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TRIJEMINAL SCHWANNOMA OF THE CAVEMOUS SINUS. CASE REPORT

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ABSTRACT

A case of cavernous sinus shwannoma is presented. A 31-years-old female presented with a mass causing proptosis, oculomotor nerve paresis and retro-orbital headaches. Diagnosis was confirmed by radiological imaging. The patient was treated with surgery. The clinicoradiological features and the possible therapeutic options are discussed.

Keyword: Shwannoma', Skull base; Cavernous sinus; Surgery

1. INTRODUCTION

Trigeminal schwannoma is a benign tumor, first described by Smith in 1938 [1]. It is the most common amont nonvestibular schwannomas and account for 1 to 8% of intracranial tumors.[2.3] Incidence of trigeminal schwannoma is more in the middle aged population with peak incidence in the âge group 38^-0 years, commonly seen in women.[6]

Schwannomas are slow growing nerve sheath tumors. When originating in the CS, they most commonly arise from cranial nerves III, VI or V2[4]

Imaging plays an essential rôle in the évaluation of suspected CS pathology, both for lésion characterization and délinéation of extent and for planning the surgical approach [7]. MRI with and without contrast is the imaging modality of choice for a suspected cavemous sinus mass. [5]

The treatment of choice is complété resection of the tumour. Management of trigeminal schwannomas include - observation, microsurgical resection, radiosurgery, fractionated conformai radiotherapy or a combination of the above. Management strategy dépends upon the case history and clinical présentation and varies from case to case [8].

2. OBSERVATION

Mme B.A 31 years old , with no comorbidities,

Having developed irreducible, non-pulsatile, right-sided axial proptosis that has been progressing for 6 months, associated with retro-ocular pain and latéral gaze palsy due to involvement of the VI nerve which regressed with anti-edema treatment. Associated with hpoesthesia in the V1 territory with intact pupillary light reflex, and preserved visual acuity.

MRI scann shows a tissue tumor projecting from the right cavernous sinus, maesuring 26x19.5 mm axially and extending

22.5 mm in height, roughly oval and well-defîned, appearing isointense on Tl and FLAIR images, and heterogeneous iso intense on T2 images due to the presence of a few hyperintense areas, without corresponding enhacement on diffusion- weighted imaging. This tumor présents the following relationships :fîlls the superior orbital fissure and compressess the right optic nerve, Displaces and compresses the ICA in its cavernous option, partially fills the right Meckel's cave. And finally reaches contact with the temporal cérébral parenchyma.

Surgical intervention was performed via a left subtemporal approach through a frontopterional bone flap, followed by zigomatic bone removal and drilling of the outer third of the lesser sphenoid bone.

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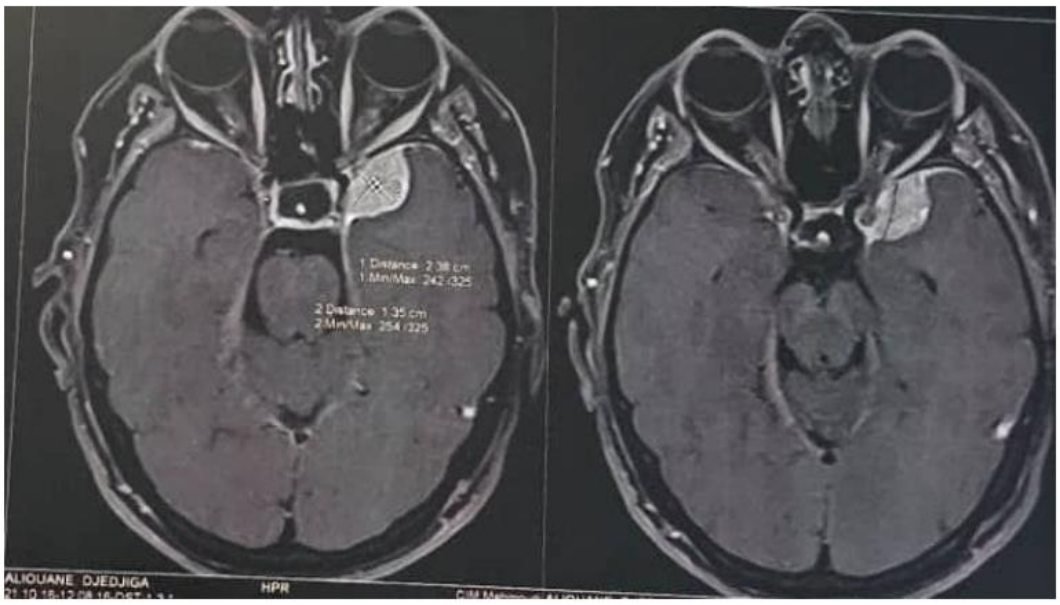


Figure. 1. Preoperative T1-weighted, gadolinium-enhanced MRI (axial section)

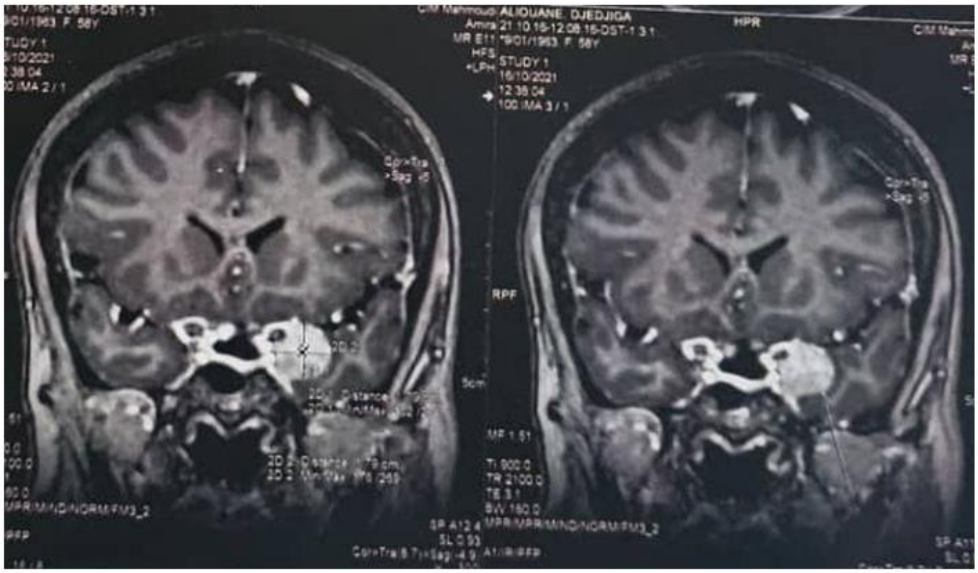


Figure. 2. Frontal T2-weighted MR image.

3. DISCUSSION

Trigeminal nerve is the largest cranial nerve. It is a mixed sensory-motor nerve receiving sensory input from the face and providing motor supply to the muscles of mastication.[8.9] The nerve can be divided into

five segments: intra-axial (brain stem), cistemal, Meckel's cave and cavemous sinus, skull base and extra cranial segments. The Gasserian or semilunar ganglion is located in the inferior aspect of Meckel's cave and gives off three branches: ophthalmic (VI), maxillary (V2) and mandibular (V3).[8.10] Trigeminal schwannomas are rare tumors, accounting for 1-2% of ail intracranial schwannomas and tend

to occur predominantly in the 4th-6th décades, slightly more common in females.[] These are benign tumors and hâve a slow insidious course.

Trigeminal neuralgia is a spécifie painful syndrome defined by the International Association as a "sudden, usually unilatéral, severe, brief, stabbing, récurrent pain in the distribution

of one or more branches of the trigeminal

nerve". Other common clinical symptoms are numbness

or buming sensation along the distribution of the nerve or one of its branches. Longstanding tumors may also présent with motor symptoms like difficulty in chewing and déviation of the jaw.[10] If the tumour is involving trigeminal nerve in the cavernous sinus it can lead to dysfunction of cranial nerves III, IV and VI as in our case ,whereas enlargement in prepontine cistem may lead to compressive effects on cranial nerves VII, VIII and IX. [11.13]

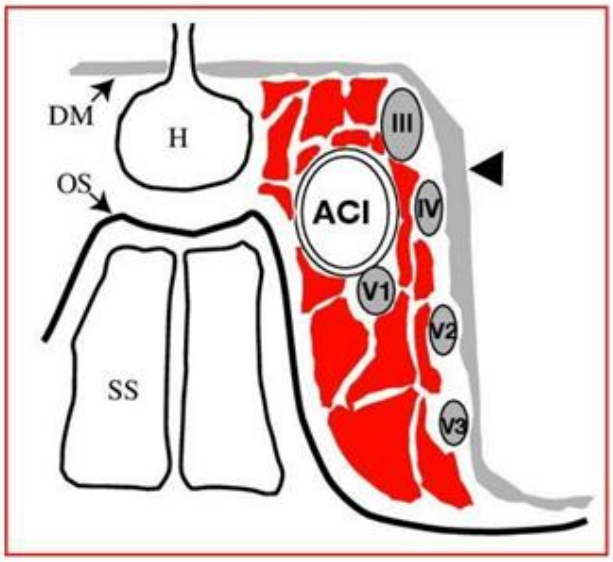


Figure 3 .cavernous sinus.

located at the base of the skull, on either side of the sella turcica, playing a crucial rôle in cérébral venous drainage. It is traversed by the internai carotid artery and cranial nerves III, IV, V1, V2, and V.

Preoperative neuroradiological évaluation is of utmost importance for establishing a diagnosis and deciding on the surgical approach for trigeminal schwannoma. MRI is the imaging modality of choice and is usually diagnostic in the appropriate clinical setting.

Tumors usually appear as isointense or hypointense on Tl- weighted images, hyperintense on T2-weighted images with avid enhancement after contrast injection.

Sometimes the lésion may be mixed solid-cystic or predominantly cystic on imaging. In addition to routine MRI sequences, it is important to acquire thin

T2-weighted CISS 3D axial sequence in patients with clinical suspicion of a trigeminal nerve lésion for better évaluation of the cistemal segment of the nerve. CT scan is supplementary to the MRI imaging, particularly for tumors located in the skull

base. On CT scan, they usually appear as uniformly enhancing masses with remodeling of the adjacent bone.[12]

The differential diagnosis includes meningiomas, epidermoid cysts, metastasis, chondrosarcomas, chordomas, chondromas, vestibular schwannomas, and maxillary sinus tumour s.

Jefferson[14] divided trigeminal tumours into 4 groups based on their anatomical location into posterior fossa (root

type), combined posterior fossa-middle fossa (dumbbell type), middle fossa (ganglion type), and peripheral (division type). Samii et al. [15] classifîed it into 4 categories based on radiological fîndings into the following - Type A, intracranial tumour predominantly in the middle fossa; Type B, intracranial tumour predominantly in the posterior fossa; Type C, intracranial dumbbell-shaped tumour in the middle and posterior fossa; and Type D, extracranial tumour with intracranial extensions. Ramina et al. [16] modifîed the clas-sification given by Samii et al.: Type A, predominantly an extracranial tumour with small extension into the middle fossa; Type B, an intracranial tumour predominantly in the middle fossa with extracranial extension; Type C, a tumour in the middle fossa; Type D, a tumour in the posterior fossa; Type E, a tumour with middle and posterior fossa extensions; and Type F, a tumour with extracranial, middle, and posterior fossa extensions.

Tumour types are ordered according to the level of difficulty as A, C, D, B, E, and F (Fig.2). According

to this modified classification, Type A tumours hâve the lowest level of difficulty and Type F tumours, the highest.

The surgical approaches commonly used, based on the aforementioned classification are as follows: Type A tumours

- extradural approach, Type B tumours - intradural middle fossa approach, Type C tumours - middle fossa approach, Type D tumours - retrosigmoid approach, Type E tumours - presigmoid approach and type F tumours usually require a combination of approaches [16].

A more recent classification into six different types has been proposed.

Type P: posterior fossa tumor in the subdural space

Type M: middle fossa tumor in the interdural space

Type E: extracranial tumor in the épidural space

E 1 : tumor in the orbit

E2: tumor in the pterygopalatine and inffatemporal fossae

Type MP: dumbbell-shaped tumor in the middle and posterior fossae

Type ME: dumbbell-shaped tumor in the middle fossa and extracranial space

Type MPE: tumor involving the posterior fossa, middle fossa, and extracranial space[19]

Chen et al noted 62% tumor s in the middle fossa or both the middle fossa and the posterior fossa.Goel reported 81% tumors occupying the middle fossa and the posterior fossa. Wanibuchi reported 66% tumors occupying the middle fossa and the posterior fossa. The trigeminal schwannomas usually displace the neighboring neural and vascular structures, as opposed to the invasion of the venous spaces of the cavemous sinus or encasement of the cavemous sinus portion of the internai carotid artery (ICA) [20]

Management strategy for trigeminal schwannomas includes the following [17].

Clinical observation followed by MRI follow up for incidental tumours.

Surgical removal.

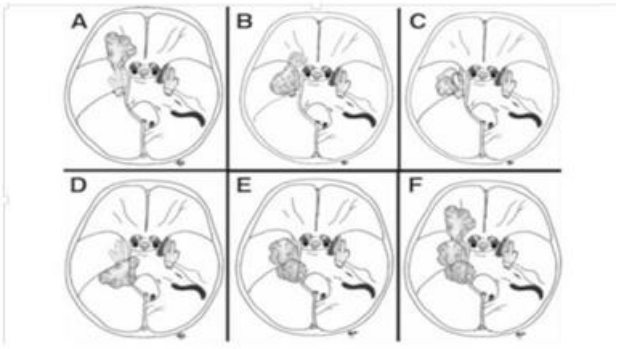
Radiotherapy.

Radiosurgery.

Skull base approaches and microsurgical dissections help in complété or near total surgical removal in more than 70% patients. Cavemous sinus involvement is one of the reasons for incomplète tumour removal. Trigeminal hypoesthesia is the most frequent presenting symptom post-surgery. Diplopia, CSF leak, meningitis and hydrocephalus are the possible complications. Radiosurgery is reserved for small, non resectable and residual tumours within the cavemous sinus.

However no patient will be cured with this benign tumour with radiotherapy or radiosurgery [18].

Figure4 : Classification of trigeminal schwannomas - Ramina et al.



Surgical Results:

In cases with marked expansion of Meckel cave and a dominant middle fossa component, a purely anterior approach becomes a viable option via either a pterional or FTOZ (frontotemporal-orbitozygomatic) craniotomy utilizing an extradural dissection.

In this situation, the expanded Meckel cave becomes the corridor by which the posterior fossa component is

resected. For tumors extending more anteriorly (as in our case)

in the middle fossa and more inferiorly in the infratemporal fossa, a zygomatic osteotomy can be added to the subtemporal approach and partial resection of the middle fossa floor can expose most of

the inferior extent of trigeminal schwannoma.

Dedicated infratemporal fossa approaches such as described by Fisher[21] are rarely, if ever, needed in the resection of trigeminal schwannomas. Not to be forgotten that these are benign tumors in which a subtotal resection and postoperative radiosurgery is

often a very reasonable strategy.

Outcome from surgical resection has been reported in several sériés, and although some authors hâve reported improvement in preoperative hypoesthesia[22.23.24] in most cases, the presenting symptoms persisted or worsened after surgical

resection. Goel et al [23] reported improvement in the preoperative trigeminal sensory déficit in 40% of the patients but

worsening in 27%. Wanibuchi et al [24] noted improvement in 16%, no change in 73%, worsening in 12%, and new hypesthesia in 22% of patients. Chen et al reported unchanged facial hypesthesia in 72% and improved in 28%. Other frequent symptoms include those related to involvement of adjacent cranial nerves in the cavemous sinus. [22.23.24] Wanibuchi et al reported diplopia in 20% (86% due to déficit of the abducens nerve and 14% due to déficit of the oculomotor nerve). Al- Mefty et al 15 noted diplopia in 52% (40% due to sixth nerve déficit). Chen at al reported diplopia in 18% of patients (80% due to abducens nerve déficit and 20% due to oculomotor

nerve). The diplopia improved in 70% of patients postoperatively.

In a recent study of 20 patients, Samii et al reported improvement in facial pain in ail patients and cerebellar ataxia in three of four patients. [25]

Jeong et al noted improved or unchanged trigeminal symptoms in 51% but worsening of facial hypesthesia in most of the cases. Fukaya et al studied

57 patients who had surgical resection for trigeminal schwannoma. While 42 of 45 patients (93%) who had skull base surgery achieved a complété tumor resection, 39 of 57 patients (68%) developed complications that included cranial neuropathies, brain contusion, or CSF leakage. Sharma et al evaluated 68 patents with trigeminal schwannomas who had

surgery. [26] These authors reported 76% rate of complété tumor resection, 2% mortality, and 15% permanent morbidity.

Tablel Trigeminal schwannoma: surgical sériés

Author	Year	Tumor classification and No. of cases					Radical removal (%)	Mortality (%)	Morbidity (%)
		A	B	C	D	Total			
McCormick et al[9]	1988	6	5	2	1	14	6 (43%)	0	78
Pollack et al[20]	1989	6	4	5	1	16	12(75%)	0	6
Dotent[21]	1994	NA	NA	NA	NA	40	40(100%)	0	25
Konovalov et al[22]	1996	42	26	30	13	111	86 (77%)	3	87
Yoshida and Kawase[5]	1999	4	5	10	8	27	20 (74%)	0	74
Coel et al[7]	2003	29	7	30	7	73	51 (70%)	3	7
Pamir et al[1]	2007	5	2	9	2	18	17(94%)	0	28
Wanibuchi et al[8]	2012	39	22	33	14	105	86 (82%)	0	9
Chen et al[6]	2014	13	10	21	11	55	52 (95%)	0	5
Fukaya et al[7]	2010	15	12	26	4	57	46 (81%)	2	68
Samii et al[4]	2014	8	1	8	4	20	15(75%)	0	4
Jeong et al[6]	2014	20	20	9	0	49	47 (95.9%)	0	18

Figure 5 : Pterional approach

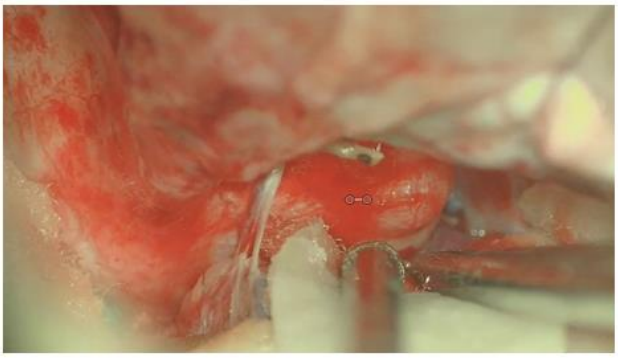


Figure 7 :Schwannoma of the cavemous sinus

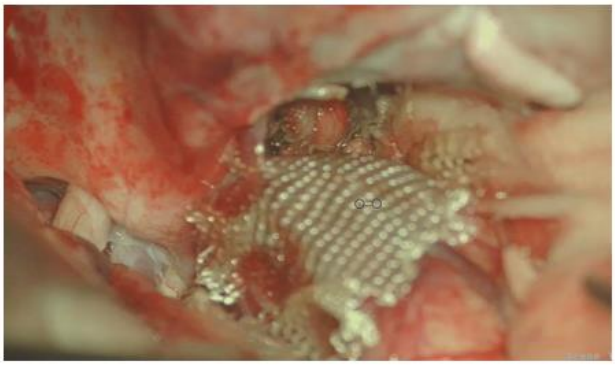


Figure 8 :operating cavity after tumor resection

3. CONCLUSION :

Improvement in neurologie symptoms, préservation of cranial nerve fonction, and control of mass effect are the primary goals of management for trigeminal schwannomas. An individualized management strategy by giving the patient the best chance of achieving fois goal. Where appropriate, a complété surgical resection is the treatment of choice, but fois may not be possible in ail cases. Radiosurgery is an option as primary management for small- to moderate-sized tumors and can be used for postoperative residuals or récurrences. For larger trigeminal schwannomas, initial surgical resection followed by SRS for residual tumor is an effective option, is associated with fewer cranial neuropathies, and greatly improves the patient's overall outcome.

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