tumors into the superior orbital fissure and cavernous sinus limited their surgical resection; thus they were left behind. On the basis of these surgical findings, schwannoma of optic nerve was highly suspected.

*Histopatholgical Findings.* Microscopically, the tumor mass composed of moderately packed elongated spindle cells in interlocking fascicles (Antoni A), intermingled with loosely meshwork textured tissue with mixoid stroma (Antoni B) (Fig. 3). Immunohistochemical study showed that the tumor cell was strongly positive for S-100 protein reactivity (Fig.4). These pathological findings confirmed a schwannoma

*Postoperative Course.* The patient experienced ptosis and ophthalmoplegia postopertively. The ophthalmoplegia was completely recovered 3 months after surgery, while the
ptosis was partially improved. The proptosis was completely disappeared within two months after surgery.

**Discussion**

Optic nerve has been considered as a impossible site for schwannoma since there are no Schwann cells in the optic nerve.\(^4,7,14\) Therefore schwannoma presumably could not occur in the optic nerve.\(^4,7\) However to the best of our knowledge, there were five cases of optic nerve schwannoma had been reported.\(^4,5,11,13\) 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 optic nerve. Many histopathogenetic hypothesis of unusual location of this schwannoma had been proposed.\(^4\) 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.\(^4,11,14\) Another possible origin of Schwann cells in this schwannoma could be from perivascular nerves plexus innervating central retinal artery.\(^13\) Kim et al.\(^4\) believed that the origin of Schwann cell in their two cases with optic sheath schwannoma was from perivascular nerve plexus in the central retinal artery. Although there is no conclusive evidence to support the hypothesis, the authors presume that orbital optic nerve sheath schwannoma in the case reported here originated from perivascular nerve plexus innervating central retinal artery, as reported by Kim et al.\(^3\)

Schwannoma of the orbit usually present with slowly progressive proptosis associated with limited ocular movement.\(^1,2,7\) This slowly growing tumor causes late visual impairment.\(^2,7,12\) However, in schwannoma originating from optic nerve, decreasing of vision would be the first presenting feature\(^11,13\) as in our case. Kulkarni et al.\(^4\) reported one case of optic nerve schwannoma presented with earlier blurring of vision followed by proptosis.

The exceedingly rare occurrence of this tumor makes it difficult to diagnose on the basis of imaging findings alone.\(^1,2,3,4,9,11\) 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 tumor and to plan surgical strategies.\(^1,2,9,11\) Therefore, histopathological examination is essential to confirm the diagnosis of schwannoma, otherwise it would be clinically confusing.\(^2,3,4,11\) The histological distinction between schwannoma and other neurogenic tumors is not difficult.\(^1\) The histopathology of the tumor in authors’ case was characteristic of schwannoma, which was shown by the presence of both Antoni-A and Antoni-B patterns.\(^1,3,4,7\) 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.\(^1,2,9,13\) In this case, the authors observed that the medial part of optic nerve fused with the mass just few millimetres behind the globe and the continuity of nerve was obscured due to it was engulfed by the mass itself. These surgical findings make the authors believe that this presenting schwannoma originates from optic nerve.

Since schwannomas are well encapsulated tumors, complete surgical removal is usually recommended.\(^1,4,5,7,8,11,13\) In the authors’ case, the extension of tumor into the superior orbital
fissure and cavernous sinus limited the extent of surgical resection as reported by Rose at al.\textsuperscript{10} and Butt et al.\textsuperscript{1} Since schwannomas are benign tumors with a slow growth, therefore they rarely reoccur after a local excision even if the capsule is left behind.\textsuperscript{1,5,9}

**Conclusion**

To the best of authors’ knowledge, there are only five cases of optic nerve schwannoma have so far been published in the English-language literature. Among the five cases only three were located in the orbit. Now, we are reporting another one case. As this tumor originates from the optic nerve, visual loss would appeared first. The tissue diagnosis is essential in this tumor since imaging studies alone would not be able to confirm the diagnosis. The exceedingly rare occurrence of this tumor should be born in mind to include this tumor in the differential diagnosis of periorbital tumors.
References

Legends

Fig. 1A Preoperative clinical photograph of patient demonstrating proptosis of the right eye (with permission from the patient)
Fig. 1B Postoperative clinical photograph of patient showing that the proptosis of the right eye was completely disappeared and the ptosis was completely improved (with permission from the patient)
Fig. 2A (axial view) and 2B (coronal view) orbital CT scan of the patient showed an ovoid tumor filling almost all orbit with posterior side of the tumor completely attached to the apex.
Fig. 2C Postoperative orbital CT scan of the patient showed a remnant tumor at the apex of the orbit.
Fig. 3 Intraoperative photograph of the orbital tumor showed the optic nerve (arrow head) with its medial side fused with the tumor (black arrow)
Fig. 4A Light microscopy revealed biphasic pattern of dense cellular Antoni A and hypocellular Antoni B, which consistent with schwannoma (H & E, x100)
Fig 4B Immunostaining of the tumor cell showed diffuse staining of S 100 (x400)