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Case Report

Hybrid nerve sheath tumor in the orbit: A case report and review of literature

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ABSTRACT

Background: In neurosurgical practice, we rarely encounter hybrid nerve sheath tumors (HNST) in the orbit. We recently had a patient of this rare tumor. We believe that this is the first report where we resected the tumor transcranially.

Case Description: A 54-year-old male presented with the left proptosis and intraconal tumor of 43 mm. We performed fronto-orbital craniotomy to resect the tumor mass. His proptosis completely improved and discharged home with a modified Rankin Scale of 1.

Conclusion: Transcranial resection of orbital HNST was a safe and effective way to treat. Since we do not have much data regarding this rare tumor, we need to accumulate more cases.

Keywords: Exophthalmos, Hybrid peripheral nerve sheath tumor, Pterional

INTRODUCTION

Few case reports have been made regarding the hybrid nerve sheath tumor (HNST) in the orbit. HNST was officially enrolled as a distinct tumor entity relatively recently[8] and is defined as benign peripheral nerve sheath tumors with combined features of more than one conventional type, of which neurofibroma/schwannoma and perineurioma/schwannoma being more common.[2]

CASE REPORT

History and examination

A 54-year-old male had the left proptosis pointed out on his regular health checkup. Subsequent computed tomography showed tumor in the left orbit. He was referred to our hospital for further evaluation. Physical examination was unremarkable except for the left proptosis with normal visual acuity (20/320 oculus dexter and 20/320 oculus sinister) and field. He had no diplopia, nystagmus, or stigmata of neurofibromatosis. Computed tomography showed an apparent proptosis of the left eye [Figure 1a]. Magnetic resonance imaging revealed well-demarcated tumor of 43 mm compressing the optic nerve medially [Figure 1b and c]. Digital subtraction angiogram showed no apparent tumor blush or abnormal vessels. However, VasoCT showed

some feeders from the ophthalmic artery, which is coursing around the anterior pole of the tumor [Figure 1d]. The differential diagnosis included cavernous hemangioma, schwannoma, and meningioma. Considering the tumor size and proptosis, we decided to perform surgical resection for definite diagnosis and to prevent visual dysfunction.

Operation

We administered general endotracheal anesthesia and placed lumbar drain. We positioned the patient supine with the head turned 20° to the right and fixed in Mayfield head holder. A curvilinear skin incision was made from just anterior to the tragus to the forehead behind the hairline. Skin flap and the temporalis muscle were reflected in two layers and frontotemporal craniotomy was done. Orbital bar was removed with bone saw lateral to the supraorbital foramen. The greater wing of the sphenoid was rongeured off until temporal dura was exposed. The lesser wing of the sphenoid was rongeured off toward sphenoid ridge until meningo-orbital band was exposed. The superior and lateral wall of the orbit was rongeured off to expose the periorbita. We incised the periorbita in between the superior and lateral rectus muscles and identified the tumor [Figure 2a and b]. The tumor was grayish, firm, and the vascularity was mild. Internal decompression using cavitron ultrasound surgical aspirator [Figure 2c] was performed followed by the tumor dissection from the adjacent tissue and resection in a piecemeal fashion. The optic nerve was identified medially displaced near the orbital apex [Figure 2d]. After resection, we closed the wound in layers. Estimated blood loss was minimal.

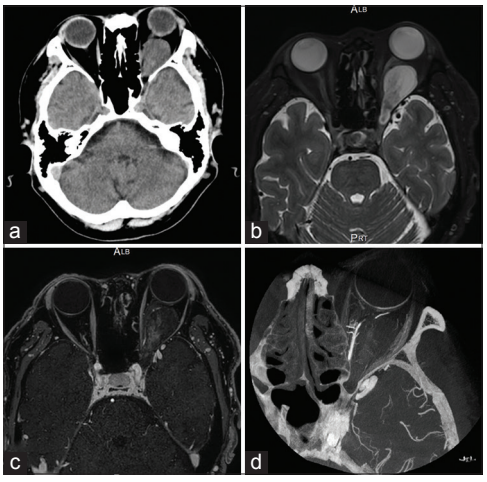

Figure 1: Computed tomography showed the left intraconal orbital tumor compressing the globe with resultant exophthalmos (a). T2-weighted magnetic resonance imaging (MRI) showed a hyperintense tumor (b). Gadolinium-enhanced fat suppression T1-weighted MRI showed a heterogeneous enhancement (c). VasoCT showed some feeders from the ophthalmic artery (d).

Histopathological findings

Histology showed loosely proliferating spindle tumor cells (neurofibromatosis [NF] component) and nuclear palisading tumor cells (schwannomatous component). No clear border between the two components was observed. On immunohistochemical analysis, S100 protein was slightly and strongly positive in NF and schwannomatous component, respectively. Neurofilament staining showed axonal filament in NF component [Figure 3]. Epithelial membrane antigen was negative. Ki67 was 1.2% in hot spots. The histopathological diagnosis of HNST, the WHO Grade I was made.

Postoperative course

His postoperative course was uneventful except for abducens nerve palsy and ptosis. The postoperative imaging study showed completely improved proptosis and substantially reduced tumor as well as decompression of the optic nerve [Figure 4]. He was discharged at a modified Rankin Scale of 1. His abducens nerve palsy and ptosis recovered completely 4 months after surgery.

DISCUSSION

To the best of our knowledge, this is the second report of intraconal orbital HNST. HNST was first described in 1998 and only recently included in the WHO classification in 2013. Due to its rarity, the exact incidence is not known. However, based on the available literature, it is diagnosed most commonly in young adults with no gender predilection.[8] It often arises in the skin and is much less common within the central nervous system.[2]

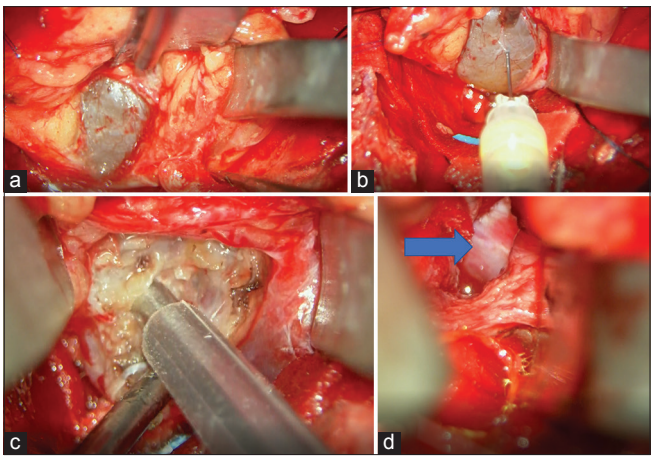

Figure 2: Tumor capsule was revealed (a). We aspirated the tumor (b), which was not effective. Note the gelatinous tumor content (c). After the appropriate amount of tumor resection, optic nerve (arrow) was observed (d).

Table 1: Previous case reports of orbital HNST.

Author, Year	Age/ Sex	Presenting symptoms	Size (cm)	Site side	Surgery EOR	Characteristics of mass lesion	Pathologic components	F/U period (mos)	Postoperative course	Stigmata of neurofibromatoses
Youens *et al.*,[12] 2008	51 F	Right brow pain, hypesthesia	2.1	Extraconal right	Anterior orbitotomy ND	Well demarcated, firm, gray-tan myxoid nodule	Neurofibroma, schwannoma	ND	Hypesthesia improved	None
Stevenson *et al.*,[6] 2019	68 F	Incidental	2.9	Extraconal right	Anterior orbitotomy GTR	Well demarcated, soft, and yellow-gray color; supplied by ICA branches; supraorbital nerve origin	Neurofibroma, schwannoma	1 (still under f/u)	Hypesthesia	None
Verhelst *et al.*,[9] 2017	39 M	Diplopia, visual loss, limited eye movement	2.5	Intraconal left	Transconjunctival ND	Thin capsule; red-bluish mass	Neurofibroma, schwannoma	ND	Visual acuity/ diplopia improved	None
Taubenslag *et al.*,[7] 2017	31 M	Ptosis, proptosis	3.4	Extraconal left	Anterior orbitotomy GTR	Homogeneous, smooth, tan-yellow, unencapsulated, gelatinous; supraorbital nerve origin	Neurofibroma, schwannoma	Lost to f/u	ND	None
Our case	54 M	Proptosis	4.3	Intraconal left	Frontotemporal craniotomy STR	Firm, gray-tan, gelatinous, mild vascularity	Neurofibroma, schwannoma	6 (still under f/u)	Proptosis improved; mild CN VI palsy, no relapse	None

Note our case was the largest in tumor size, which necessitated to perform transcranial resection. CN: Cranial nerve, EOR: Extent of resection, F: Female, f/u: Follow-up, GTR: Gross total resection, M: Male, mos: Months, ND: Not described, STR: Subtotal resection, ICA: Internal carotid artery, HNST: Hybrid nerve sheath tumor

HNST from pathological standpoint

HNST is mainly a mixture of two or more out of the following: neurofibroma, perineurioma, and schwannoma.[2,4] Of note, other types of benign peripheral nerve sheath tumors also constitute the spectrum of HNST, such as neurothekeoma/perineurioma, granular cell tumor/perineurioma, and perineurioma/schwannoma admixed with melanocytic elements.[1,3,10,11] The past reports on HNST in the orbit are summarized in Table 1.[6,7,9,12] Two out of five orbital HNSTs were intraconal. All cases were the mixture of neurofibroma and schwannoma, and none had perineurioma component. Neurofibroma/schwannoma mixture is typically associated with neurofibromatoses, especially schwannomatosis (NF type 3).[2,4] As a matter of fact, methylome and chromosomal profile of neurofibroma/schwannoma was similar to that of schwannoma.[5] However, none, including ours, had signs of neurofibromatoses, which suggests that orbital HNST may occur in isolation.

HNST from clinical standpoint

In this paper, we described the intraoperative detail of orbital HNST, which was resected transcranially. All of the previous orbital HNSTs were resected through anterior orbitotomy. However, in our case, the tumor size was the largest of the reported cases [Table 1], and we considered it to be appropriate to perform frontotemporal craniotomy to get a wider corridor. We did not stick to gross total resection because opening the annulus of Zinn may have the risk of ophthalmoplegia. As orbital HNST is rarely reported, we do not know the relapse or recurrence rate of these tumors, which leads to the necessity of the tight follow-up. Further, accumulation of cases on the orbital HNST is necessary so as to understand these rare tumors.[7,8] The entity of HNST should be widely known to those dealing with orbital lesion including but not limited to neurosurgeons and ophthalmologists.

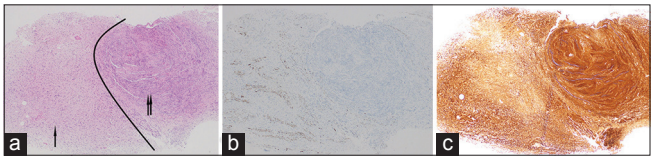

Figure 3: Hematoxylin and eosin stain showed neurofibromatosis (arrow) and schwannomatous components (double arrow). Arbitrary black line is drawn (a). Neurofilament stain showed positivity in the neurofibromatosis component (b). S100 stain showed strong positivity in the schwannomatous component (c). Original ×40.

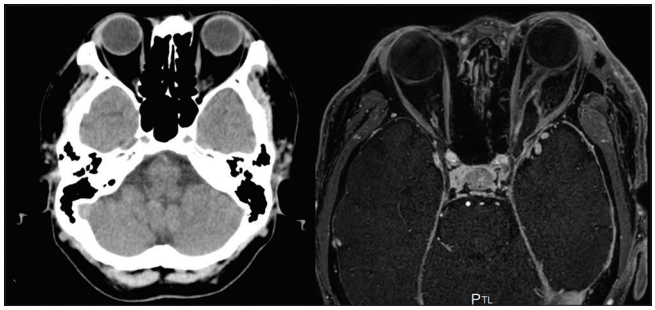

Figure 4: Postoperative imaging. Computed tomography showed the proptosis disappeared (left). Gadolinium-enhanced fat suppression T1-weighted magnetic resonance imaging showed subtotally resected tumor and well-visualized optic nerve. Note the optic nerve compression was relieved (right).

CONCLUSION

HNST should be included in the differential diagnosis of well-demarcated orbital tumor.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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