CASE REPORT

Atypical Presentation and Treatment of Large Facial Neuroma: Case Report and Review

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ABSTRACT

Background: Facial nerve neuromas (FNN) are significantly rare brain tumors. Our objective is to report a case of an atypical presentation and treatment of a large facial nerve neuroma. We review current literature and discuss issues pertaining to postoperative outcomes of large facial neuromas.

Method: We retrospectively reviewed a case of patient who presented in our center with atypical symptoms of a large intracranial facial neuroma (schwannoma), our approach to treatment,outcome and combined it with review of current literature. A review of the current literature was performed by extrapolating data relating to clinical and management details pertaining to FNN, currently listed on the Google scholar, Clinical- Trials.gov database, and PubMed. This information was tabulated and compared with our experience.

Conclusion: Large facial nerves neuromas (FNN) are very rare brain tumors which may coexist with the patient without debilitating symptoms. When diagnosed, individualized approached to archive maximal resection while preserving surrounding cranial nerves and brain stem function must be the goal. Intraoperative patient positioning, is vital for attaining maximal view, brain relaxation and minimal bleeding.

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INTRODUCTION

Facial nerves neuromas (FNN) or Facial nerve schwannomas (FNS) are generally significantly very slow growing rare brain tumors. They are a schwannoma that arise from the facial nerve sheath which may involve its intracranial, intratemporal or extracranial segments . Bastinade M et al in a large case series, described in detail clinical presentations of patients presenting with FNN. Herein, we report the clinical picture, diagnosis, treatment, and outcome of a patient who presented in our center with a large FNN and atypical presentations.

METHOD

We retrospectively reviewed a case of patient who presented in our center with atypical symptoms of a large intracranial facial schwannoma and combined it with review of current literature. A review of the current literature was performed by extrapolating data relating to clinical and management details pertaining to LFN, currently listed on the Google scholar, Clinical- Trials.gov database, and PubMed. This information was tabulated and compared with our experience.

CASE REPORT

A 24-year-old male was referred to our center from a local hospital after discovery of a large cerebellar pontine angle space occupying lesion during a routine work medical checkup. The patient

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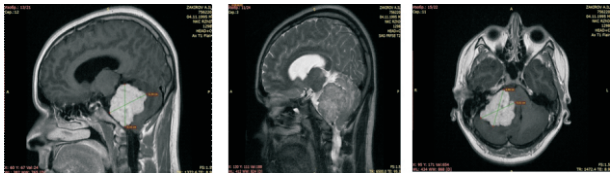
explained to have had a history of mild intermittent frontal headaches which did not interfere with his daily activities. Furthermore, neither he nor his wife noticed any changes in appearance. He denied having hearing impairment, tinnitus, facial tic, twitching, formication, imbalance, ataxia, pain, and nostalgia. Physical examination showed no somatic pathology and examination of other systems was unremarkable. The patient had normal facial nerve function. Laboratory values are normal. MRI of brain revealed a cerebellar pontine angle space occupying lesion (5cm × 4cm) closely attached to the brain stem with occlusive hydrocephalus.***(figure 1)***



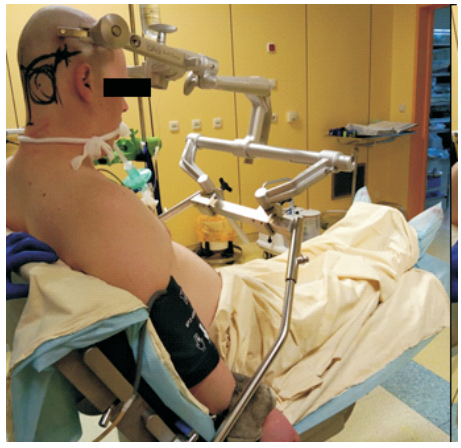
T1WI (sagittal) *T2WI (Sagittal)* *T1WI(AXIAL)*

Post- operative Coronal view ***Postoperative axial view***

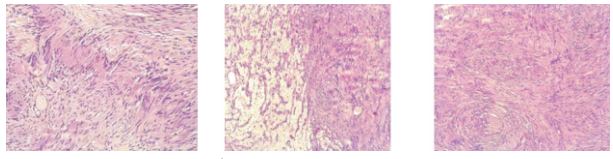
Figure 1 : Cerebellar pontine angel tumor and mild occlusive hydrocephalus

Treatment

Surgery was the mainstay of treatment. Intraoperatively the patient was placed in a sitting position ***(figure 2)***. A lateral suboccipital approach was deemed favorable in order to preserve facial and hearing function and while at the same time have early clear visualization of the bulbar group of nerves. Unavailability of intraoperative neurophysiological monitoring meant that early visualization was invaluable. Using microsurgical techniques, the tumor was debulked piecemeal, freeing it superiorly and laterally from the tentorium. The 5th and 8th nerve were easily separated from the tumor. However, the 7th nerve was intimately engulfed as depicted in ***figure 4***. Using CUSA and a nerve hook, the 7th nerve was separated from the tumor. Lastly, the closely adhesive medial aspect of the tumor to the brainstem was freed totally and conclusive gross total resection was archived without the need for nerve repair. Tissue specimens were sent for histopathology where an impression of schwannoma was made ***(figure 3)***. After surgery, the patient had facial nerve paralysis of House-Brackmann (HB) Grade III. At three months post-operative follow up, he had facial paralysis of HB Grade I.



Lateral Suboccipital approach, in sitting position head clamped using Doro fixation



a ×200 b ×100 c × 100

slide1. Neuroma.
Fig. 3a. Verocay body. Hematoxylin and eosin stain. × 200. Fig. 3b. Hypercellular zone with the formation of fascicular structures. Range Antoni A. Coloring hematoxylin and eosin. × 100. Fig. 3c. The junction area of the areas of Antoni A and B. Coloring hematoxylin and eosin. × 100.

DISCUSSION

Due to their subtle presentation, FNN are sometimes difficult to diagnose and requires a high degree of clinical suspicion. The presenting symptoms most frequently depend on tumor location, size, and histology. Large FNN are generally considered to be

more than 35mm in size or larger[4]. In this case the size was above 40mm - 50mm. At this size, apparent clinical features are expectant to have already manifested. Facial nerve neuromas that arise in the internal auditory canal (IAC) and cerebellopontine angle may manifest with a progressive sensorineural hearing loss similar to that caused by an acoustic tumor. The true diagnosis in such locations may be established intraoperatively. Unsurprisingly, during a large series conducted by McMonagle et al, patients in whom large FNN was diagnosed came complaining of hearing loss (either sensorineural or conductive hearing). Recurrent Bell's palsy, manifesting with unsatisfactory facial nerve recovery with each episode is another common complaint[5]. In the case of our patient, he presented more or less asymptomatic which is atypical for the size of the tumor. Kubota et al described 2 similar cases and attributed this to adaptive mechanisms the facial nerve assumes due to the slow growth of the tumor[3]. However, in their case, the patients were 40 and 64 years old and had other correlating symptoms such as vertigo, tinnitus and numbness of the of the tongue. As such it is expected that the size of the tumor would correspond to the age of the patient. In our case the patient's age and presentation do not correlate with the large FNN thus making it atypical in its presentation. That ***Table 1*** gives a review of literature of large FNN common clinical presentations, intraoperative approaches, and pre- and post-operative HB grades. As highlighted, other authors preferred retrosigmoid, infratemporal, middle fossa, transpetrosal, translabyrinthine or transmastoid approaches. We opted for a modified lateral suboccipital approach in a sitting position which could allow for more angles of view, reduction of venous pressures, and thus archiving better optimal brain relaxation. This in turn gave us early visibility of the 7th, 8th, 9th, 10th and 11th cranial nerves ***(figure 4)***, minimal venous bleeding, and shorter access to the CPA. We were able to archive total tumor resection without the need to sacrifice the facial nerve and surrounding vital structures which were closely adhered to the tumor. We were able to archive HB grade 1 three months post operatively, hearing preservation and minimal sequelae.

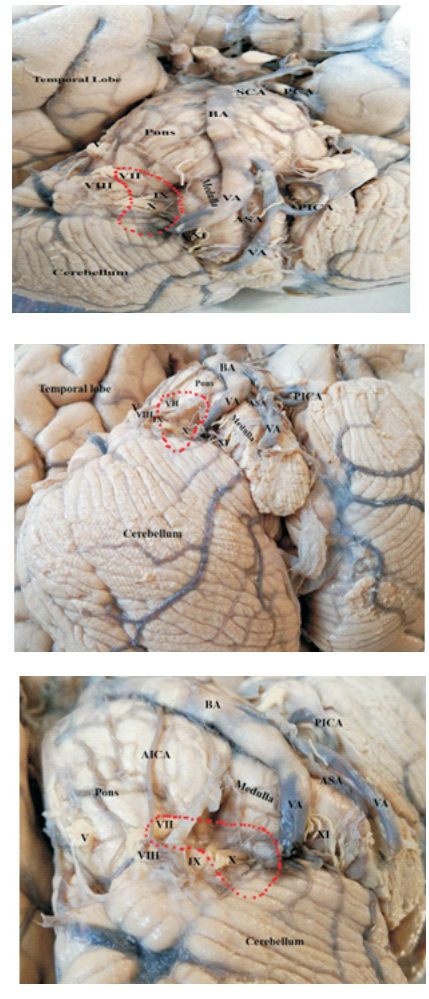


Figure 4: *BA- Basilar artery, PICA-Posterior Inferior Cerebellar Artery, VA-Vertebral Artery, ASA- Anterior Spinal Artery, SCA- Superior cerebellar Artery, V, VII, VIII, IX, - Nerves, Red boundary --- Tumor location.*

CONCLUSION

Large facial nerves neuromas (FNN) are very rare brain tumors which may coexist with the patient without debilitating symptoms. When diagnosed, individualized approached to achieve maximal resection while preserving surrounding cranial nerves and brain stem function must be the goal. Intraoperative patient positioning, is vital for attaining maximal view, brain relaxation and minimal bleeding.

Table 1 :

REVIEW OF LITERATURE

AUTHOR & YEAR	PATIENTs	SYMPTOMS	APPOACH	HB OUTCOME TOTOL NUMBER OF PATIENTS T/S (cm)	HB	Initial	Final	FNR
McMonagle et al 2008[5]	53	Facial weakness ,Tinnitus, Facial tic, twitching, formication CHL Imbalance, ataxia Pain, headache, otalgia Vertigo 'Dead ear' Recurrent facial weakness Synkinesis Epiphora Parotid mass Facial numbness Dysgeusia Otorrhoea IX palsy Xerostomia Sore throat Asymptomatic	TL RS TM, TMCF		I II III IV V VI LTFF	269	16 7 17 6 0 4 3	15
Soon H. Park et al 2014[7]	2	Tinnitus, facial palsy, infra auricular mass.	TM, TM+TP	(5×4) (3×2.5)	I II	1 1	0 2	10
Guyan A et al 2104[2]	12	facial nerve symptoms, including twitching, spasm, and palsy, cholesteatomas, hearing imparement	CyberKnife TM		I II III IV V VI	4 2 2 1 1 2	4 1 3 2 0 2	12
Theodore R et al 2011[6]	56	Hearing loss Tinnitus FN weakness Vertigo Facial hypesthesia	(TL) (54.7%) TM (11.3%), c (MCF) (11.3%), MCF (9.4%), retrosigmoid (9.4%), TL/MCF (3.8%)		I II III IV V VI	33 4 7 4 2 6	7 3 3 29	10
. Kubota et al 2005[3]	2	Vertigo tinnitus, right tinnitus, lingual paresthesia, and conductive hearing impairment	A right anterior transpetrosal		I II III IV	2	2	

FN: facial nerve; FN: facial nerve Reconstruction; ; HB: House–Brackmann; ITF: infratemporal fossa approach; MF: middle fossa approach; SN: sural nerve; SP: subtotal petrosectomy; preop: preoperative; postop: postoperative; TL: translabyrinthine approach; TM: transmastoid approach

ETHICS DECLARATION

Conflict of interest

The authors declare that they have no conflict of interest.

Ethical approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent

For this type of study, formal consent is not required.

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REFERENCES

1. Bartindale M, Heiferman J, Joyce C, Balasubramanian N, Anderson D, Leonetti J: The Natural History of Facial Schwannomas: A Meta-Analysis of Case Series. *J Neurol Surg B Skull Base* 80: 458-468, 2019.
2. Channer GA, Herman B, Telischi FF, Zeitler D, Angeli SI: Management outcomes of facial nerve tumors: comparative outcomes with observation, CyberKnife, and surgical management. *Otolaryngol Head Neck Surg* 147: 525-530, 2012.
3. Kubota Y, Kawamata T, Kubo O, Kasuya H, Muragaki Y, Hori T: Large facial nerve schwannomas without facial palsy: case reports and review of the literature. *Neurosurg Rev* 28: 234-238, 2005.
4. Mass SC, Wiet RJ, Dinces E: Complications of the Translabyrinthine Approach for the Removal of Acoustic Neuromas. *Archives of Otolaryngology–Head & Neck Surgery* 125: 801-804, 1999.
5. McMonagle B, Al-Sanosi A, Croxson G, Fagan P: Facial schwannoma: results of a large case series and review. *J Laryngol Otol* 122: 1139-1150, 2008.
6. McRackan TR, Rivas A, Wanna GB, Yoo MJ, Bennett ML, Dietrich MS, et al.: Facial nerve outcomes in facial nerve schwannomas. *Otol Neurotol* 33: 78-82, 2012.
7. Park SH, Kim J, Moon IS, Lee WS: The best candidates for nerve-sparing stripping surgery for facial nerve schwannoma. *Laryngoscope* 124: 2610-2615, 201