Case Report

Rapidly Growing Cystic Vestibular Schwannoma With Sudden Onset Facial Palsy, Ten Years After Subtotal Excision

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An elderly male patient diagnosed with a right-sided cystic vestibular schwannoma (CVS) at our center underwent a translabyrinthine approach with a subtotal excision to preserve the facial nerve (FN). The tumor grew slowly for the first 9 years but in the subsequent 2 years grew rapidly, with the patient developing a FN paralysis. Using the previous approach, a second surgery was done and the tumor was excised, leaving behind a sheath of tumor on the facial and lower cranial nerves. This case demonstrates that CVSs show unpredictable growth patterns and need to be followed up for a longer period of time.

Key Words: Cystic vestibular schwannomas (CVSs), enlarged translabyrinthine approach, subtotal resection, rapid growth, facial nerve (FN) paralysis.

INTRODUCTION

Cystic vestibular schwannomas (CVSs) constitute about 5.7% of all vestibular schwannomas (VSs).1 CVSs generally are more aggressive than solid VSs and can present atypically, often with a brief clinical history or a preoperative facial nerve (FN) paralysis. Furthermore, they are notorious for rapid growth, resulting in a sudden deterioration in symptoms, unpredictable expansion of the cystic component, and hemorrhage—leading to severe mass effect and hydrocephalus.2–4

Patient underwent a translabyrinthine approach at our center, with a subtotal excision to preserve the FN. The tumor grew slowly for the first 9 years but in the subsequent 2 years grew rapidly, with the patient developing a FN paralysis.

The tumor showed a very slow growth for 9 years, followed by 2 years of very rapid growth with a sudden onset FN paralysis. A second surgery was performed using the previous approach and the tumor was excised; once again, a sheath of tumor was left behind on the facial and lower cranial nerves. This case is unique because it is the only case in a series of over 150 cases of incomplete VS resections (out of 3,200 operated VS) at our center that presented with a rapid growth after a lengthy period of follow-up. This case demonstrates the treatment dilemmas in managing a CVS, its unpredictable growth patterns, and the need for a longer period of follow-up.

CASE REPORT

In 2006, a 66-year-old male patient was diagnosed with a CVS measuring 25.0 mm × 16.6 mm on magnetic resonance imaging (MRI) (Fig. 1). The patient was operated via an enlarged translabyrinthine approach at our center, a quaternary referral skull-base institute in Piacenza, Italy. Intraoperatively, the FN was found to be stretched and pushed anterosuperiorly. A small piece of tumor (up to 5% of the original tumor) was left behind on the intracanalicular segment of the FN to preserve the FN function, thereby leading to a subtotal excision.

Postoperatively, the patient had a normal FN function. The patient was followed up with an MRI in the first year and subsequently once a year for 9 years (until March 2015). The remnant tumor grew at an average growth rate of 0.4 × 0.2 mm to reach a size of 20.6 × 7.6 mm in 9 years without causing any symptoms. Considering the patient’s age, normal FN function, and morbidity associated with the revision surgery, a decision was made to defer surgery and the policy of wait-and-scan was continued. However, at the next annual follow-up (February 2016), the tumor had grown rapidly to 25 × 9.4 mm (Fig. 2). Surgery was advised, but the patient chose not to get operated because he was asymptomatic.

Ten months later, the patient presented with a sudden onset of FN paralysis to reach a grade of House-
Brackmann (HB) VI. An MRI of the brain showed a multicystic lesion measuring 42.9 × 24.9 mm in the right cerebellopontine angle (CPA), with the tumor indenting the pons (Figs. 3 and 4). The medial globular cystic component measured 29.4 × 27 mm.

The patient was posted again for surgery; this time, a near-total resection was achieved wherein all the tumor was removed except for the tumor capsule around the lower cranial nerves and the intracanalicular segment of the FN, with an objective of saving the nerves. The postoperative period was uneventful. On the last follow-up after 3 months (Fig. 5), there was no improvement in the FN function.

DISCUSSION

CVSs constitute 5.7% to 48% of VSs.1,3,5–7 In our own series, the incidence was reported to be 6.8%.8

Etiopathology

The exact mechanism of formation of CVS is unclear. Various theories, such as isolated or repeated hemorrhages, hyaline, fatty or mucinous degeneration, and microcystic changes, have been proposed to be the causative factors. Furthermore, cystic enlargement has been attributed to the fluid accumulation either due to the osmotic effect or impaired blood–tumor barrier, resulting in extravasation of serum proteins. Cyst formation in VS
tumors and cysts located medial to the internal auditory that peripheral thin-walled, adherent, large cystic also is supported by Gagliardo et al.,9 who reported a cranial nerves, as observed intraoperatively. This fact resulted in an adhesion over the FN and other lower more medially in the CPA. This further might have ing intense pressure in a narrow area) at the periphery This must have resulted in a voluminous bleeding (caus- more a case of macrohemorrhage than microhemorrhage. expansion of the cyst in the last 10 months, it could is difficult to ascertain the real cause behind the rapid growth rate of tumor cells itself, as revealed by Ki-67 staining.6 When hemorrhage is the cause, in most instances it likely is a microhemorrhage, which forms into a cyst over a period of time. Although in our case it is difficult to ascertain the real cause behind the rapid expansion of the cyst in the last 10 months, it could more a case of macrohemorrhage than microhemorrhage. This must have resulted in a voluminous bleeding (caus- ing intense pressure in a narrow area) at the periphery of the cyst wall, leading to the creation of a larger cyst more medially in the CPA. This further might have resulted in an adhesion over the FN and other lower cranial nerves, as observed intraoperatively. This fact also is supported by Gagliardo et al.,9 who reported a case of rapid growth in a CVS in an elderly hypertensive patient with involvement of cranial nerves V to X. They hypothesized that the rapid growth could be due to mac- rohemorrhage, leading to mechanical conflict with the adjacent cranial nerves. However, the patient in the present report did not have hypertension.

Subtotal Resection/Near-Total Resection in Cystic Vestibular Schwannoma

The CVSs exhibiting fluid–fluid levels on MRI suggesting intratumoral bleeding have been implicated for adhesions to surrounding neurovascular structures such as the FN and brainstem.10 Aggressively dissecting the tumor capsule from the cranial nerves, the anterior inferior cerebellar artery, or an indented brainstem to achieve a total removal can be dangerous.1,11 It is prudent to leave behind the tumor capsule on these neurovascular structures. In our experience,8 we have found that peripheral thin-walled, adherent, large cystic tumors and cysts located medial to the internal auditory canal are best dealt with by a subtotal or a near-total resection.

Facial Nerve Outcomes After Subtotal Resection/ Near-Total Resection in Cystic Vestibular Schwannoma

Poor FN outcomes have been reported in the CVS compared to the solid VS of comparable size due to many factors, such as difficulty in obtaining an arach- noid plane, unusual position of the nerve resulting from displacement by the tumor, and low resistance offered by the tumor after evacuation of the cyst.4,5 However, in our series the long-term FN outcomes of CVS when com- pared with solid VS did not show any significant differ- ence, likely because we prefer to leave behind the tumor capsule on the FN in order to preserve its function.9 In another study on residual VS constituting 111 cases, we found that 62 (55.86%) of tumors were cystic12 and that the FN function was HB grade I through II in 33 (49.25%) and 18 (47.37%) in subtotal and near-total resection, respectively, over a 1 year follow-up.

Long-term Follow-up (Regrowth) in Subtotal/ Near-Total Resection in Cystic Vestibular Schwannoma

The incidence of regrowth of residual tumors was 6.3% (7 cases all in subtotal resection, including the present one in CVS) in our study involving 111 cases of residual VS, of which six (30%) were noted in solid VSs (20 patients) as against only one (5.56%) noted in CVS (18 cases). We recommended a minimum follow-up period of 7 to 10 years for all cases of residual VS to observe for regrowth for the same reason.12 The present case was an exception to all other CVSs in our series because it was the only case that showed dramatic growth after a period of slow growth for as many as 9 years. A study by Kameyama et al.13 demonstrated that of the 19 residual cases, 10 showed regrowth, of which five were CVS that showed a growth doubling time of 4.5 years as compared to 15 years observed with solid VS. Most of them required reoperation.

CONCLUSION

Residual CVSs are unpredictable and can show growth anytime in their follow-up period, making management especially tricky in the elderly. Although micro- hemorrhages are common, it is important to be aware that macrohemorrhages also can occur, and if so can cause serious neurovascular complications in a very brief period. Periodic yearly follow-up, even after more than 10 years, may be necessary to observe for rapid progress- ion of the residual lesion.

BIBLIOGRAPHY


