

Technical notes & surgical techniques

Vestibular schwannoma extending into the bulb of the jugular foramen

Eduardo Gavioli^a, Rafael Glikstein^{b,*}, Charles Agbi^c, Shawn Malone^d

^a The Ottawa Cancer Centre, Ottawa, ON, Canada

^b Department of Radiology, The Ottawa Hospital, Ottawa, ON, Canada

^c Department of Neurosurgery, The Ottawa Hospital, Ottawa, ON, Canada

^d Department of Radiation Oncology, The Ottawa Cancer Centre, Ottawa, ON, Canada



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1. Introduction

Vestibular Schwannoma, also known as Acoustic Neuroma, are benign brain tumors originating from the vestibular branch of the 8th cranial nerve presenting with a varying clinical course. Those tumors arise from the interface between the Schwann cells, which produce myelin sheath covering the peripheral and cranial nerves, and the oligodendroglia [2]. Schwannomas in general can be either intracranial or extracranial. Intracranial tumors arise from the cerebellopontine angle (CPA) and/or the internal auditory canal (IAC). Vestibular Schwannoma comprise approximately 8% of all intracranial tumors and, they tend to present between the fourth to sixth decades of life. The growth rate as described in the literature is similar to meningiomas, considered to be around 2 to 3 mm per year.

2. Case report

A 47-year-old patient presenting with gradual and progressive right-sided hearing loss that started eight years prior to the diagnosis. Associated symptoms included tinnitus, ear pain and decreased sensation over the right side of her face. As the patient lives in a remote community, her initial assessment was limited to physical examination without imaging investigation and she was initially managed with a hearing aid. Her hearing loss continued to progress and, she eventually became deaf on her right ear. Investigation with a brain MRI on 21st of September 2017 showed a large T1hypointense, heterogeneously T2 hyperintense enhancing mass measuring $4.0 \times 3.8 \times 3.6$ cm in the right internal auditory canal and cerebellopontine angle causing

significant mass effect on the brainstem, right middle cerebellar peduncle, and cerebellum. The lesion also had an inferior extension to the right cochlea and it was abutting the right jugular bulb and the internal jugular vein (Figs. 1 and 2).

On 23rd of November/2017 the patient underwent a right sided retrosigmoid craniotomy for debulking of cerebellopontine angle tumor with ultrasonic aspiration. Intraoperative neuromonitoring of the cranial nerves were performed to assess the facial nerve. Residual tumor was left in the interface with the seventh cranial nerve which was completely involved by the tumor. Postoperatively the facial nerve function was completely preserved. The pathological report showed a Schwannoma, WHO grade I with fragments of tumor of Schwann cell lineage, and several foci of Antoni A with some palisading; there was neither atypia nor mitotic activity.

The postoperative MRI from 25th of November/2017 showed residual enhancing mass at the cerebellopontine angle measuring $2.2 \text{ cm} \times 0.6 \text{ cm}$ (Fig. 3). At this stage the patient elected to remain on imaging surveillance declining adjuvant radiotherapy post-resection.

Surveillance MRI from 23rd of May/2018 showed progression of the residual mass now measuring $2.4 \text{ cm} \times 1.5 \text{ cm} \times 1.6 \text{ cm}$, extending into the right jugular foramen and internal auditory canal causing extensive remodeling (Fig. 4), there was also superior extension into the right prepontine cistern where the mass was abutting the root entry zone of the right trigeminal nerve. A repeat MRI from 08th of July/2018 showed stable findings. In the end of July/2018 the patient underwent Cyberknife radiosurgical treatment with a prescribed dose of 1800 cGy in 3 consecutive fractions.

* Corresponding author at: Department of Medical Imaging, The Ottawa Hospital General Campus, Room M2429a, 501 Smyth Rd, Ottawa, ON, K1H 8L6, Canada.
E-mail address: rglikstein@toh.ca (R. Glikstein).

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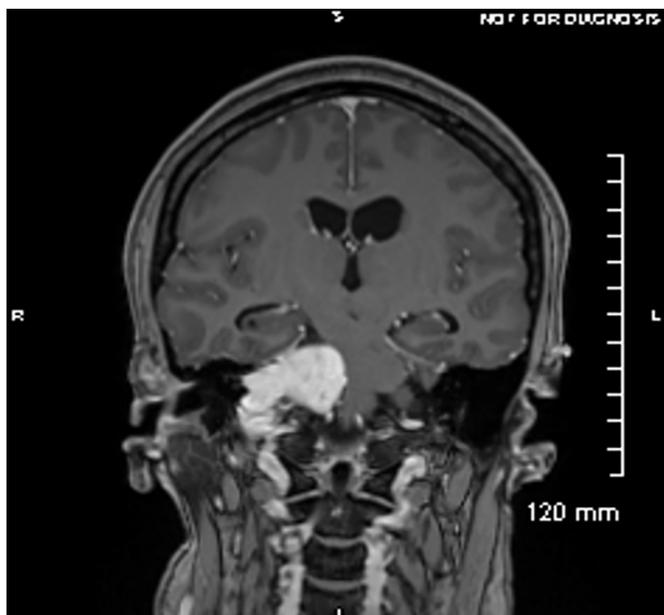


Fig. 1. Tumor extending inferiorly the right cochlea abutting the jugular bulb and the internal jugular vein.

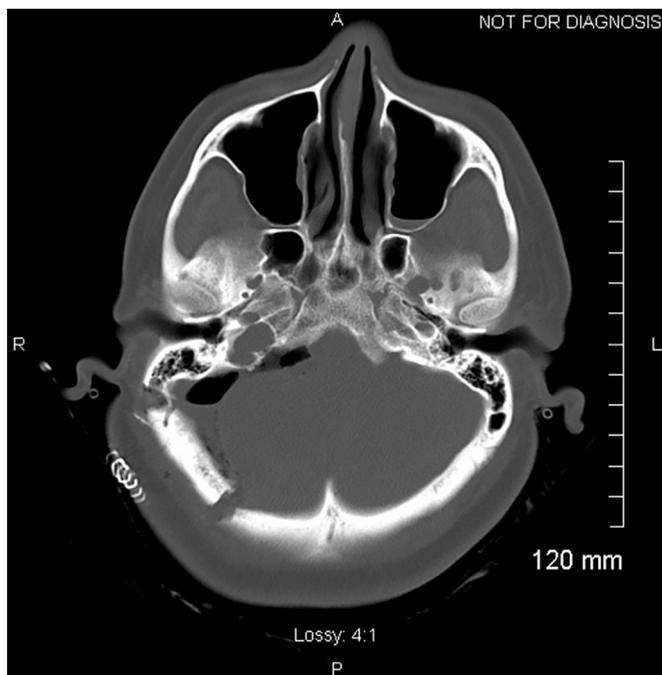


Fig. 3. Extensive remodeling of the internal auditory canal.

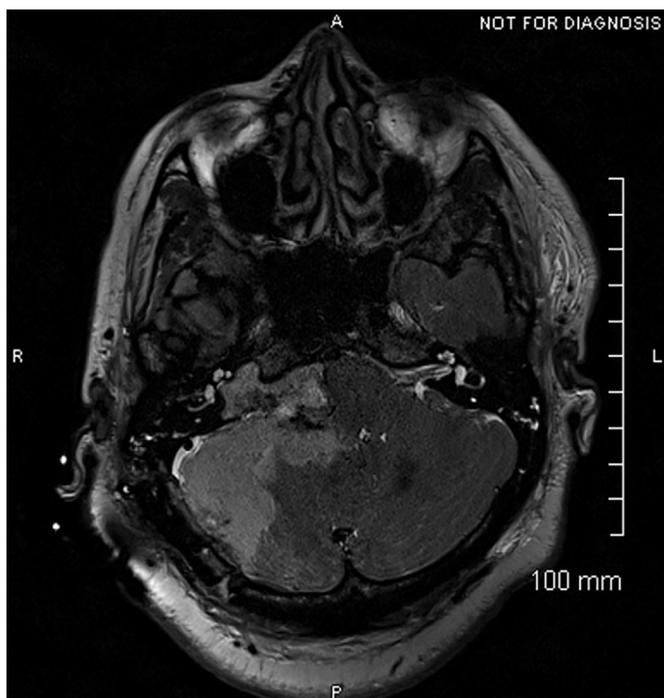


Fig. 2. Tumor extending inferiorly the right cochlea abutting the jugular bulb and the internal jugular vein.

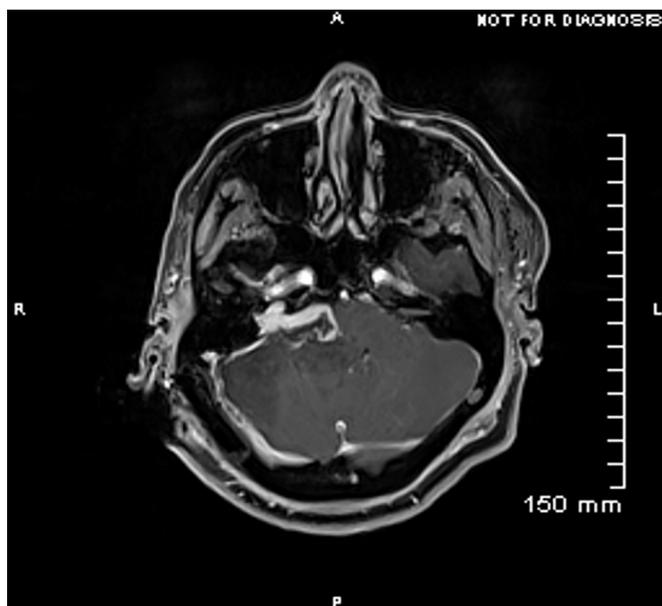


Fig. 4. Residual enhancement abutting the right jugular bulb and internal jugular vein.

3. Discussion

Vestibular Schwannomas can present with enlargement and/or erosion of the internal auditory canal [1]. What was unusual in this case was the extent of the remodeling of the internal auditory canal, with a 3.0 cm diameter. Also, the tumor extension into the jugular foramen abutting the internal jugular vein and tracking it inferiorly.

Vestibular Schwannomas are known to have a mean yearly growth pattern of 2 to 3 mm per year. In this case, in spite of being a WHO grade I tumor, the growth in tumor diameter over one year was around 10 mm, which is unusual for a grade I tumor.

According to the medical literature, fractionated radiosurgery offers a tumor local control above 90%. *Combs et al* report a 5-year tumor control rate of 93% in 106 patients treated in a single institution with median follow-up of 48 months. This patient was treated with CyberKnife Radiosurgery [3] with a prescribed dose of 1800 cGy in 3 consecutive fractions. The gross tumor volume coverage by the prescribed dose was 99.99% and, the planning target volume covered by the prescribed dose was 98.29% (Fig. 5). Organs at risk dose constraints were respected with the volume of brainstem receiving 1800 cGy was limited to 0.17 cc (dose constraint used: D1cc < 1800 cGy). The patient tolerated the treatment without issues.

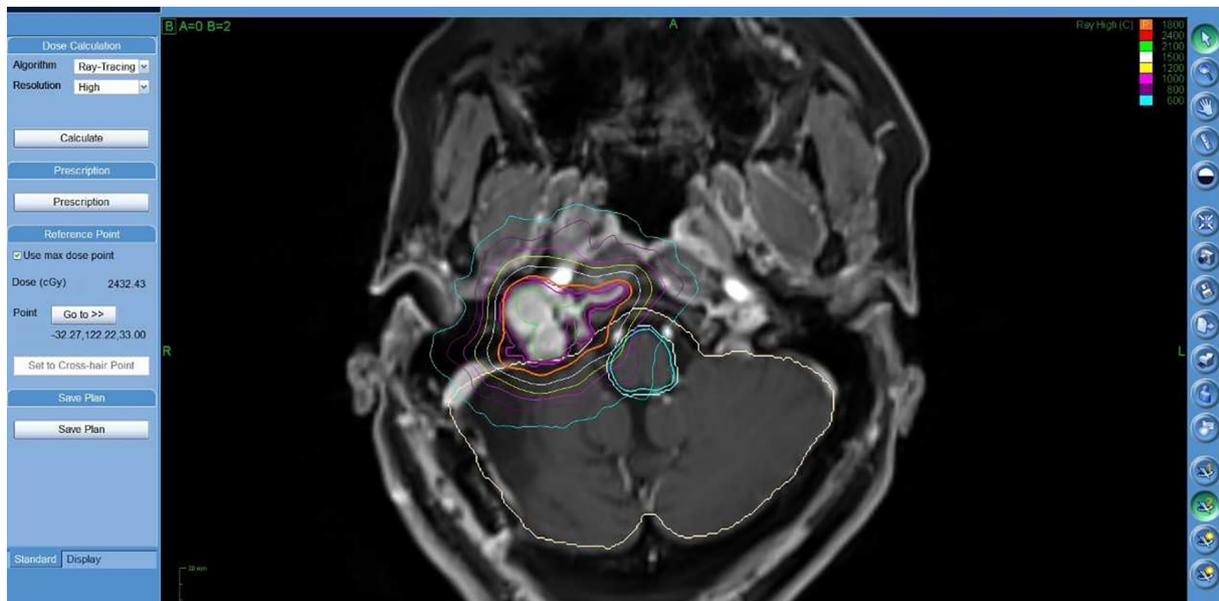


Fig. 5. Isodoselines distribution on Cyberknife planning.

4. Conclusion

The unusual presentation and learning points pertaining to this case include the extension into the jugular foramen extending below the level of the skull base, the extensive bone remodeling and erosion of the IAC and, the recurrence growth pattern rate of 10 mm in one year for a WHO grade I acoustic neuroma.

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