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Case Report

Optic Nerve Hemangioblastomas–A Review of Visual Outcomes

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ABSTRACT

Supratentorial hemangioblastomas (HBLs) are exceedingly uncommon. They account for less than 5% of all central nervous system HBLs. The commonest location is the cerebral cortex. Rarely, they can involve the visual pathway. Most of the reports have focused on the differential diagnosis, since depending on the location they can often resemble optic nerve gliomas or meningiomas of the optic nerve sheath. In this study, we describe a 67-year-old man with progressive diminution of vision in the left eye with an optic nerve hemangioblastoma. There was no history of von Hippel-Lindau disease. We discuss the diagnostic challenges, and review the visual presentation and visual outcomes in these patients. This report, we believe, may add further clarity in counseling patients with an optic nerve lesion and the factors in surgical decision-making.

KEYWORDS: Hemangioblastoma, Optic nerve, Visual acuity, Visual fields, Surgery, Supratentorial

INTRODUCTION

Hemangioblastomas (HBLs) are benign vascular tumors of the central nervous system accounting for approximately 2% of all intracranial neoplasms (3). They are often associated with von Hippel-Lindau (VHL) disease (1). Only 22 cases of optic nerve hemangioblastoma (ONH) have been reported in the literature. They often mimic meningiomas or optic nerve gliomas and are frequently misdiagnosed preoperatively. There has been only one other case of a HBL of the optic nerve producing optic tract oedema (9). We describe a case of an ONH without VHL, mimicking a meningioma preoperatively with optic tract edema. We review the visual outcomes of these patients as they have significant clinical implications. Such a review has not been done before in the literature.

CASE REPORT

A 67-year-old man presented with a 1-year history of episodic blurring of vision in the left eye. He also complained of seeing halos around objects as well as headaches. His visual acuity was 20/60 in the left eye and 20/20 in the right eye and visual field testing revealed concentric narrowing. Fundoscopic examination revealed left primary optic atrophy. Endocrine examination was normal.

The magnetic resonance imaging (MRI) (Figure 1A-D) showed a 1.8 cm extra-axial, well-defined, slightly lobulated lesion adjacent to the left anterior clinoid process and planum sphenoidale, compressing the optic nerve and the chiasm. The anterior cerebral artery (A1) was abutting the posterior surface of the tumor but was not encased. Most of the tumor was hypointense on T1-weighted images, enhanced uniformly, and showed heterogeneous intensity on T2-weighted images. There was a small cystic component to the tumor postero-inferiorly. The tumor did not have an obvious dural attachment and a dural-tail could not be identified. It was clearly separate from the pituitary gland and pituitary stalk. There was compression of the left optic nerve with no clear plane of separation. Likewise there was edema along the optic tracts bilaterally, more so on the left side. The preoperative

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diagnosis was that of meningioma, although several imaging features of the lesion were atypical for meningioma, including lack of dural base, a dural tail and the presence of a cystic component of the tumor.

In view of his visual symptoms and symptomatic nature of the lesion, surgery was recommended. A mini-pterional craniotomy was carried out and the lesion was exposed by splitting the sylvian fissure. The lesion was seen to be arising directly from within the left optic nerve with no dural attachment. There was extension into the optic canal. It was reddish, vascular with small feeding vessels from the anterior cerebral and anterior communicating arteries, which were coagulated and cut. It had all the characteristics of a hemangioblastoma. Although it was firmly attached to the optic nerve, a total excision was achieved after careful dissection from the optic nerve.

The histopathology demonstrated a highly vascular tumor composed of proliferating capillaries and epitheloid and foamy stromal cells with nuclear atypia. The cells were positive for inhibin immunostain and negative for glial fibrillary acidic protein (GFAP), epithelial membrane antigen (EMA) and PAX-8. The diagnosis was consistent with hemangioblastoma. A diagnosis of VHL was ruled out in the absence of family history of VHL and normal blood work and an abdominal computed tomography (CT) scan. Genetic testing was not done.

Postoperatively his vision was significantly worse with only perception of light in the left eye but he retained normal vision in the right eye. This remained unchanged at a 3-month follow up. The postoperative MRI showed no residual tumor with near complete resolution of the optic tract oedema (Figures 2A, B; 3A, B).

■ DISCUSSION

HBLs are uncommon tumors of the central nervous system of uncertain histogenesis and principally composed of stromal and endothelial components (3). The cell of origin is believed to be the hemangioblast subset of mesodermal cells that are embryologically arrested (5). They typically occur in the cerebellum, brain stem and spinal cord. Supratentorial HBL's (60% associated with VHL) are exceedingly rare. Two thirds of these are solid and one third cystic. While most of these lesions are cerebral in location, a cranial nerve origin of these tumors is infrequent (8). Only 22 cases of HBL (including our case) arising from the optic nerve have been described, 18 of which are in the English literature and 4 reports are in Japanese (9). HBLs arising from the sella, stalk and hypothalamus has also been reported but are excluded from this review, which attempts to focus primarily on visual outcomes of pure ONHs.

The most common tumor of the optic nerve is the optic nerve glioma. While most often these are benign, malignant variants

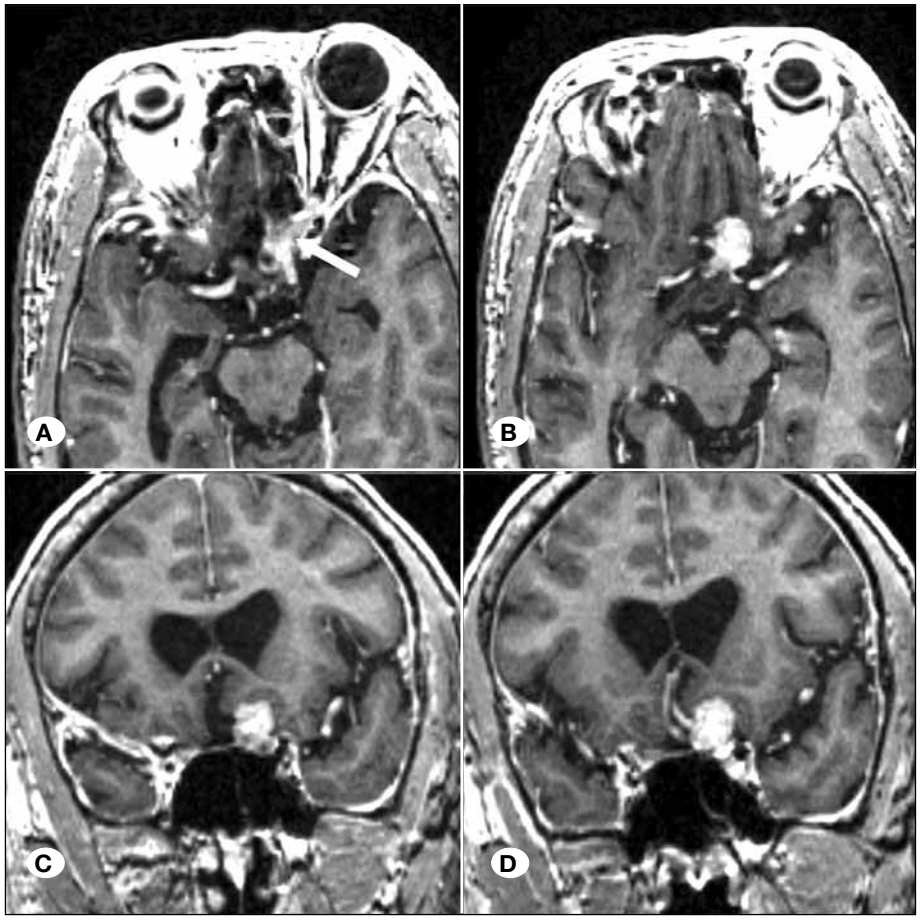

Figure 1: A, B) Preoperative gadolinium enhanced axial MRI of a 67-year-old man showing a well-defined 1.8 cm enhancing lesion arising from the optic nerve and extending to involve the optic canal (arrow). **C, D)** Gadolinium-enhanced coronal MRI showing close proximity to the branches of the anterior cerebral artery.

do occur. Other less frequently occurring tumors include gangliogliomas, medulloepitheliomas, lymphoma, metastasis, sarcoidosis and hemangioblastomas. Those that arise from the optic nerve sheath include meningiomas, schwanommas and hemangiopericytomas (2). Most cases of ONH were diagnosed post-operatively on histopathology. In the majority of patients, the preoperative diagnosis was either a glioma or meningioma.

The classic HBL has an intensely enhancing nodule along the wall of a non-enhancing cyst. Larger HBLs often have visible irregular shaped flow-voids adjacent to the nodule although this feature is usually not seen with smaller HBLs. If seen, flow voids alongside a cystic tumor are highly suggestive of HBL, especially with a background of VHL. Although the lack of a dural base and dural-tail, and small cystic component were all considered atypical for meningioma, this was still considered the most likely diagnosis pre-operatively. The presence of a cystic component in a suprasellar tumor also invokes consideration of craniopharyngioma and pilocytic astrocytoma. The clear separation from the stalk made the former diagnosis unlikely. Optic tract edema, a prominent finding in this case, can be observed with any tumor that compresses the nerves or chiasm, and thus this finding is not particularly helpful in the differential diagnosis. Of note, optic tract edema is uncommon with optic gliomas.

Various surgical approaches have been described. Purely intraorbital tumors can be removed using a lateral orbitotomy approach, while those with an intracranial extension require craniotomy. The endoscopic approach has met with mixed outcomes often with the inability to resect the tumor completely and with postoperative worsening in vision (1). While tumors that arise inferior to the nerve, elevating it superiorly, could possibly be approached endoscopically, this is not feasible in tumors arising from the superior surface of the nerve and abutting the orbital surface of the frontal lobe. These tumors have been reported to invade optic nerve tissue (4). Significant visual deterioration, including blindness, can occur post-operatively either secondary to physical manipulation of the nerve, or more often by interruption of its complex vasculature. The visual outcome depends on the origin of the lesion and whether it arises from within the nerve or more superficially, where it can be dissected off the pia. While in the former case there is a high risk for severe visual compromise, in the latter case, patients have a better chance for visual recovery. In our patient, the lesion had an extensive and deep attachment to the optic nerve. While dissection was possible and total

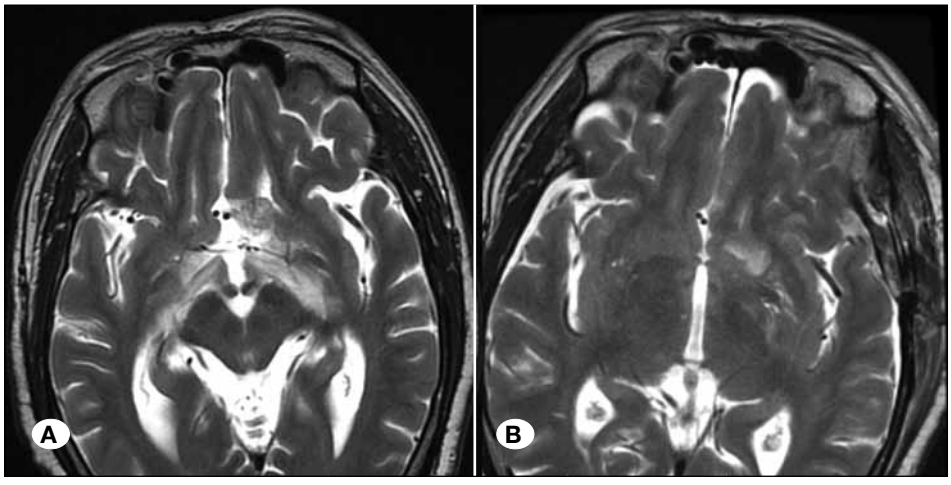

Figure 2: A, B) Pre and postoperative T2-weighted axial images showing bilateral optic tract oedema with complete resolution of the same after surgery.

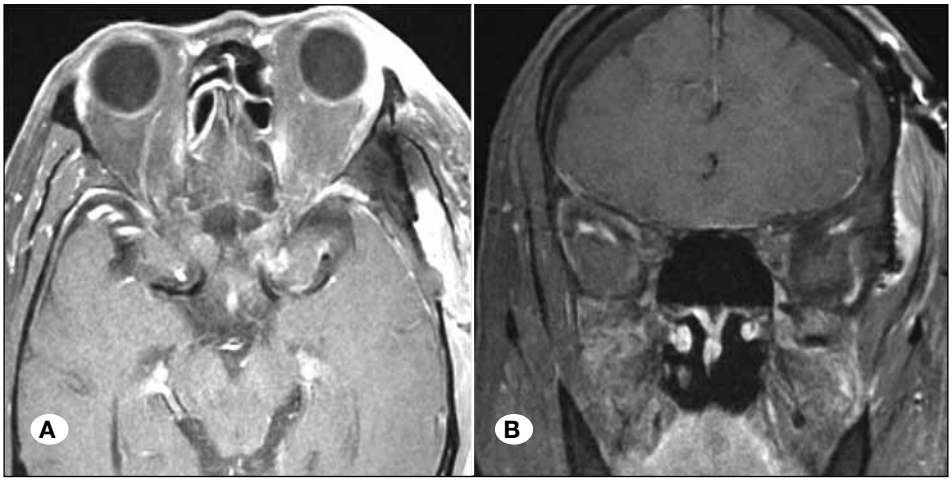

Figure 3: A, B) Postoperative Gadolinium-enhanced axial and coronal MRI showing no residual tumor.

Table I: Details of Visual Presentation and Outcome in 22 Patients with ONH Published in the Literature

Author	Size	Location	Pre op VA	Pre op VF	Post op VA	Post op VF	Follow up	VHL	Pre op diagnosis
Stefani (1974)	11 mm	IC	5/35	Enlarged blind spot	No sx	No Sx	Blind with no sx	No	Retrobulbar neuritis
Lauten (1981)	15 mm	IC/PC	PL+	NR	NR after sx	NR	NR	No	Glioma
Eckstein (1981)	23 mm	IO	PL+	Temporal hemianopia	PL- after orbital exenteration	PL-	NR	No	Vascular neoplasm
In (1982)	29 mm	IO	PL-	PL-	PL- after TC surgery	Blind	NR	Yes	Hemangioma/ Meningioma
Tanaka (1984)	NR	IO	20/200	Inferior quadrantanopia	PL- after TC sx, optic nerve resection	PL-	NR	Yes	Hemangioma/ Meningioma
Nerad (1988)	NR	IO	20/60	NR	20/200 after TC biopsy	NR	5 months	Yes	HBL
Hotta (1989)	16 mm	IO	PL-	PL-	PL after TC surgery	PL-	NR	Yes	Hemangioma
Ginzburg (1992)	10 mm	PC	Reduced VA	NR	PL- after TC sx	PL-	NR	Yes	Glioma
Rubio (1994)	20 mm	IO/IC	PL-	PL-	PL-	PL-	NR	Yes	NR
Miyagami (1994)	1 cm	IC/PC	20/20	Normal	NR	NR	5 years	Yes	HBL
Kerr (1995)	10 mm	IC	20/400	Temporal hemianopia	CF at 30 cm after TC surgery	Further constriction	2 yrs	Yes	Meningioma
Raila (1997)	10 mm	PC	20/20	Normal	20/20	Normal	5 months	Yes	HBL
Kato (2004)	NR	IO/IC/PC	PL-	PL-	PL- after TC Sx and optic nerve resection	PL-	NR	No	Meningioma/ HBL
Higashida (2007)	NR	IO	HM alone	Tunnel vision	Unchanged after TC surgery	Same as pre op	NR	No	Glioma meningioma schwannoma
Barrett (2008)	45 mm	IO/IC	PL-	NR	PL- after TC Sx	PL-	NR	Yes	Glioma
Meyerle (2008)	18 mm	IO	20/50	Triquadrant defect	Died before Sx from pheochromocytoma	Died	NR	Yes	HBL
	NR	PC	20/30	NR	20/20 after EEA	Nasal quad defect	NR	Yes	HBL
	10	IC	CF at 3 feet	NR	20/800 after EEA to 20/320 at 6 months	NR	6 months	Yes	HBL
	10 mm	IO	20/20	Normal	20/15 after sx	Normal	12 years	Yes	HBL
Prabhu (2009)	3.5 cm	PC	6/60	Temporal field cut	Unchanged after TC sx	Same as pre op	3 months	Yes	Meningioma
Zywicke (2012)	10 mm	IC/PC	20/70	Normal	HM + after TC surgery	Field loss	NR	Yes	Meningioma
Staub (2014)	12 mm	IO/IC/PC	PL-	PL-	PL- after TC sx	PL-	NR		Glioma
Turel (2016)	18	PC/IC	20/60	Normal	HM Positive	Temporal field	3 months	No	Meningioma

***VA:** Visual Acuity, **VF:** Visual Fields, **VHL:** von Hippel Lindau, **IO:** intraorbital, **IC:** intracanalicular, **PC:** Pre-chiasmal, **Sx:** Surgery, **PL:** perception of Light, **HM:** Hand movements, **TC:** Transcranial, **EEA:** Endoscopic endonasal approach, **HBL:** Hemangioblastoma, **NR:** Not reported.*

removal was achieved, the significant worsening of vision post-operatively was likely related to the devascularization of its blood supply.

Details of visual outcome with follow-up are described in Table I. There is inconsistent documentation of detailed visual acuity, fields and fundoscopic findings both pre-and postoperatively among the reports and hence several columns are not reported (NR).

VHL patients who present with loss of vision not consistent with a retinal examination should be evaluated for an ONH. The initial reports on ONHs published in the 70's and 80's were mostly of patients who were operated only after the vision was severely compromised to the extent that it was non-functional. In patients with minor visual symptoms, the operation was often delayed until they were almost blind or when proptosis and headache were the main symptoms. Most other reports of patients with significantly compromised but functional vision, who underwent surgery, were worse post-operatively. In only 2 patients did the vision remain unchanged (1,6). One patient had improvement in vision (from 20/30 to 20/20 with a persistent visual field defect) after surgical excision (1). The visual outcomes also depended on the anatomical location of the tumor. Tumors with an intracanalicular component (IC) had a worse outcome than tumors with an intraorbital (IO) or a pre-chiasmal (PC) location. The two cases in which vision improved were both PC in location with no IC extension.

Raila et al. (7) have described a successful excision in an asymptomatic patient with a pre-chiasmatic ONH. Their rationale and indication for surgery was prevention of visual deterioration since left untreated most eventually lead to blindness. It is unclear from the literature whether surgery should be recommended when the vision is minimally affected or when it is severely compromised. Another consideration is the visual function in the other eye since patients with VHL can harbor retinal angiomas. Most authors recommend close radiological and ophthalmological follow-up after the diagnosis of an ONH is made and would consider surgery based on the visual progression and growth in the size of the lesion.

Survival is significantly longer with total excision than sub-total resection and adjuvant therapy. The outcome of subtotal resection and fractionated radiotherapy for ONH has not been separately documented due to their rarity. Radiosurgery would not be considered as a primary treatment option in patients with good vision. In patients with no functional vision, this has been used as a form of treatment with reasonable tumor control rates. However due to their infrequent occurrence no distinction was made from the cohort of supratentorial HBLs.

In an analysis of all supratentorial HBLs carried out by Mills et al. (3), the five-year progression-free survival for gross total resection and sub-total resection were 100% and 53% respectively. Solid tumors seem to have a higher rate of recurrence. The major issue is whether to consider a radical resection in the face of relatively normal vision and a lesion invading the optic nerve tissue where the risk of visual deterioration post-operatively is very high. For pre-chiasmal tumors that arise distal in the optic nerve and do not invade the optic nerve, it may be reasonable to offer surgery to prevent visual deterioration. However, for tumors within the optic canal, unless there is evidence of significant growth our recommendation would be to delay surgery since visual outcomes with surgery are universally poor.

CONCLUSION

HBL affecting the optic nerves is rare and should be considered in the differential diagnosis of patients with visual disturbance, not only in patients with VHL but also in sporadic cases. They may be suspected radiologically by the presence of flow voids and the absence of a dural attachment. The choice of surgical approach (endoscopic versus transcranial) depends on its location in relation to the optic nerve. The visual outcome depends on the preoperative visual status and the location of the tumor in relation to the optic nerve and canal. Most patients show worsening vision after surgery, thus mandating careful discussion with the patient.

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