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Case Report

Abducens nerve schwannoma of the cavernous sinus: A case report and literature review

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INTRODUCTION

Schwannoma of the abducens nerve in the cavernous sinus (with or without extension into other anatomical corridors such as the orbit or prepontine cistern) is an uncommon entity.[22] Since 1982, 10 cases within the cavernous sinus proper[5,10,17,19-22,30,32] and six cisternocavernous cases[1,8,18,29,32,35] have been reported. Due to limited numbers of these tumors, multiple classification methods were proposed based on imaging or symptomatology. This was to achieve a better management approach and consequently improve outcome of patients. Hence, we report a case of sixth cranial nerve schwannoma within the cavernous sinus, with a review of literature.

CASE REPORT

A 50-year-old woman with no known medical history presented with a 2-year history of double vision and left facial numbness that started 6 months before presentation. On examination, she had a left sixth cranial nerve palsy manifested by the left eye esotropia with limited abduction on lateral gaze, paresthesia over distribution of ophthalmic and maxillary divisions of the left trigeminal nerve. There were no other positive signs on examination with no stigmata of neurofibromatosis. An ophthalmologist conducted a formal ophthalmological examination that did not reveal any additional
deficit. Magnetic resonance imaging showed heterogeneous T2 hyperintense lobulated mass at the left cavernous sinus extending into Meckel’s cave with bony remodeling. The mass also extended into left preptontine cistern, with avid contrast enhancement no imaging evidence of intratumoral cystic degeneration or dural tail. The images are shown in [Figure 1].

Considering the clinical status of patient and imaging findings, we decided to proceed with surgery. Lumbar drain was inserted preoperatively, and 40 cc of CSF was drained before craniotomy. Then, the patient underwent left frontotemporal craniotomy, through the peritoneal approach, drilling of lateral sphenoid wing was performed. Initially, the extradural approach was utilized but the dura was tense. A decision was made to open the dura and drain some CSF through the Sylvian fissure. After careful retraction, the wall of the cavernous sinus was coagulated and opened. Initially, the meningeal dural layer was opened followed by endosteal layer. The lesion was identified immediately underneath, and we performed internal debulking while the capsule was rolled over and removed after gentle dissection circumferentially. The abducens nerve was not visualized during resection. Pathology confirmed tumor to be a schwannoma WHO Grade I [Video 1].

She had a stable postoperative course, with immediate improvement of the left facial paresthesia. Left sixth cranial nerve palsy was gradually improving at 3 months follow-up. No postoperative MRI is performed yet.

DISCUSSION

Cavernous sinus neoplasms are a very uncommon entity. They originate from structures within the sinus. In addition, they invade the sinus from surrounding or distant sources. Most encountered lesions in this area are meningiomas, followed by pituitary adenomas, trigeminal schwannomas, chordoma, chondrosarcoma, and angiofibroma. Others include epidermoid, plasmacytoma, hemangioma, and schwannomas arising from other nerves.[26] Metastatic lesions from pulmonary or renal origins have also been reported. However, majority of tumors in cavernous sinus are benign[19] Intracranial schwannomas have a predilection for the posterior fossa, mainly cerebellopontine angle, or preptontine cistern, where cranial nerves course. Most affected nerve is vestibulocochlear nerve (particularly, the vestibular division),[19,27] followed by trigeminal,[19,24] oculomotor, trochlear, and abducens nerves. When a schwannoma arises from abducens nerve, cavernous segment are more likely to be involved.

In 1981, Bing-Huan described one of the earliest cases of an abducens nerve schwannoma, involving cerebellopontine segment.[2] A year later, Leunda et al. reported the first case of cavernous sinus schwannoma of the sixth cranial nerve.[18] Since then, only 16 cases of abducens schwannomas arising from cavernous segment have been reported [Table 1]. Cranial schwannomas originate in transitional segments where Schwann cells take over from oligodendroglia, to myelinate cranial nerves. This segment is not consistent in terms of anatomical location, across all cranial nerves. In the case of abducens nerve, rest of cranial nerves innervating ocular muscles schwannomas do not arise at the previously mentioned zones, which for these nerves occur 1–4 mm from brainstem.[19] Given the limited number of cases, data on natural history or rate of growth of abducens schwannomas in cavernous sinus are not well documented. Unfortunately, our patient reached neurosurgical care, symptomatic, and required surgical intervention, without a period of preoperative follow-up.

Patients with cavernous sinus pathology present with wide array of signs and symptoms related to critical neural and vascular structures in this anatomical region. Our patient presented with left sixth cranial nerve palsy and left facial parasthesia in the distribution of ophthalmic and maxillary divisions of trigeminal nerve. We attributed these symptoms to the presence of a schwannoma arising from sixth cranial nerve and causing mass effect on two divisions of the trigeminal nerve within the narrow space of the cavernous sinus. By this, postoperative left facial paresthesia had subsided, while the sixth cranial nerve recovery took a slower course. It is important to conduct a careful preoperative physical examination looking for the third, fourth, fifth, and sixth cranial nerves palsies. In addition, looking for the manifestations of Horner’s syndrome (due to involvement of internal carotid sympathetic plexus)[33] or stigmata of neurofibromatosis, which our patient did not have. Based on the symptomatology, Tung et al. categorized these lesions into two types. Type 1 originated from cavernous

Figure 1: (a-d) Preoperative magnetic resonance imaging.

Magnetic resonance imaging: Heterogeneous T2 hyperintense lobulated mass at the left cavernous sinus extending into Meckel’s cave with bony remodeling, also with extension into left preptontine cistern, with avid contrast enhancement with no imaging evidence of intratumoral cystic degeneration or dural tail.
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![Image: A page from a document with text and a table]

Table 1: Review of literature of sixth cranial nerve schwannomas in cavernous sinus.

| Author            | Age/gender | Deficit                        | Location                              | Approach         | Degree of resection | Recovery                        |
|-------------------|------------|--------------------------------|---------------------------------------|------------------|---------------------|---------------------------------|
| Sun et al.,[30] 2017 | 18, F      | Abducens palsy Partial oculomotor palsy | Cavernous sinus proper Cavernous sinus proper | Orbital zygomatic | GTR                  | Recovered                       |
| Chowdhury et al.,[3] 2014 | 38, F      | Abducens palsy                  | Cavernous sinus proper Cavernous sinus proper | Subtemporal Extrudal | GTR                  | Persistent                      |
| Chowdhury et al.,[3] 2014 | 30, M      | Abducens paresis                | Cavernous sinus proper Cavernous sinus proper | Frontotemporal   | STR                  | Not documented                  |
| Moses et al.,[21] 2011 | 65, M      | Abducens palsy Mandibular division of trigeminal nerve hypoesthesia. Abducens palsy Mandibular division of trigeminal nerve paresthesia | Cavernous sinus proper Zygomatic anterior transpetrosal |                  |                     |                                 |
| Nakagawa et al.,[22] 2004 | 36, F      | Abducens palsy Mandibular division of trigeminal nerve paresthesia |                      | GTR              |                     | Abducens palsy recovered; Trigeminal involvement improved Persistent |
| Mascarenhas et al.,[30] 2004 | 39, F      | Abducens palsy                  | Cavernous sinus proper Cavernous sinus proper | Orbitozygomatic intrudal | GTR                  | Persistent                      |
| Lo and Besser,[19] 2001 | 19, M      | Abducens palsy                  | Cavernous sinus proper Cavernous sinus proper | Subtemporal intrudal Frontotemporal | STR                  | Persistent                      |
| Lanotte et al.,[17] 1992 | 62, M      | Oculomotor palsy Abducens palsy Ophthalmic and maxillary division of trigeminal nerve paresthesia | Caverous sinus proper | GTR              |                     | Persistent                      |
| Tung et al.,[32] 1991 | 45, F      | Abducens palsy                  | Cavernous sinus proper | Frontotemporal   | STR                  | Persistent                      |
| Hansman et al.,[14] 1986 | 58, M      | Abducens palsy                  | Cavernous sinus proper | Not documented   | Not documented       | Not documented                  |
| Wang et al.,[35] 2015 | 68, M      | No deficit.                     | Cisternocavernous Type 2 Cervous sinus proper | Retrosigmoid     | STR                  | Abducens paresis                |
| Shibao et al.,[29] 2014 | 36, F      | Sensorineural hearing impairment Ophthalamic and maxillary division of trigeminal nerve paresthesia | Cisternocavernous Type 2 | Anterior transpetrosal | STR                  | Improved hearing Abducens palsy |
| Acharya et al.,[1] 2003 | 40, F      | Abducens palsy                  | Cisternocavernous Type 1 | Subtemporal     | STR                  | Persistent                      |
| Tung et al.,[32] 1991 | 35, M      | Abducens palsy                  | Cisternocavernous Type 1 | Frontotemporal | GTR                  | Persistent                      |
| Ginsberg et al.,[8] 1988 | 47, F      | Nystagmus Facial nerve palsy    | Cisternocavernous Type 2 Cervous sinus proper | Not documented   | Not documented       | Not documented                  |
| Leunda et al.,[18] 1982 | 10, M      | Abducens palsy                  | Cisternocavernous Type 1 | Subtemporal     | GTR                  | Persistent                      |

GTR: Gross total resection, STR: Subtotal resection

Imaging appearance in our case was consistent with previously described radiological characteristics of schwannomas.[13] Features on MRI include heterogeneous T2 hyperintensity[6] (related to size of the lesion, the larger the lesion, the more heterogeneous it appears) and strong sinus presenting with abducens palsy and diplopia. Type 2 originated mainly from prepointine cistern before cavernous segment, presenting with signs of increased intracranial pressure and obstructive hydrocephalus with sixth nerve palsy, other cranial nerves might be involved.[32]
contrast enhancement.[11] Bony remodeling has also been described, which was noted in our case. Classically described dumbbell shape is attributed to lesion traversing anatomical corridors such as Dorello's canal or in other schwannomas, through Meckel's cave.[11] Celi et al. divided sixth cranial nerve cavernous sinus schwannomas into three types; cisternal, cavernous, and mixed (cavernous and cisternal segments of the nerve).[3] We proposed a classification in two categories. These included cavernous sinus proper (where tumor confined to borders of cavernous sinus without extension to Meckel's cave or orbit) and cisternocavernous (type one involves cavernous sinus and extends into prepontine cistern with bulk of the mass in cavernous sinus, while type two involves cavernous sinus and extends into prepontine cistern with bulk of the mass in prepontine cistern) [Table 2]. This classification helps to guide surgical approach, expected extent of resection, and feasibility of nonsurgical options.

This unique pathology requires an in-depth knowledge of the cavernous sinus and its membranous anatomy, allowing safer approaches to facilitate maximum tumor resection.[11,22,23] The pyramidal shape of the cavernous sinus plays an integral role in how different lesions within the same area cause different manifestations and are tackled differently. The first relevant anatomical characteristic of the cavernous sinus is that two dural layers form the sinus. These include an inner layer (periosteal or endosteal) and an outer membranous layer.[6,22] The membranous layer is further divided into two layers, superficial (dura propria) and deep. An overlap of these layers creates compartments in the region of the cavernous sinus, which leads us to the second relevant anatomical characteristics of the sinus – the distribution of contents of the sinus within these compartments. The sixth cranial nerve runs within the sinus proper, accompanied by the cavernous segment of the internal carotid artery and sympathetic plexus. On the other hand, the first and second divisions of the trigeminal nerve, trochlear, and oculomotor nerves follow a course within the lateral wall, separated from sinus proper by the deep membranous layer.[6,34] This places abducens nerve schwannomas in a separate category from other schwannomas in this area. The growth of abducens schwannomas might cause narrowing of the adjacent internal carotid artery, rather than only displacing it by mass effect as compared to schwannoma of other cranial nerve in lateral wall of the sinus.

Multiple factors influence the decision for surgery such as patient's age, clinical status (including chronic illnesses), ability to tolerate such surgery, and neurological condition at presentation. Indications for surgery in cavernous sinus schwannomas include presence or worsening of cranial nerve deficits high intracranial pressure caused by large lesions or those extending into prepontine cistern. The main goal of surgery is to provide maximum safe resection, while preserving critical neural structures near the tumor. Moreover, selecting the appropriate approach depends on multiple factors. One of the most important factors is the volumetric distribution of the tumor. Lesions that are confined to the cavernous sinus proper are better addressed through frontotemporal (pterional); either extradural or combined (intra, extradural), supraorbital craniotomy (subfrontal intradural), and numerous other approaches described in literature.[13] In contrast, lesions where bulk of tumor is in prepontine cistern require different approaches, while keeping in mind goal of surgery. These involve the decompression of brainstem or resolving obstructive hydrocephalus caused by mass effect, taking priority over cavernous sinus tumor debulking. In our case, we utilized the ptetional route, a combined extra and intradural approach.

Sun et al. described several complications caused by injury to critical neurovascular structures in the cavernous sinus.[10] Some complications affect the degree of resection, like venous bleeding. This hinders resolution of preoperative neurological deficits, a major aim of surgery. Venous bleeding is best avoided by following the plane of the dural layers and gentle dissection beyond the deep layer of the lateral wall. Usually, packing of bleeding from venous structures is sufficient to control it.[10] We recommend a careful manipulation of the perforator vessels near the tumor to prevent vasospasms. It also important to avoid injury to the inferolateral trunk vessels, which supplies the oculomotor and trochlear nerves within the sinus.[10] Inferolateral trunk is associated with the abducens nerve cavernous segment, it is important to identify it and preserve it during resection.[10] Other complications like Horner's syndrome precipitated by sympathetic fibers injury have been described as well.[10]

Multiple factors are related to the preservation of the sixth cranial nerve function during surgery. The abducens nerve splits into several fibers as it passes lateral to internal carotid artery within the cavernous sinus.[11] Higher chance of recovery is associated with tumors in cavernous sinus proper exclusively due to sparing of the nerve in the Dorello's canal.[22] Dual nerve trunks in the cavernous

| Table 2: Proposed classification of sixth cranial nerve schwannoma in cavernous sinus. |
|------------------------------------------|----------------------------------------------------------------------------------|
| **Category**                | **Definition**                                                                       |
| Cavernous sinus proper       | Confined to borders of cavernous sinus without extension to Meckel's cave or orbit |
| Cisternocavernous type 1     | Involves cavernous sinus and extends into prepontine cistern with bulk of the mass in cavernous sinus |
| Cisternocavernous type 2     | Involves cavernous sinus and extends into prepontine cistern with bulk of the mass in prepontine cistern |
segment of abducens nerve have been reported, which might account for postoperative preservation of function, in instances where the nerve is incised. In cases where the nerve was completely transected, reconstruction of the nerve is recommended with acceptable functionality. Radiosurgery is a good option in patients who are exposed to unacceptable risk with surgery or who prefer a less invasive treatment modality. Counseling the patient on surgical and less invasive modalities is recommended. In 1996, Chakrabarti et al. documented the first recorded treatment of abducens schwannoma by stereotactic radiosurgery. This patient suffered from a recurrent sixth cranial nerve schwannoma following surgical resection. On follow-up, radiological response was evident by a necrotic center on MRI accompanied by an improved neurological condition. Further reposts followed similar findings. However, there is still limited number of patients and the lack of data on natural history makes it difficult to compare it against different treatment modalities.

CONCLUSION
Abducens nerve schwannoma of the cavernous sinus is a rare and challenging tumor. However, it is amenable to surgical intervention with favorable neurological outcomes.

Declaration of patient consent
Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest
There are no conflicts of interest.

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