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Case Report

An unusual case of repeated intracranial hemorrhage in vestibular schwannoma

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Abstract

Background: Symptomatic intratumoral hemorrhage (ITH) in vestibular schwannoma (VS) is rare. A repeated hemorrhage is, therefore, even more exceptional. Repeated ITH has been reported in four cases thus far in English literature. Here, we describe a patient with a Koos grade D VS who presented to our Skull Base team with repeated ITH and an unexpected disease course.

Case Description: A 76-year-old woman presented with hearing loss due to polycystic VS on the left side. Five years later, the patient was presented with facial palsy caused by hemorrhage in the VS. The patient had an eventful medical history that necessitated the use of anti-coagulants. The patient suffered from three subsequent hemorrhages preoperatively and one hemorrhage 36 h postoperatively.

Conclusion: We have experienced multiple repeated hemorrhages in a patient with a polycystic VS, and despite surgical intervention, the outcome was unfavorable.

Key Words: Acoustic neuroma, repeated intratumoral hemorrhages, vestibular schwannoma

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INTRODUCTION

Vestibular schwannomas (VS) are benign masses of the 8th cranial nerve (CN VIII). Approximately 8–10% of all intracranial tumors are VS. They are considered to be the most common tumor in the cerebellopontine angle (CPA), as they constitute 75% of the tumors found in this area.[4] Clinical presentation varies from sudden asymmetric hearing loss to gait ataxia. Intratumoral hemorrhage (ITH) within VS is considered a rare event.[4] However, repeated ITH is even more uncommon. To our knowledge, repeated ITHs with two instances of bleeding have been reported in four cases in the literature.[1-3,7] Different theories have been proposed to explain the pathogenesis of the hemorrhage. An invasion of the vascular wall by tumor cells and vascular proliferation followed by necrosis are plausible explanations.[1] Our skull base surgery team was recently confronted with an unusual case of a patient with multiple repeated ITHs with an unexpected disease course.

CASE DESCRIPTION

A 76-year-old female patient was diagnosed with a left polycystic VS Koos grade D in the cerebellopontine angle

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in 2011 [Figure 1a and b]. The patient suffered from severe hearing loss, which was also the presenting symptom. Because of advanced age and serious cardiovascular comorbidity, a wait and scan policy was followed.

The medical history of the patient included hypertension, hypercholesterolemia, thrombosis of the right carotid artery, acute myocardial infarction, total knee replacement, thoracic and lumbar fractures, diabetic retinopathy, aortic valve replacement, coronary artery bypass surgery, and postoperative arterial fibrillation. The patient used an extensive list of medications, which included anti-coagulants.

The VS remained stable on follow-up. In May 2016, the patient presented with a severe headache and a facial palsy House and Brackmann (HB) grade 5. Computerized tomography (CT) scan showed an ITH [Figure 1c]. The patient was then hospitalized in a peripheral hospital. The anti-coagulant therapy was stopped accordingly. After a couple of days, the facial palsy improved to a HB grade 3 and the patient was discharged. However, 1 week later, the partial facial palsy progressed to a HB grade 6. The CT scan revealed a second hemorrhage in the tumor [Figure 1d]. The patient was then referred to the Maastricht University Medical Centre for treatment. Neurological examination upon arrival revealed an optimal EMV score with a facial palsy HB grade 6. Other cranial nerves were intact. A few days later, the patient deteriorated to a score of E3M6V3. The CT scan showed another bleeding resulting in a strong mass effect on the brainstem [Figure 1e]. In addition, there was a bilateral dilation of the lateral ventricles and the third ventricle consistent with hydrocephalus. An emergency retrosigmoid approach was employed to evacuate the hematoma and resect the VS as much as possible. In addition to these measures, an occipital external ventricular drainage was applied. During surgery, the tumor appeared very necrotic. One day following surgery, the patient recovered to an optimal EMV score. Approximately 36 hours after surgery, the patient deteriorated rapidly. A CT scan showed another hemorrhage in the same area, however, more directed toward the brainstem [Figure 1f]. Unfortunately, the condition of the patient worsened quickly and the patient deceased.

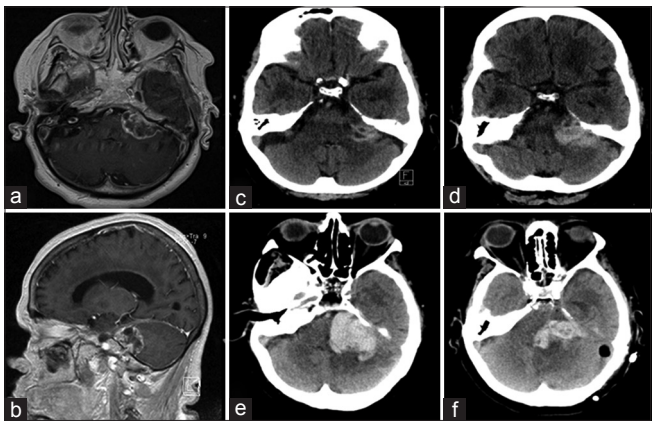

Figure 1: (a) An axial T1 weighted magnetic resonance imaging (MRI) of the polycystic vestibular schwannoma with a mass effect. (b) A T1 weighted MRI sagittal view of the vestibular schwannoma. (c) Computerized tomography imaging of the first intratumoral hemorrhage in vestibular schwannoma; (d) Second hemorrhage; (e) Third hemorrhage; (f): fourth hemorrhage 36 h after intervention

Histopathological examination confirmed the diagnosis of VS. Furthermore, it showed an extensive hemorrhage with small focal fragments of fusiform proliferation and thick-walled dilated blood vessels. Immunological examination showed a positive S100 and a low mitotic activity (MIB1) in tumor areas. Glial fibrillary acidic protein (GFAP) staining was negative. The tumor area showed inflammation and contained eosinophil cytoplasm with elongated nuclei.

DISCUSSION

ITH in VS is uncommon.[4] Repeated bleeding nonetheless is extremely rare. Our case is the 5th to be reported in literature thus far, and the first with multiple repeated ITHs, as far as we are aware of. Different risk factors have been suggested for ITH in VS which include tumor size (>2 cm), abnormal vasculature, radiosurgery, and rapid growth of the tumor. A few cases have reported that the use of anticoagulants, methotrexate, cocaine, and trauma can result in ITH.[4] Three of these factors apply to our patient. They include large tumor size, abnormal vasculature, and the use of anti-coagulants. Hitherto, tumor size and abnormal vasculature composition seem to be the most important factors. All five cases made note of a relatively large tumor and an abnormal vasculature. However, it has been postulated that the change in the size of the tumor is attributed to microhemorrhages which then lead to its expansion.[6] All patients reported sudden headaches at the time of the recurrence of the bleeding.

Cystic vestibular schwannoma (CVS) has been described to have a more rapid expansion rate than the solid ones, faster involvement of nerves, and expression of symptoms as well as erratic behavior. It is also postulated that the formation of the cyst is due ITH.[5] This, in addition to the use of anti-coagulants, could have played a role in the formation of the multiple ITHs in this case.

A notable similarity between our case and that of Mandl[3] is that both the cases had ITH 5 years after initial diagnosis. In the case of Takeuchi,[7] it was reported that the intermission between the first hemorrhage and the repeated one is at least 1 month. Our case has shown that this is not always the case. Four cases, including ours, have reported hearing loss to be one of the initial symptoms.[2,3,5] The HB grade was different in all cases,

and thus does not seem to be a good prognostic factor. It is nonetheless noteworthy that one group[5] reported a postoperative hemorrhage in one patient with CVS which necessitated surgical interference. However, the authors did not report any preoperative bleeding.

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Conflicts of interest
There are no conflicts of interest.

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