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INVITED EDITORIAL

RARE INSTANCES OF LONG TERM RECURRENCE IN TWO CASES OF VESTIBULAR SCHWANNOMA AFTER TOTAL TUMOR REMOVAL (13 AND 16 YEARS AFTER SURGERY)

*Sampath Chandra Prasad, **Eyad Abu Nahleh, ***Karthikeyan Balasubramanian, ****Mario Sanna

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ABSTRACT:

We report two rare cases of long term recurrence of vestibular schwannomas after gross total removal.Two patients, a 66 year old female and a 61 year old male, underwent surgery for vestibular schwannoma. In both patients an enlarged translabyrinthine approach was performed to achieve a gross total removal of vestibular schwannoma. Both the patients developed a tumor recurrence 13 and 16 years after surgery respectively. Recurrences after gross total tumor removal with Enlarged Translabyrinthine Approach is uncommon. Though different centres have different protocols, generally follow-up is restricted to a maximum of ten years after surgery since most recurrences tend to occur within this time period. This report points out to the fact that patients need to be put on prolonged follow up for at least 15 years even after gross total removal.

Running Title – Recurrence of vestibular schwannoma.

Key words – Vestibular Schwannoma, Recurrence, long term, hearing preservation.

INTRODUCTION:

Vestibular Schwanomas (VS) are the most common benign tumors of the cerebellopontine angle (CPA).[1] The translabyrinthine approaches (TLA) popularized by William House and the technical refinements brought about thereafter that led to the development of the enlarged translabyrinthine approach (ETLA) have made access to the tumor safe and exposure of even giant VS possible with minimal surgical sequelae. Recurrence rates after gross total removal(GTR) are extremely low[2] leading a few authors to advocate cessation of serial Magnetic Resonance Imaging (MRI) follow-up scans after a few years of surgery for VS after GTR. Here we present two cases wherein the patients developed a recurrent VS, 13 and 16 years after total removal with an ETLA.

Case 1 – A66 year old female patient presented to our centre in March 2000, with history of right sided hearing loss, ipsilateral tinnitus and incapacitating vertigo. ENT examination was unremarkable. Pure tone audiometry revealed a moderate to severe high frequency sensory neural hearing loss (SNHL) in the right ear. Brainstem Evoked Response Audiometry (BERA) on the same ear revealed an inter-aural delay in the Vth wave. A MRI revealed the presence of an intracanaliculartumor of approximately one cm in size. A diagnosis of a right sided VS was made. After explaining all the available modalities of treatment for VS to the patient, an approach of wait-and-scan was adopted. In March 2001, a repeat MRI showed that the tumor had grown by five millimetres (Figure-1). In the same month the patient underwent ETLA for excision of tumor. A GTR was achieved with preservation of both facial and cochlear nerves. Postoperatively, the facial nerve was House Brackmann (HB) grade I. The postoperative period was uneventful. She was followed up with serial MRI scans in the first, third, fifth and seventhyear after surgery, all of which were negative for recurrence (Figure-2 A,B). The next scan that was done in March 2014 and this time there was a clear demonstration of

Author Affiliations:

*Department of Otology and Skull Base Surgery, GruppoOtologico, Piacenza-Rome, Italy

**Department of ENT, Jordanian Royal Medical Services, Amman, Jordan

Corresponding author:

Dr. Sampath Chandra Prasad

MS, DNB, FEB-ORLHNS

Department of Otology and Skull Base Surgery, GruppoOtologico,

C/o Casa Di Cura Piacenza Privata SPA

Via- Emmanueli, 42, Piacenza, Italy 29121

E-mail: sampathcp@gmail.com

Ph: + 39 3898486174.

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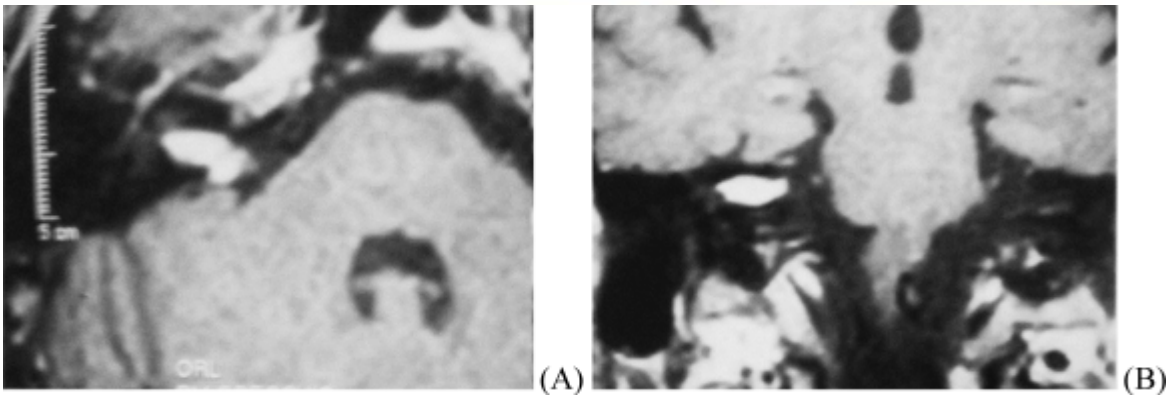

Fig.- 1 – Gadolinium enhanced T1 weighted MRI (A: axial, B: coronal) done in 2001 that showsa one cm VS in the right CPA.

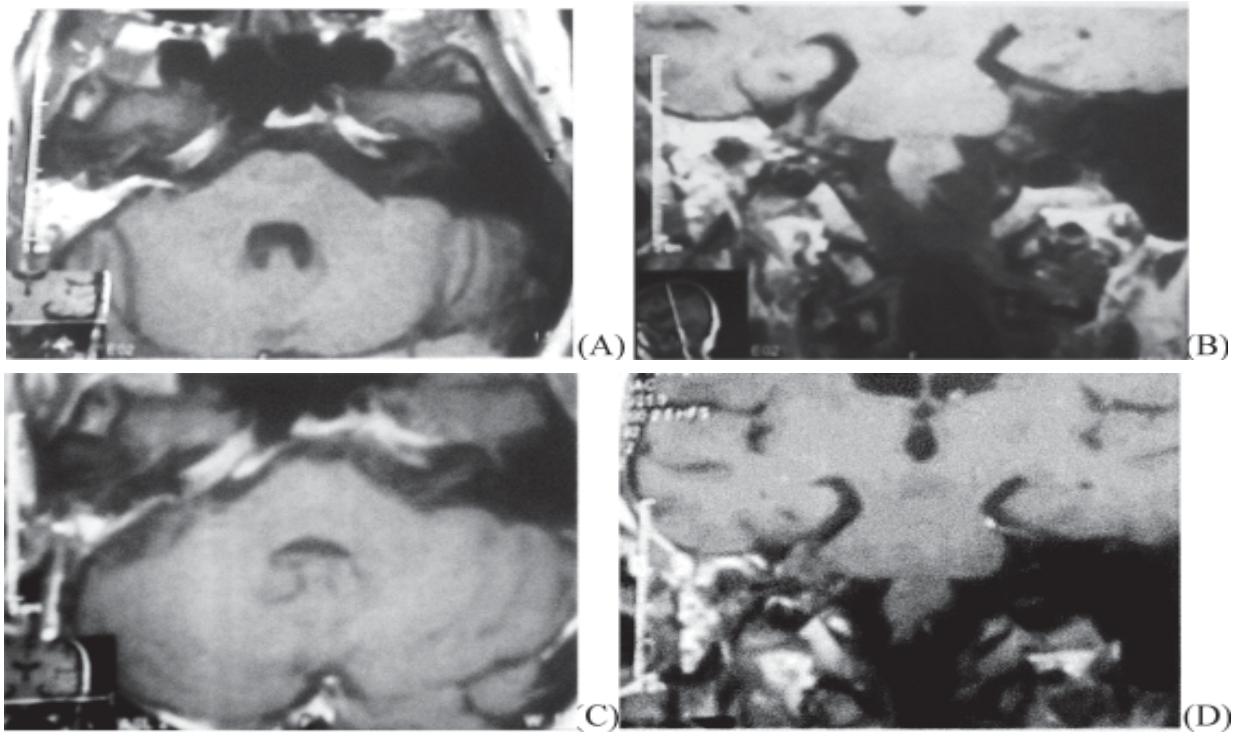

Figure 2 –Follow up MRI in 2002 (A: axial, B: coronal) shows no evidence of residual or recurrence of tumor after ETLA, B: Follow up MRI in 2004(C: axial, D: coronal) also shows no evidence of recurrence of tumor.

a one centimetre tumor in the CPA (Figure-3). The patient has been put on 'wait-and-scan' and the next scan is scheduled in March 2015.

Case 2 – In 1998 a61 year old male presented with tinnitus in the right ear and ipsilateral hearing loss. As in the first case, the ENT examination was unremarkable. Pure tone audiometry revealed anacusia in the right ear. Brainstem Evoked Response Audiometry (BERA) on the same ear revealed an inter-aural latency in the Vth wave. An MRI showed a 2 centimetre tumor in the right CPA. A diagnosis of a VS was made. In April 1998 the lesion was totally excised using an ETLA. As in the first case, the facial and cochlear nerves were preserved. On the 1st postoperative day, the patient experienced a facial weakness of HB grade II which improved on the 2nd day postoperative. The postoperative period was uneventful. Follow up MRI showed no recurrence in the first, third, fifth and seventh years(Figure-4A) postoperatively. In 2011, the patient underwent a coronary artery bypass surgery in another centre during which time a brain MRI was done. Despite the evidence of a small lesion in the CPA, this was reported as normal. In 2014 the patient came to our centre for a routine check-up during which the earlier scan was reviewed and the missed tumor was detected. A repeat MRI done one year later in April 2014 that confirmed

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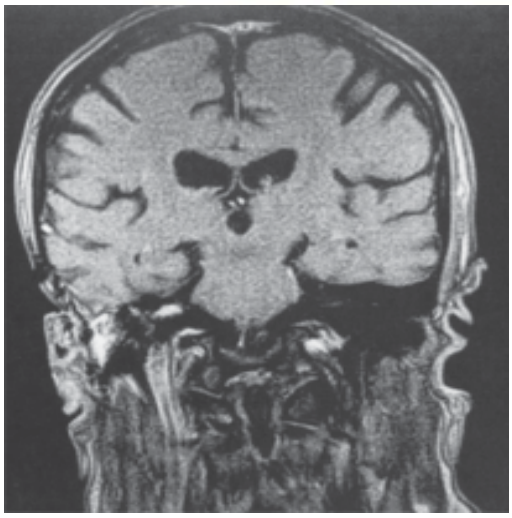

Fig.-3 – Follow up MRI in 2014 showsrecurrence of the tumor.

the recurrence(Figure-4 B). In the same month, the patient underwent a revision surgery using the earlier ETLA approach and the tumorwas totally removed.

DISCUSSION:

Over the last few decades, the ELTA has taken centre stage in the surgical treatment of VSs. This approach provides an unparalleled exposure of the CPA and also brings with it additional benefits ofproviding a high degree of safety to the facial nerve and the evasion of cerebellar retraction. Reported recurrence rates after TLA or ETLAare between 0% and 0.5%[2,3] which is very low when compared to other approaches to the CPA like the suboccipital (SO) /retrosigmoid (RS) or the middle cranial fossa (MCF) approaches.

The completeness of tumor removal during surgery was defined and classified in the report of the Consensus meeting on systems for reporting results in acoustic neuroma held in Tokyo in 2001.[4] This classification, which was revised in our earlier publication,[5] categorizes removal of tumor during surgery into four groups: 1) Gross total removal, 2) Near-total removal, 3) Subtotal removal and 4) Partial removal. The word 'recurrence' can be used only if there is demonstration of tumor during follow-up after GTR and the word 'residual' for the tumor that is left behind after incomplete excisions. The incidence of recurrence after GTRs with ETLA was0.05% (1/2011) in our earlier reported series of 2400 cases published in 2012. With the addition of the two cases against the updated series of ETLA, this rate comes down to 0.04% (3/2555).

The reasons for recurrences after GTR, despite the wide exposure achievedin ETLA, are unclear. Histological analysis have shown that VS lacks a true capsule and the tumor periphery is formed only by compressed neoplastic cells.[6] Tumor specimens have shown microscopic tumor ingrowth of the cochlear nerve[7] and demonstrated histologically inseparable planes between the facial nerve and the multifocal tumor. It must be noted that in both our cases the facial and cochlear nerves were preserved during the surgery. The vestibular and cochlear components of the eighth cranial nerve usually enter the Internal Auditory Canal (IAC) as a single unit. They divide into separate nerves within the IAC. The point of separation may vary among individuals. Only in the three to four centimeters of the most lateral part of the IAC do the vestibularand cochlear nerves usually divide into distinct, identifiable structures. At the fundus of the IAC, the vestibular nerve is separated into superior and inferior divisions by the crista falciformis.Therefore, the tumor may microscopically follow these nerves into their own canal in the fundus of the IAC. Visualization of this area requires removal of part of the bony partition between the vestibule and the lateral IAC.

Often because of the previous sacrifice of the vestibular and cochlear nerves, the recurrent tumor is asymptomatic until it reaches a large size and hence it is important to diagnose this by follow-up serial scanning with MRI. MRI with Gadolinium contrast is the gold standard for diagnosing VS with sensitivity and specificity reaching 100% and this

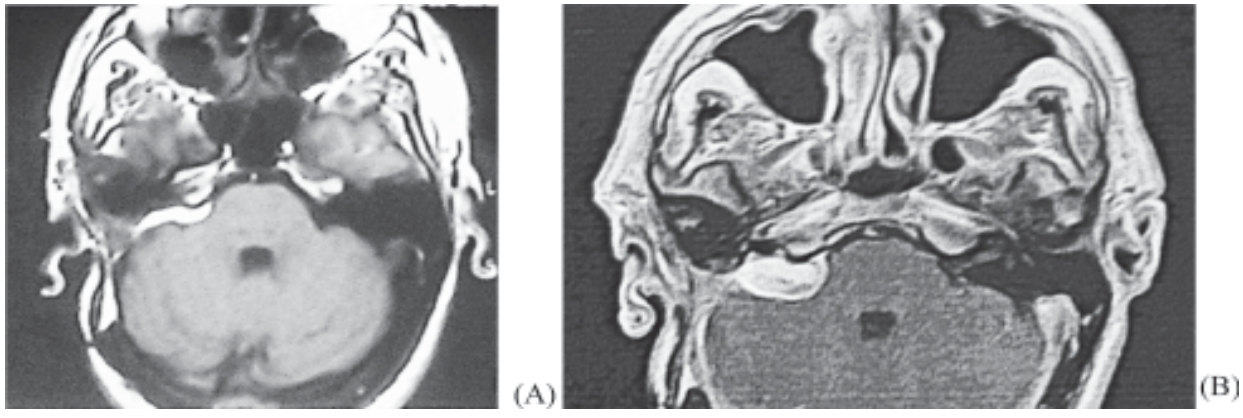

Fig.-4 –Follow-up MRIin 2005(A) shows no evidence of residual or recurrence of VS after ETLA. Follow-up MRI in 2014

holds true even for detecting recurrences.However, in detecting recurrences, there are certain difficulties that come in the way of an accurate diagnosis. The presence of scar tissue, fat or artefacts after surgery may also give rise to enhancements on post contrast T1 weighted MRI. The dural response to the surgical manipulation, development of granulation tissue and chemical inflammation induced by blood (hematoma)may be responsible for this.[3] Several authors reports that persistent nonspecific postoperative enhancement is commonplace following VS resection[3] but a linear enhancement is usually due to dural enhancement or scarring while a nodular enhancement correlates with a tumor.[8,9] Carlson et al.[10] reviewed over 200 postsurgical VS patients and correlated patterns of enhancement with tumor recurrence. Of them 66.0% displayed enhancing lesion regression and 3.5% showed complete resolution of enhancement.

The MRI protocols for follow-up of VS after surgery is also a matter of debate. It is obvious that a long term follow-up is necessary to differentiate between the recurrent/residualtumor and other factors that lead to enhancement. Despite the widespread clinical use of MRI in the last 2 decades, there remains no consensus regarding optimal timing or frequency of postoperative MRI surveillance. At the GruppoOtologico, we used to perform scans at one, three and five years after surgery for GTRs and discontinue follow-up is there was no evidence of recurrence. However with our latest experience we have modified this protocol. We now advice our patients to undergo scans at one, three and five, ten and fifteen years after surgery or in between in case of symptoms. The follow-up protocolmust be followed strictly especially after hearing preservation surgery due to a higher probability of recurrence. While Shelton[11] reported 0.3% recurrence rate, with an average time of recurrence of 10 years, Mazzoni et al.[9] claimed that the majority of recurrence VS regrowth between three and seven years from the surgery. After this timetumor recurrence was theoretically possible but extremely improbable.Like us, Carlson et al.[10] also recommend a repeat imaging at 15 years for linear enhancement during follow-up. Barring Roche et al.[12]who reported 9.2% recurrence rate after TLA for VSwith a follow-up ranging from 8 to 16 years there have been no reports of recurrences after 10 years of follow-up, nor have been authors advocating a follow-up for greater than 10 years. However, considering that the incidence of long-term recurrence (after 10 years) after GTR of VS is still very low (2/2553 cases; 0.08%) the cost-benefit ratio of following up all VS for five more years must also be considered.

CONCLUSION:

Recurrences after GTRs with ETLA for VS is extremely rare. We present two cases of recurrences after GTRs with ETLA 13 and 16 years after surgery for VS. This report points to the fact that prolonged follow up in such cases is necessary for detection of late recurrence of the tumor,more so in hearing preservation surgery. We suggest up to 15 years of follow-up with gadolinium-enhanced MRI scanning at the intervals of one, three and five years after surgery and once every five years thereafter for 15 years.

DISCLOSURES:

(a) Competing interests/Interests of Conflict- None
(b) Sponsorships – None
(c) Funding - None
(d) Written consent of patient- taken
(e) Animal rights- Not applicable

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