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Fast-Growing Vestibular Schwannoma

ABSTRACT—A case of a Jehovah's witness affected by an intracanalicular vestibular schwannoma with an extremely fast growth rate is presented. Nine months after presentation, the tumor reached 23 mm in the cerebellopontine angle. A partial removal through a retrosigmoid approach was planned. Because of the presence of a dominant high jugular bulb masquerading the internal auditory canal, the intracanalicular portion of the tumor was left in place. The residual tumor grew 12 mm in 2 months. Even after a gross total removal through a middle cranial fossa approach, the tumor recurred, reaching the size of 30 mm in 17 months. A modified transcochlear approach was then performed, and the patient was free of disease at the last radiologic follow-up, 8 months after the surgery. We illustrate our strategy in treating this aggressive benign lesion with unusual behavior.

Vestibular schwannoma (VS) is the most frequent lesion of the cerebellopontine angle (CPA), representing 80–90% of the tumors in this site.^{1,2} Its main treatment consists of surgical removal that can be accomplished by means of different approaches, depending on tumor size, preoperative hearing, patient age, and state of contralateral hearing.³ However, due to the fact that VSs are generally slow-growing lesions, in case of small tumors or in old patients a conservative management is sometimes suggested.^{4,5}

We present a case of a fast-growing VS complicated by the presence of a high jugular bulb in a Jehovah's witness.

CASE REPORT

In May 1996 a 44-year-old woman, a Jehovah's witness, came to our office with the diagnosis of a right internal auditory canal (IAC) tumor. Her magnetic resonance imaging (MRI), performed in another hospital (December 1995), revealed the presence of a neoformation completely filling the right IAC, consistent with a VS (Fig. 1). She complained of unsteadiness and tinnitus in the right ear for many years and progressive right hearing loss for the last 7 months. Audiologic examination revealed a right deafness; electronystagmography

Figure 1. Coronal MRI (December 1995) showing the intracanalicular tumor.

showed a weak response to caloric stimulation on the right side.

The patient was scheduled for translabyrinthine removal on June 1996. Because of her religious belief she refused to sign the authorization for a blood transfusion in case of necessity. The operation started and after the labyrinthectomy it was noted that the presence of a high jugular bulb with an overlapping diverticulum obstructed completely the route to the IAC. Because of the potential risk in displacing inferiorly a so high jugular bulb without knowing the patency of the torcular herophili and the size of the contralateral sigmoid sinus, it was decided to stop the surgery and to perform an angiographic examination. In fact, in case of lack of communication between the transverse sinuses or in case of hypoplasia of the contralateral venous system, a closure or thrombosis of the jugular bulb may be unsafe for the risk of a venous sufficiency of the temporal lobe⁶ or the development of a benign intracranial hypertension.⁷ The patient underwent selective angiography with study of the venous phase confirming the presence of a high jugular bulb with diverticulum and revealing the sigmoid sinus and the jugular bulb of the tumor side being much larger than the opposite side (Fig. 2).

Considering hazardous the option of jugular bulb displacement, the possibility of a middle cranial fossa (MCF) approach for removal of the tumor was discussed. The patient consented to undergo the new surgery, but this was delayed because her hemoglobin level was 10 g/dl, considered unsafe because of the patient's religious belief and her refusal to undergo any blood transfusion. Erythropoietin therapy was started under the control of the general practitioner.

In September 1996, 9 months after the first MRI, she underwent a new scan, and the tumor had reached the size of 23 mm in the CPA (Fig. 3). A different strategy was planned and it was decided to perform a staged procedure: the first stage by means of a retrosigmoid (RS) approach for the extrameatal portion and a second

Figure 2. The venous phase of selective angiography confirms the intraoperative finding of a high jugular bulb with diverticulum. The contralateral sinus and bulb appear much smaller than the tumor side.

stage through a MCF approach for the remaining intracanalicular portion.

The RS approach was accomplished in January 1997; intraoperatively the tumor was estimated to be much larger than the last MRI size. The patient was discharged on postoperative day 5, with a House-Brackmann grade VI facial palsy⁸ and a moderate weakness of the VI and X cranial nerves. A postoperative MRI performed before discharge showed a residual tumor limited to the IAC (Fig. 4). The histologic examination confirmed the radiologic diagnosis of VS, with a mixed pattern according to Antoni.

The second stage was planned 2 months later, on March 1997. On admission the patient had completely recovered from the weakness of the VI and X cranial nerves; facial function was still grade VI. Due to the fast growth rate previously showed by the tumor, the day before surgery she underwent a new MRI, detecting a new growth of the tumor, reaching again the size of 12 mm

Figure 3. Axial MRI (September 1996); the tumor reached 23 mm in size.

Figure 4. Axial MRI (January 1997) a few days after the retrosigmoid removal of the extracanalicular portion of the tumor. Note that the only tumor left is inside the IAC.

in the CPA (Fig. 5). However, a total removal was accomplished. The postoperative period was uneventful, and the patient was discharged on postoperative day 5. The histologic examination confirmed the previous diagnosis.

At the follow-up visit, 6 months after the last surgery, she was doing well, but the facial palsy was still present. The control MRI was negative for any residual tumor (Fig. 6). In July 1998, 10 months later, the patient underwent another MRI that showed a 15-mm recurrent VS (Fig. 7). However, the patient refused to contact our office for a consultation until February 1999, when she started to complain of marked instability. At that time the facial nerve function had recovered to grade IV. She underwent another MRI (7 months after the last one) that confirmed the presence of a recurrent tumor now reaching the size of 30 mm in the CPA (Fig. 8).

Figure 5. Axial MRI (March 1997), taken only 2 months after the scan showed in Figure 4, with a new growth of 12 mm.

Figure 6. Axial MRI performed 6 months after removal of the intracanalicular portion of the vestibular schwannoma (November 1997). No tumor remnants are visible.

To obtain the largest exposure for tumor removal, a modified transcochlear approach^{9,10} was then planned. A gross total removal was achieved, but it was not possible to preserve anatomically the facial nerve or to identify the medial segment of the nerve for grafting. The postoperative MRI was negative for residual tumor (Fig. 9). The patient was discharged on postoperative day 8 with a facial nerve palsy. The histologic examination confirmed again the diagnosis of VS without any sign of malignancy.

Two new MRIs performed 4 and 8 months after the last surgery were negative for any residual lesion (Fig. 10). However, the patient has been advised to repeat another scan in 8 months time.

DISCUSSION

In an attempt to select cases suitable for conservative treatment, the growth rate of VS has been the topic

Figure 7. Coronal MRI (July 1998); a recurrent vestibular schwannoma is present in the anterior compartment of the CPA, reaching 15 mm in diameter.

Figure 8. Coronal MRI (February 1999); the recurrent tumor grew until the size of 30 mm in the CPA.

of some investigation.^{4,5,11-13} However, a regular behavior has not been recognized yet, with different patterns occurring among similar tumors and also in the same tumor over the years. Usually, the fastest growth rate has been identified in cystic tumors, due to the increasing size of the cystic component, and in neurofibromatosis type 2 cases.³ Taking into consideration all kinds of VSs, the growth rate reported in literature ranges between 0 and 30 mm/year.⁴ The occurrence of a tumor increasing in size more than 10 mm/year is considered extremely rare. We are presently following six patients with a tumor in the only hearing ear, and the mean growth rate is 0.8 mm/year. Some of these cases have been previously published.¹³

In the literature, few studies have investigated the fate of small tumor remnants left unintentionally or on purpose during the surgery. The results showed different behaviors, leading to claim that a strict observation is necessary during the years after the surgery.^{5,14}

Figure 9. Axial MRI, T2-weighted images, before patient discharge after the last operation through a modified transcochlear approach. A postsurgical edema is visible at the level of the brainstem.

Figure 10. Axial MRI (May 1999), 4 months after the last operation, showing no evidence of recurrence. The postoperative edema has completely recovered.

As far as we know, the case presented in the report is exceptional regarding the growth rate of the original, residual, and recurrent tumors (23 mm in 9 months, 12 mm in 2 months, 15 mm in 10 months, and 15 mm in 7 months, respectively). The histologic examination failed to identify any peculiarity or any sign of malignancy. The only feature in the clinical history was the assumption of erythropoietin.

The rare possibility of fast tumor growth should be taken into consideration when adopting the wait and see policy. In our opinion, this policy could be questionable in cases in which the tumor at the first MRI is already over 1 cm in the CPA, if not in selected circumstances, as aged or poor general conditions patients.

The case reported was made more complex by the contemporary occurrence of the venous anomaly and patient refusal to authorize any blood transfusion. A high jugular bulb is sometimes encountered during CPA surgery, and some technical steps for its management have been described if it is discovered during translabyrinthine and RS approaches.^{15,16} However, in this particular case, due to the small size of the opposite sigmoid sinus, bulb displacement was avoided during the surgeries because it was considered hazardous. In fact, in this anatomic condition, thrombosis of the bulb and sigmoid sinus may originate an obstruction of the venous outflow, with the consequent risk of complications like venous infarction of the temporal lobe or the development of a benign intracranial hypertension.^{6,7,17}

Finally, it is the first case in our experience (460 VSs operated on from April 1987 to July 1999) and probably the first case reported in the literature in which all the main surgical approaches have been performed in the same patient for a VS removal. Due to the particular clinical history, the patient will remain under strict radiologic monitoring.

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REFERENCES

1. Moffat DA, Saunders JE, McElveen JT, McFerran DJ, Hardy DG. Unusual cerebellopontine tumors. *J Laryngol Otol* 1993;107:1087-1098
2. Kohan D, Downey LL, Lim J, Cohen NL, Elowitz E. Uncommon lesions presenting as tumors of the internal auditory canal and cerebellopontine angle. *Am J Otol* 1997;18:386-392
3. Sanna M, Saleh E, Panizza B, Russo A, Taibah A. *Atlas of Acoustic Neurinoma Microsurgery*. Stuttgart/New York: Georg Thieme Verlag, 1998. (controllare)
4. Charabi S, Thomsen J, Mantoni M, et al. Acoustic neuroma (vestibular schwannoma): Growth and surgical and nonsurgical consequences of the wait and see policy. *Otolaryngol Head Neck Surg* 1995;113:5-14
5. Rosemberg SI, Silverstein H, Gordon MA, et al. A comparison of growth rates of acoustic neuromas: Nonsurgical patients vs. subtotal resection. *Otolaryngol Head Neck Surg* 1993;109:482-487
6. Ohata K, Haque M, Morino M, et al. Occlusion of the sigmoid sinus after surgery via the presigmoidal-transpetrosal approach. *J Neurosurg* 1998;89:575-584
7. Brookes GB, Graham MD. Benign intracranial hypertension complicating glomus jugulare tumor surgery. *Am J Otol* 1984;5:350-354
8. House JW, Brackmann DE. Facial nerve grading system. *Otolaryngol Head Neck Surg* 1985;93:146-147
9. Sanna M, Mazzoni A, Gamoletti R. The system of the modified trancochlear approaches to the petroclival area and the pre-pontine cistern. *Skull Base Surg* 1996;6:221-225
10. Sanna M, Saleh E, Russo A, Taibah AK. *Atlas of Temporal Bone and Skull Base Surgery*. Stuttgart/New York: Georg Thieme Verlag, 1995
11. Charabi S, Engel P, Jacobsen GK, Tos M, Thomsen J. Growth rate of acoustic neuroma expressed by KI-67 nuclear antigen versus symptom duration. *Ann Otol Rhinol Laryngol* 1993;102:805-809
12. Thomsen J, Tos M. Acoustic neuroma: Clinical aspects, audiovestibular assessment, diagnostic delay, and growth rate. *Am J Otol* 1990;11:12-19
13. Sanjaya B, Sandeep K, Taibah A, Russo A, Sanna M. Vestibular schwannoma and the only hearing ear. *J Laryngol Otol* 1996;110:366-369
14. Mazzoni A, Calabrese V, Moschini L. Residual and recurrent acoustic neuroma in hearing preservation procedures: Neurological and surgical findings. *Skull Base Surg* 1996;6:105-112
15. Shao K, Tatagiba M, Samii M. Surgical management of high jugular bulb in acoustic neurinoma via retrosigmoid approach. *Neurosurgery* 1993;32:32-37
16. Saleh E, Aristegui M, Taibah A, Mazzoni A, Sanna M. Management of the high jugular bulb in the translabyrinthine approach. *Otolaryngol Head Neck Surg* 1994;110:397-399
17. Keiper GL Jr, Sherman JD, Tomsick TA, Tew JM Jr. Dural sinus thrombosis and pseudotumor cerebri: Unexpected complications of suboccipital craniotomy and translabyrinthine craniectomy. *J Neurosurg* 1999;91:192-197