



Case Report

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

Zeyad M. Alhussain, Shatha K. Alharbi, Faisal Farrash

Department of Neuroscience, Division of Neurological Surgery, King Faisal Specialist Hospital and Research Center, Riyadh, Saudi Arabia.

E-mail: *Zeyad M. Alhussain - zeyad.alhussain@gmail.com; Shatha K. Alharbi - shatha.k.alharbi@gmail.com; Faisal Farrash - dr.farrash@gmail.com



***Corresponding author:**

Zeyad M. Alhussain,
Department of Neuroscience,
Division of Neurological
Surgery, King Faisal Specialist
Hospital and Research Center,
Riyadh, Saudi Arabia.

zeyad.alhussain@gmail.com

Received : 19 June 2020

Accepted : 30 September 2020

Published : 25 November 2020

DOI

10.25259/SNI_362_2020

Videos available on:

www.surgicalneurologyint.com

Quick Response Code:



ABSTRACT

Background: Schwannomas of the abducens nerve are a rare pathology and are encountered less within the cavernous sinus. We describe a case of sixth cranial nerve schwannoma, in the cavernous sinus.

Case Description: A 50-year-old lady, presented with 2 years history of double vision and left facial numbness that started 6 months before presentation, found to have hyperintense lobulated mass at the left cavernous sinus extending into Meckel's cave with bony remodeling on magnetic resonance imaging. She underwent left frontotemporal craniotomy, combined extra-intradural approach, gross total resection. She had a gradual recovery of the sixth cranial nerve function.

Conclusion: Abducens nerve schwannoma of the cavernous sinus is a rare and challenging tumor. However amenable to surgical intervention with favorable neurological outcome.

Keywords: Abducens nerve, Cavernous sinus, Classification, Neurilemoma, Neurinoma, Neuroma, Schwannoma

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

This is an open-access article distributed under the terms of the Creative Commons Attribution-Non Commercial-Share Alike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

©2020 Published by Scientific Scholar on behalf of Surgical Neurology International

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 prepontine 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,

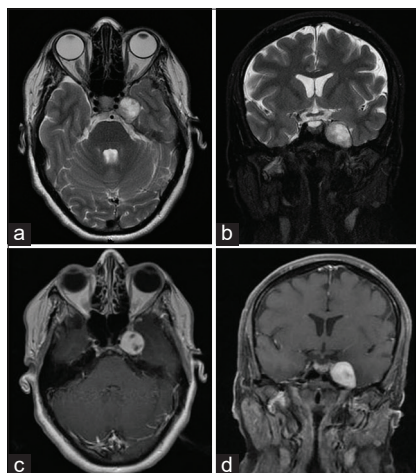


Figure 1: (a-d) Preoperative magnetic resonance imaging.

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 prepontine cistern, with avid contrast enhancement with no imaging evidence of intratumoral cystic degeneration or dural tail.

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.^[24] Metastatic lesions from pulmonary or renal origins have also been reported. However, majority of tumors in cavernous sinus are benign.^[7] Intracranial schwannomas have a predilection for the posterior fossa, mainly cerebellopontine angle, or prepontine 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.^[3] 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 paresthesia 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

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

Author	Age/ gender	Deficit	Location	Approach	Degree of resection	Recovery
Sun <i>et al.</i> , ^[30] 2017	18, F	Abducens palsy Partial oculomotor palsy	Cavernous sinus proper	Orbital zygomatic	GTR	Recovered
Chowdhury <i>et al.</i> , ^[5] 2014	38, F	Abducens palsy	Cavernous sinus proper	Subtemporal Extradural	GTR	Persistent
Chowdhury <i>et al.</i> , ^[5] 2014	30, M	Abducens paresis	Cavernous sinus proper	Subtemporal Extradural	GTR	Worsened
Moses <i>et al.</i> , ^[21] 2011	65, M	Abducens palsy Mandibular division of trigeminal nerve hypoesthesia.	Cavernous sinus proper	Frontotemporal	STR	Not documented
Nakagawa <i>et al.</i> , ^[22] 2004	36, F	Abducens palsy Mandibular division of trigeminal nerve paresthesia	Cavernous sinus proper	Zygomatic anterior transpetrosal	GTR	Abducens palsy recovered; Trigeminal involvement improved Persistent
Mascarenhas <i>et al.</i> , ^[20] 2004	39, F	Abducens palsy	Cavernous sinus proper	Orbitozygomatic intradural	GTR	Persistent
Lo and Besser, ^[19] 2001	19, M	Abducens palsy	Cavernous sinus proper	Subtemporal intradural	STR	Persistent
Lanotte <i>et al.</i> , ^[17] 1992	62, M	Oculomotor palsy Abducens palsy Ophthalmic and maxillary division of trigeminal nerve paresthesia	Cavernous sinus proper	Frontotemporal	GTR	Persistent
Tung <i>et al.</i> , ^[32] 1991	45, F	Abducens palsy	Cavernous sinus proper	Frontotemporal	STR	Persistent
Hansman <i>et al.</i> , ^[10] 1986	58, M	Abducens palsy	Cavernous sinus proper	Not documented	Not documented	Not documented
Wang <i>et al.</i> , ^[35] 2015	68, M	No deficit.	Cisternocavernous Type 2	Retrosigmoid	STR	Abducens paresis
Shibao <i>et al.</i> , ^[29] 2014	36, F	Sensorineural hearing impairment Ophthalmic and maxillary division of trigeminal nerve paresthesia	Cisternocavernous Type 2	Anterior transpetrosal	STR	Improved hearing Abducens palsy
Acharya <i>et al.</i> , ^[11] 2003	40, F	Abducens palsy	Cisternocavernous Type 1	Subtemporal	STR	Persistent
Tung <i>et al.</i> , ^[32] 1991	35, M	Abducens palsy	Cisternocavernous Type 1	Frontotemporal	GTR	Persistent
Ginsberg <i>et al.</i> , ^[8] 1988	47, F	Nystagmus Facial nerve palsy	Cisternocavernous Type 2	Not documented	Not documented	Not documented
Leunda <i>et al.</i> , ^[18] 1982	10, M	Abducens palsy	Cisternocavernous Type 1	Subtemporal	GTR	Persistent

GTR: Gross total resection, STR: Subtotal resection

sinus presenting with abducens palsy and diplopia. Type 2 originated mainly from prepontine 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]

Imaging appearance in our case was consistent with previously described radiological characteristics of schwannomas.^[15] 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

contrast enhancement.^[31] 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.^[31] Celli *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 pterional route, a combined extra and intradural approach.

Sun *et al.* described several complications caused by injury to critical neurovascular structures in the cavernous sinus.^[30] 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.^[30] 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.^[16] Inferolateral trunk is associated with the abducens nerve cavernous segment, it is important to identify it and preserve it during resection.^[16] Other complications like Horner’s syndrome precipitated by sympathetic fibers injury have been described as well.^[30]

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.^[9,36,37] In cases where the nerve was completely transected, reconstruction of the nerve is recommended with acceptable functionality.^[28] 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.^[12,25,26] 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.^[4] Further reposts followed similar findings.^[14,26] 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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

- Acharya R, Husain S, Chhabra SS, Patir R, Bhalla S, Seghal AD. Sixth nerve schwannoma: A case report with literature review. *Neurol Sci* 2003;24:74-9.
- Bing-Huan C. Neurinoma of the abducens nerve. *Neurosurgery* 1981;9:64-6.
- Celli P FL, Acqui M, Mastronardi L, Fortuna A, Palma L. Neurinoma of the third, fourth, and sixth cranial nerves: A survey and report of a new fourth nerve case. *Surg Neurol* 1992;38:216-24.
- Chakrabarti KB, Doughty D, Plowman PN. Stereotactic multiple arc radiotherapy. II-cranial neuroma. *Br J Neurosurg* 1996;10:577-83.
- Chowdhury FH, Haque RM, Goel AH. Abducent nerve schwannoma: A report of 3 cases. *Neurosurg Q* 2014;24:40-3.
- El-Kalliny M, van Loveren H, Keller JT, Tew JM Jr. Tumors of the lateral wall of the cavernous sinus. *J Neurosurg* 1992;77:508-14.
- Feng M, Xu D, Wang G, Wang F, Guo F. Clinicopathological features and microsurgical management of lesions located in the cavernous sinus: Analysis of 66 cases. *Turk Neurosurg* 2020;30:171-7.
- Ginsberg F, Peyster RG, Rose WS, Drapkin AJ. Sixth nerve schwannoma: MR and CT demonstration. *J Comput Assist Tomogr* 1988;12:482-4.
- Haładaj R, Skrzat SJ. Bilateral duplication of the abducens nerve-case study. *Folia Med Cracov* 2019;59:13-20.
- Hansman ML, Hoover ED, Peyster RG. Sixth nerve neuroma in the cavernous sinus: CT features. *J Comput Assist Tomogr* 1986;10:1030-2.
- Harris FS, Rhoton AL. Anatomy of the cavernous sinus. A microsurgical study. *J Neurosurg* 1976;45:169-80.
- Hayashi M, Chernov M, Tamura N, Yomo S, Ochiai T, Nagai M, *et al.* Gamma knife surgery for abducent nerve schwannoma. Report of 4 cases. *J Neurosurg* 2010;113:136-43.
- Inoue T, Rhoton AL, Theele D, Barry ME. Surgical approaches to the cavernous sinus: A microsurgical study. *Neurosurgery* 1990;26:903-32.
- Kim IY, Kondziolka D, Niranjana A, Flickinger JC, Lunsford LD. Gamma knife surgery for schwannomas originating from cranial nerves III, IV, and VI. *J Neurosurg* 2008;109:149-53.
- Korchi AM, Cuvinciuc V, Caetano J, Becker M, Lovblad KO, Vargas MI. Imaging of the cavernous sinus lesions. *Diagn Interv Imaging* 2014;95:849-59.
- Krisht A, Barnett DW, Barrow DL, Bonner G. The blood supply of the intracavernous cranial nerves: An anatomic study. *Neurosurgery* 1994;34:275-79; discussion 279.
- Lanotte M, Giordana MT, Forni C, Pagni CA. Schwannoma of the cavernous sinus. Case report and review of the literature. *J Neurosurg Sci* 1992;36:233-8.
- Leunda G, Vaquero J, Cabezudo J, Garcia-Uria J, Bravo G. Schwannoma of the oculomotor nerves. Report of four cases. *J Neurosurg* 1982;57:563-5.
- Lo PA, Harper CG, Besser M. Intracavernous schwannoma of the abducens nerve: A review of the clinical features, radiology and pathology of an unusual case. *J Clin Neurosci* 2001;8:357-60.
- Mascarenhas L, Magalhaes Z, Honavar M, Romao H, Resende M, Pereira JR, *et al.* Schwannoma of the abducens nerve in the cavernous sinus. *Acta Neurochir (Wien)* 2004;146:389-92.
- Moses JE, Vermani N, Bansal SK. Preoperative clinicoradiological diagnosis of schwannoma arising from cavernous segment of abducens nerve. *Neurol India* 2011;59:471-3.
- Nakagawa T, Uchida K, Ozveren MF, Kawase T. Abducens schwannoma inside the cavernous sinus proper: Case report. *Surg Neurol* 2004;61:559-63.
- Ozveren MF, Uchida K, Aiso S, Kawase T. Meningovenous structures of the petroclival region: Clinical importance for surgery and intravascular surgery. *Neurosurgery* 2002;50:829-37.
- Pamir MN, Kilic T, Ozek MM, Ozduman K, Ture U. Non-meningeal tumours of the cavernous sinus: A surgical analysis. *J Clin Neurosci* 2006;13:626-35.
- Peciu-Florianu I, Tuleasca C, Comps JN, Schiappacasse L, Zeverino M, Daniel RT, *et al.* Radiosurgery in trochlear and

- abducens nerve schwannomas: Case series and systematic review. *Acta Neurochir (Wien)* 2017;159:2409-18.
26. Prasad GL, Sharma MS, Kale SS, Agrawal D, Singh M, Sharma BS. Gamma knife radiosurgery in the treatment of abducens nerve schwannomas: A retrospective study. *J Neurosurg* 2016;125:832-7.
 27. Russell DS, Rubinstein LJ. *Pathology of Tumors of the Nervous System*. 7th ed. London: Edward Arnold; 1989.
 28. Sekhar LN, Lanzino G, Sen CN, Pomonis S. Reconstruction of the third through sixth cranial nerves during cavernous sinus surgery. *J Neurosurg* 1992;76:935-43.
 29. Shibao S HS, Yoshida K. Dumbbell-shaped abducens schwannoma: Case report. *Neurol Med Chir (Tokyo)* 2014;54:331-6.
 30. Sun H, Sharma K, Kalakoti P, Thakur JD, Patra DP, Konar S, *et al.* Factors associated with abducens nerve recovery in patients undergoing surgical resection of sixth nerve schwannoma: A systematic review and case illustration. *World Neurosurg* 2017;104:883-99.
 31. Tang Y, Booth T, Steward M, Solbach T, Wilhelm T. The imaging of conditions affecting the cavernous sinus. *Clin Radiol* 2010;65:937-45.
 32. Tung H, Chen T, Weiss MH. Sixth nerve schwannomas. Report of two cases. *J Neurosurg* 1991;75:638-41.
 33. Türe U, Seker A, Kurtkaya O, Pamir MN. Internal carotid plexus schwannoma of the cavernous sinus: Case report. *Neurosurgery* 2003;52:435-9.
 34. Umansky F. *The Cavernous Sinus. An Anatomical Study of Its Lateral Wall*. Vienna: Springer; 1987.
 35. Wang M, Huang H, Zhou Y. Abducens nerve schwannoma in cerebellopontine angle mimicking acoustic neuroma. *J Craniofac Surg* 2015;26:589-92.
 36. Wysiadecki G, Orkisz S, Gałązkiewicz-Stolarczyk M, Brzeziński P, Polgaj M, Topol M. The abducens nerve: Its topography and anatomical variations in intracranial course with clinical commentary. *Folia Morphol (Warsz)* 2015;74:236-44.
 37. Wysiadecki G, Polgaj M, Topol M. An unusual variant of the abducens nerve duplication with two nerve trunks merging within the orbit: A case report with comments on developmental background. *Surg Radiol Anat* 2016;38:625-9.

How to cite this article: Alhussain ZM, Alharbi SK, Farrash F. Abducens nerve schwannoma of the cavernous sinus: A case report and literature review. *Surg Neurol Int* 2020;11:402.