

The Real Story of the Fifth Nerve Neurinoma: A Review of Our Learning Curve about Surgical Approaches and Associated Shortcomings

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Abstract

Background and Importance: Our learning curve during two decades of neurosurgical practice has been changed from an abstractive to a more meaningful and conductive state. In cases of fifth nerve neurinoma, pre-operative anatomopathologic diagnosis could lead us to a pre-planned program during and after the surgery to get the safe surgical result.

Case Presentation: Representation of two complex fifth nerve neurinoma cases, untoward happenings and the way to manage patient safety. Review of literature to find a wise approach for maximum benefit is included here.

Conclusion: Both of our patients have developed iatrogenic unilateral corneal anesthesia, one of them warned of it and the second one not. We have had more problems in the way of preservation of the cornea in the warned case. We have reviewed the factors influencing safe corneal preservation after the operation of fifth nerve neurinoma which are included following items: surgical approach, Anatomopathologic location of the tumor (pre-ganglionic, ganglionic or post-ganglionic), simultaneous damage of V and VII nerve including vidian nerve, preserved corneal sensation, any combination of injury to physiologic and mechanical protectors. The cornerstones to have a safe cornea following such surgeries are pre-operative exam of fifth and 7th nerve in all aspects and also early post-operative evaluation of them including the state of the tear secretion. We encounter corneal anesthesia and epithelial defect. Iatrogenic damage of vidian nerve depending to approach selected seems to be considerable. Simultaneous damage of V and VII nerve during the surgery of large neurinoma are expected and noteworthy.

Keywords: Corneal anesthesia; Iatrogenic; Combined approach; Cerebello Pontine Angle tumor; Vidian nerve; Painless corneal perforation; Herpes ophthalmicus; Neurinoma; Interdurall location

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Background and Importance

The clinical presentation of the fifth nerve neurinoma includes a wide range from trigeminal neuralgia to generalized neurologic manifestations. The main presenting signs and symptoms detected in large neurinomas or neurinomas associated with large cystic components are divided into focal and generalized manifestations (1). Focal presentations of these neurinomas include unilateral craniofacial headache in the fifth nerve territory, classic unilateral trigeminal

neuralgia (V1,V2,V3) or any combination of them. Decreased corneal sensation, asymmetric jaw jerk accompanying cranial nerve defects such as hemifacial spasm, unilateral 6th nerve paresis are some of the other focal manifestations of the fifth nerve neurinoma (2). The Generalized symptoms are intracranial pressure (ICP) raising which can be presented with headache or vomiting, Papilledema, binocular diplopia, unsteadiness and drowsiness due to hydrocephalus. Long-Tract signs as spastic hemiparesis or quadriplegia and also

convulsion as a rare symptom are also other generalized symptoms which the fifth nerve neurinoma can be presented with them (2). Case-based approach is one of the best methods to discuss and explain the interesting points for management of complicated cases in practical neurosurgery; so we would be glad to express our practical experiences in two different fifth nerve neurinoma cases. In the first case (solid tumor), A combined cerebellopontine angle (CPA) subtemporal approach in the lateral park-bench position was used and in the second one (the cystic tumor) retrosigmoid CPA suprameatal approach in the semisitting position was used.

Case Presentation

Case 1

A 31 year old man presented with long-lasting headache, binocular diplopia, unsteadiness ataxia and quadriparesis which have been progressed for about 6 months. Bilateral papilledema was detected during physical examination. His corneal sensation was intact (corneal Reflex had not been examined) (Figure 1). We operated on the patient in lateral Park-bench position using a combined subtemporal and retrosigmoid approach. Craniectomy was performed and drilling of petrous bone was done as much as possible to reach the widest extradural exposure. Then the dura of the posterior fossa opened which revealed no intradural tumor. Subtemporaldural opening was also negative for intradural tumor.

It was concluded that the tumor should be located interdurally and so the most probable tumor was the fifth nerve neurinoma. The mass was fleshy in texture and relatively hemorrhagic. We were lucky to extract the entire tumor when we reached the distal branches of the fifth nerve. Patient recovered from anesthesia in 24 hours post-op without any major neurological deficit but it was found that Lt corneal anesthesia on the second post-op day. Vision was intact and the patient was warned about corneal anesthesia under strict corneal protection with transparent moisturizing eye ointment.

Frequent corneal exam by ophthalmologist revealed persistent corneal epithelial defect. Tarsoraphy was done for 2 weeks. Patient's cornea recovered completely but he developed paradoxical CSF leakage from the mastoid air cell origin in the third week of post-op period which sealed by fat and glue successfully. Now the patient is intact except Lt facial paresthesia and Lt corneal anesthesia. Serial CT scan and MRI (\pm GAD) one year after operation is accompanied which is clear for tumor remnant (Figure 2).

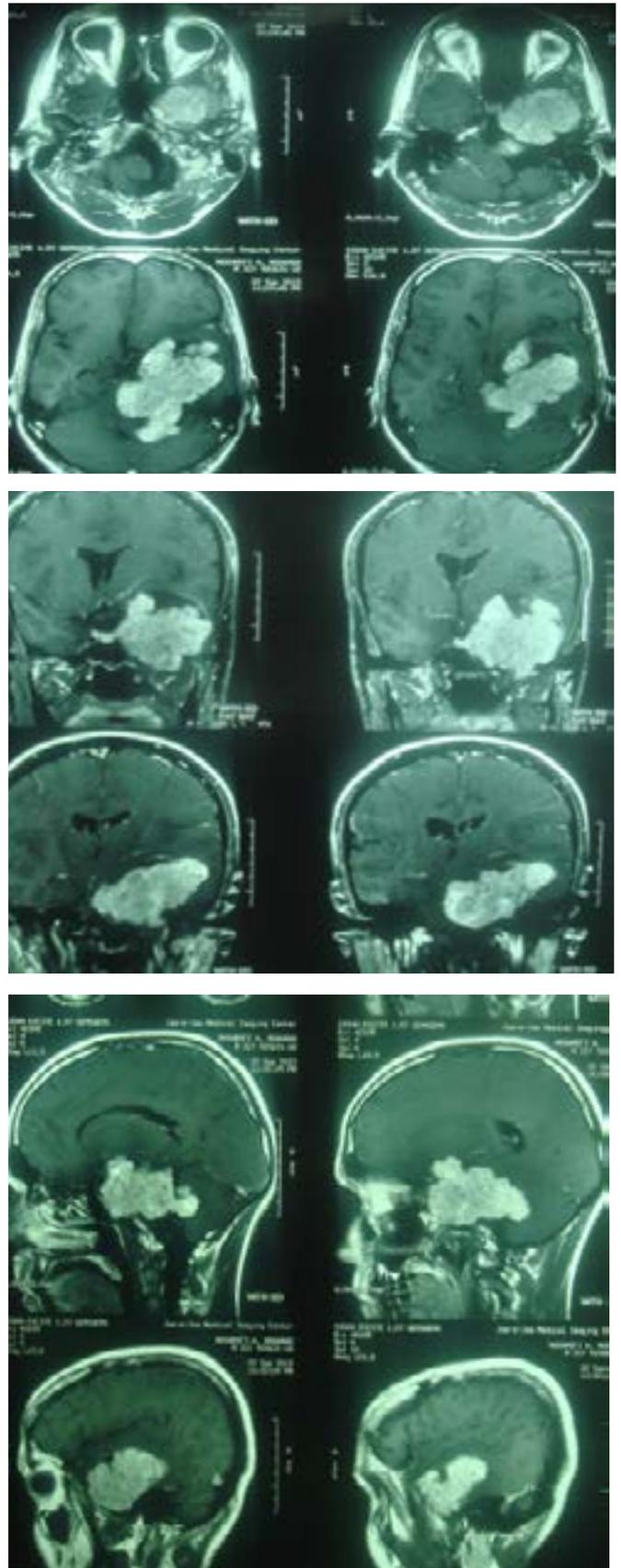


Figure 1. Pre-operation MRI

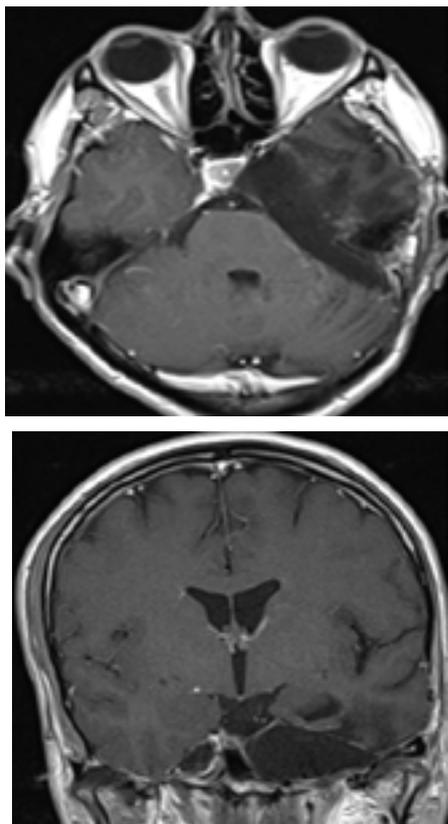


Figure 2. One-Year Post-operation MRI

Case 2

A 21 years old woman presented with unilateral craniofacial headache of long duration. She complained of binocular diplopia and atypical trigeminal neuralgia which have been progressed for about 3 months. Ipsilateral sixth nerve palsy and symmetric corneal reflex were also detected and her corneal sensation was also intact (Figure 3,4). In this patient due to the presence of large cystic component, we operated on semi-sitting position with standard retrosigmoid-subtentorial approach (Figure 5). The tumor exposed from CPA then we extended suprameatal corridor by cyst-evacuation and piecemeal evacuation of the tumor. At last we reached the small solid portion of the tumor attached to external side of the carotid artery extending to subtemporal fossa which we left it alone. The patient had quiet post-operative period and after recovery from anesthesia, we found corneal anesthesia in the right eye. Six months postoperative MRI (\pm GAD) revealed a small solid tumor remnant which was expected and also was out of access from the CPA approach. The patient is now neurologically intact except right facial paresthesia, Rt sixth nerve palsy and right corneal anesthesia. The patient was not warned of her corneal anesthesia.

The MRI revealed tumor remnant which was getting larger in the 6 month post-operation. We decided to operate on via right Pterional approach reaching anterior subtemporal fossa and so the entire tumor remnant was removed successfully (Figure 6).

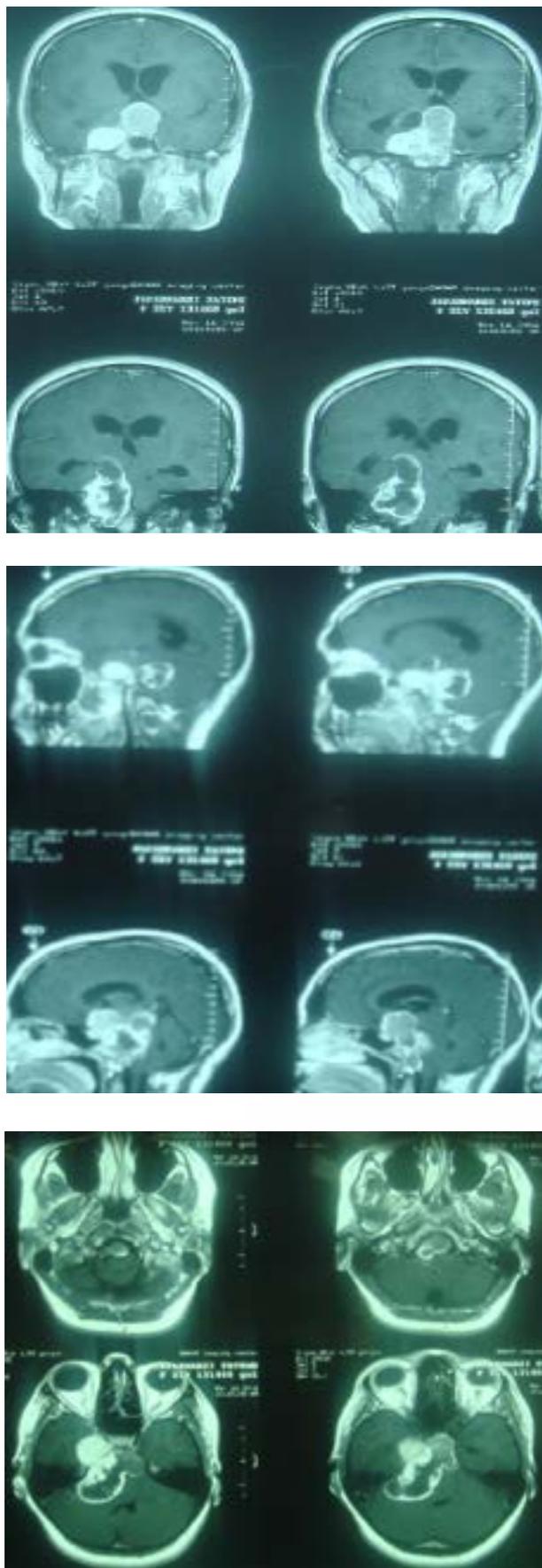


Figure 3. Pre-operation MRI



Figure 4. Pre-operation Bone Window CT Scan

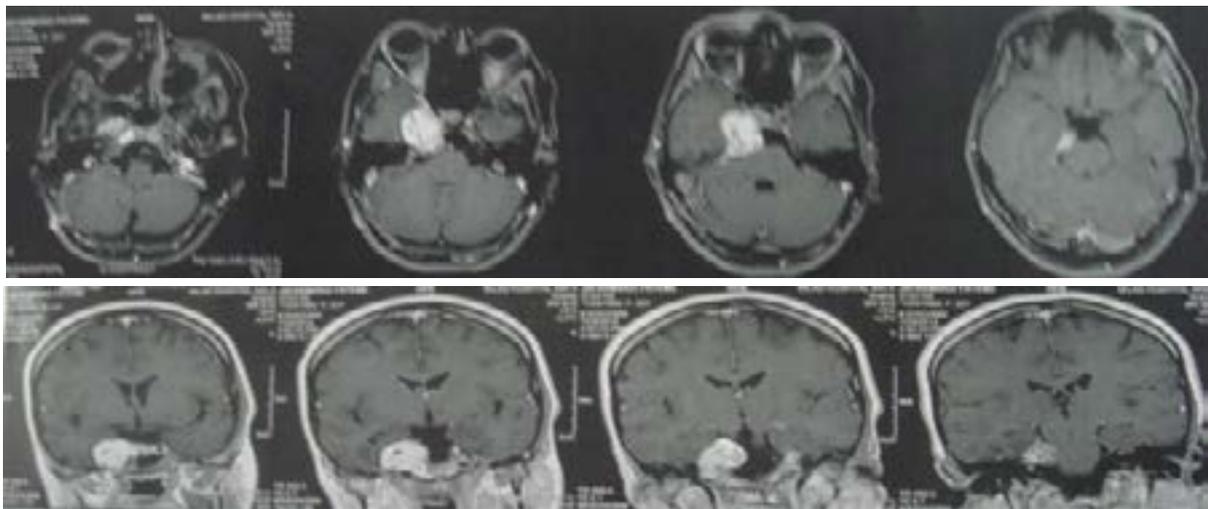


Figure 5. Six-month Intermediate Post-operation MRI

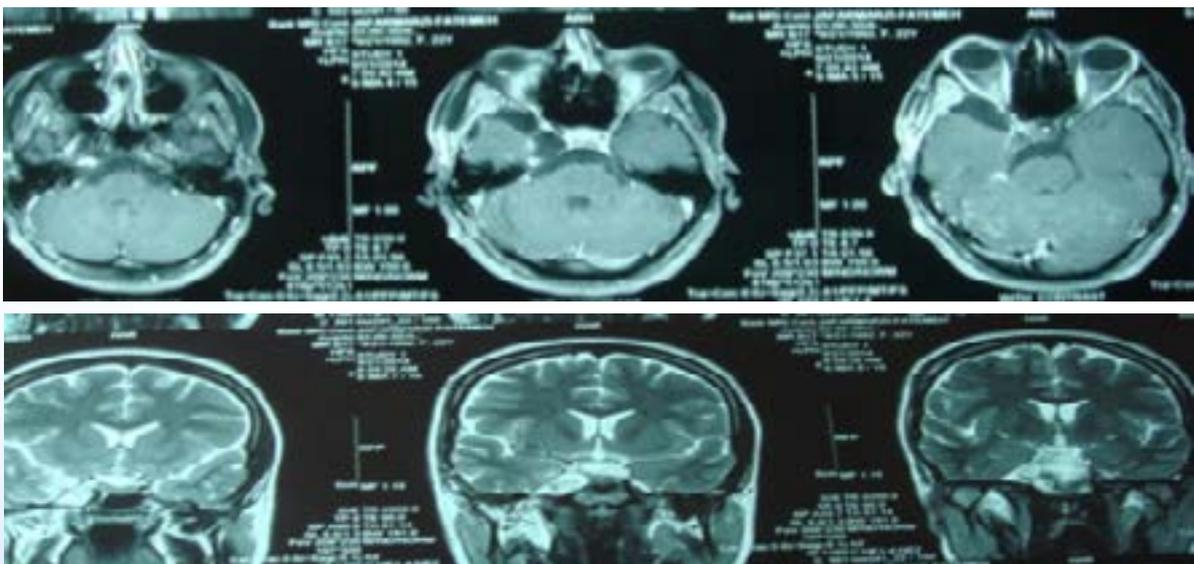


Figure 6. Nine-month Late Post-operation MRI

Discussion

Corneal anesthesia is a major threat regarding corneal preservation, so some questions may be posed as follows: 1) Is it wise to warn a neurosurgical patient about the lack of corneal sensation which was intact pre-operatively and the motor division of VII nerve is functional post-operatively? 2) Is the corneal nutrition impaired in iatrogenic corneal anesthesia cases? 3) Is the corneal preservation dependent on facial nerve function (intact vidian nerve) or the corneal somatic sensation is the only major determinant?

Secretion and production of tear:

Although the cornea is clear and seems to lack substance, it is actually a highly organized group of cells and proteins. Unlike most tissues in the body, the Cornea contains no blood vessels to nourish or protect it against infection. Instead, the cornea receives its nourishment from the tears and aqueous humor (a fluid in the anterior portion of the eye) that fills the chamber behind it (3). As we know, the secretion and production of tear is completely dependent on intact vidian nerve (a parasympathetic component of VII nerve) (4) Therefore it must be considered that in the cases of combined (Subtemporal + Retrosigmoid) approaches we should drill some of the basi temporal petrous bone where the vidian nerve may be injured inadvertently. (5) The least believable result of such a nerve injury would be xerophthalmia which the patient with intact corneal sensation complains of dry eye and burning sensation unilaterally but in the cases of iatrogenic post-op corneal anesthesia the patient may develop corneal ulceration without any painful complaint (6). In the cases of patients with 5th nerve neurinomas who undergo combined approach, we are exposed with iatrogenic post-op corneal anesthesia and at the same time we have no tear production due to vidian nerve damage which is in part approach dependent. Although the VII nerve motor function is intact but we have no enough tear to nourish and protect the cornea. On the other hand in the cases of retrosigmoid approach alone we should have intact vidian nerve and sufficient tear production to nourish the cornea is ensured (7). The role of intact V and VII nerve functions are the cornerstone of corneal preservation and protection (nutritional and mechanical view point). In preoperative period, the patient with intact corneal sensation and intact facial motor function should have intact blinking reflex. We recommend strictly to check corneal sensation, facial nerve function and preserved tearing post-operatively in order to protect the cornea (8). In the combined approach when the tumor located in pre-ganglionic or ganglionic portion, the injury of corneal sensation during operation is more likely to develop but when the tumor is located in post-ganglionic divisions, only those one involving the v1 division are likely to injure the corneal sensation (7). One extraordinary threat which we have had exposed with was the occurrence of Herpes ophthalmicus and corneal perforation without painful complaint (9). Finally the ambulated patients would have face wash and sweating so water and sweat exposure to cornea is a real danger to an anesthetic cornea in everyday activities.

Advantages and disadvantages of the approaches:

Better following of the proximal portion of the 5th nerve, better handling of subtentorial component, preparing a trans-lesional window to petroclinoidal junction in the case of cystic lesions are the advantages of Retrosigmoid (Suprameatal) approach and the threat to the vidian nerve damage is less probable than combined

approach. On the other hand better visualization of all of the mass and neighboring structures, better access to interdurally entrapped solid masses and also better handling of the neighboring vessels are the advantages of the combined (Retrosigmoid + Subtemporal) approach while the threat of simultaneous damage to V and VII nerves is considerable in this approach.

Complications

Corneal anesthesia CSF leakage (direct, paradoxical), cranial nerve damage (other than fifth) and vascular injuries are some of the surgical complications. We have been exposed with two of them in our cases.

CSF leak after a retromastoid approach can drain through open mastoid air cells into the middle ear cavity and then through the Eustachian tube to present as CSF rhinorrhea. This type of leak has been referred to as paradoxical CSF rhinorrhea (5). Watertight dural closure should be the goal following any posterior fossa approach. In case 1, we have paradoxical CSF leakage from the mastoid air cell origin in the third week of post-operation period which sealed by fat and glue successfully.

Main clues and tools indicating the diagnosis:

Any long lasting lesions involving the Meckel's cave. In other words any lesion involving the middle and the posterior fossa could be indicative of the presence of the 5th nerve neurinoma. However bone window CT scan to clarify the extent of bone destruction and to define neighboring bony structures (middle ear, Petrous Tip and Meckel's cave) and also brain MRI± GAD T1,T2 coronal and sagittal are the main diagnostic tools.

We should remember that the location of the 5th nerve neurinoma includes subtentorial Intradural, supratentorial Intradural and pure Interdural in some of them.

Conclusions

In general neurosurgery preoperative list of differential diagnosis should be limited and indicative of real anatomopathology in order to get the best result intra operatively and post-operatively. In order to have a safe surgical result, we do recommend that any masses located in the boundary of middle and posterior fossa in the Mediobasal portion (Meckel's Cave), should be defined delicately by skull base CT scan. In the cases of solid masses with different consistency we should keep in mind that, whether it is inter-dural or intra-durally located. We do also recommend pre-operative full examination of corneal sensation, facial motor function (blinking) and tearing status. Early Post-op examination of VII nerve function and corneal sensation are another recommendation. Occurrence of herpes ophthalmicus in any of such approaches is an ominous symptom in favor of the development of painless corneal perforation. At last warning the patient of corneal anesthesia in the cases of combined approaches because of the possible simultaneous damage to V and VII nerve should be considered. Consultation with an expert ophthalmologist for corneal care and temporary tarsoraphy if necessary is also recommended.

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None declared.

Conflicts of Interest

The authors declare that they have no competing interests.

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Comments

In interesting report about two cases of large schwannomas of the Meckel's cave reported during a 20 year period, the authors presented their findings and stressed upon several main points i.e. a) two stage and bicompartmenal approach to the lesions with the aim of gross total excision of the tumors, b) corneal anesthesia, the pathophysiology of lacrimation and the relation between 5th. N tumors and lacrimation. The case presentation is informative for younger neurosurgeons and the highlighted issues about the complication of corneal ulcer are also well taken. Unfortunately, the title and objectives raised by the authors have not been achieved; 1) What would be the learning curve about surgical management of 5th. nerve neuroma (in the title)? 2) What is the result of literature review on this topic (second paragraph in the abstract)? 3) How to preserve the nerves in and around the Meckel's cave (surgical techniques)? 4) Such an interesting communication needs a wide and subtitled discussion and certainly different key words

for the search in the relevant literature!! 5) Reference list need to me more inclusive. I would like to suggest the kind authors to publish another manuscript in the forthcoming issue of the Journal considering the above mentioned suggestions as the sister article and elucidate the issues more comprehensively.

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I should thank Dr. Bahrami and his colleagues for reporting these two typical and large size cases of trigeminal neuroma (1). They emphasized on the most important complications that neurosurgeons are faced with in approaches to these tumors, namely corneal anesthesia and loss of corneal reflex. They also delicately explained that how different anatomopathological location of the tumor and the type of approach may cause symptoms and complications respectively (drilling some of the basi temporal petrous bone where the vidian nerve may be injured inadvertently). Corridors that have been selected are also reasonable in both patients, and advantages and disadvantages of each approach have been discussed.

Neurosurgeons should be aware that, also total removal of the tumor is possible in some cases, pieces of the capsule with adhesion to vital structures (eg: brain stem, vascular elements) may not be safe to remove and should be left intact. Electrophysiologic monitoring of the fifth and seventh cranial nerve is essential and others may be monitored as needed. Future studies on the role of neuronavigation in outcome of resection of these lesions can be helpful.

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1. Bahrami E, Rahatlou H, Fattahi A, Astaraki Sh, Khani M, Shirvani M, Ghaempanah MJ. The Real Story of the Fifth Nerve Neurinoma: A Review of Our Learning Curve about Surgical Approaches and Associated Shortcomings. *IrJNS*. 2015;1(1):47-52.