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Case Report

Fatal delayed post-operative cerebral venous thrombosis after excision of hypoglossal nerve schwanomma

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Summary

Lower cranial nerve schwanommas are rare tumours. We present a 35 year old female patient who had a lower cranial nerve schwanomma with both intracranial and extracranial components. The internal jugular vein was injured during the dissection of the extracranial portion of the tumour. Ligation of the internal jugular vein is not associated with significant post-operative complications. Our patient however, developed retrograde cortical venous thrombosis on the 14th post-operative day resulting in multiple areas of haemorrhagic venous infarction with raised intracranial pressure. Such a delayed contiguous cortical venous thrombosis has not been reported. We present this report to highlight this event and to outline the probable causes for the same.

Keywords: Internal jugular vein; injury; ligation; thrombosis; delayed; cortical veins; lower cranial nerve; schwanomma.

Introduction

Schwanommas arising from a single lower cranial nerve are rare. Of these, the hypoglossal nerve is the most common nerve involved [6]. These patients present with wasting and paralysis of the tongue along with other cranial nerve involvement, cerebellar signs and features of brain stem compression depending upon the size of the tumour. Little is known about the natural history of these lesions. They are a challenge to the neurosurgeons due to their combined intracranial and extracranial growth. Surgical excision of these lesions is the treatment of choice. The choice of surgical approach depends upon the location, extension and the nature of the lesion. Tumours with a predominant intracranial component are approached via the lateral suboccipital route whereas those extending extracranially into the neck are removed through a cervical exposure. Ligation of the internal jugular vein is a common surgical practice while excising these tumours. This is not routinely associated with significant post-operative complications [25]. We present a patient with hypoglossal nerve schwanomma who developed internal jugular vein thrombosis post-operatively and later on had cortical venous thrombosis. This complication to our knowledge has not been described in treatment of these tumours.

Clinical details

A 35 year old female patient had a histopathologically proven 12th cranial nerve schwanomma with lower cranial nerve involvement. She had undergone partial tumour decompression twice over a period of 4 years. She had been planned for adjuvant therapy but was lost to follow up. She presented again with signs of progressive brain stem and cervico-medullary junction compression five months after her second operation. Magnetic resonance imaging (MRI) scan showed significant tumour regrowth with both (Type 4) intracranial and extracranial tumour extension (Fig. 1A, B). Intracranially, the tumour was causing significant compression and displacement of

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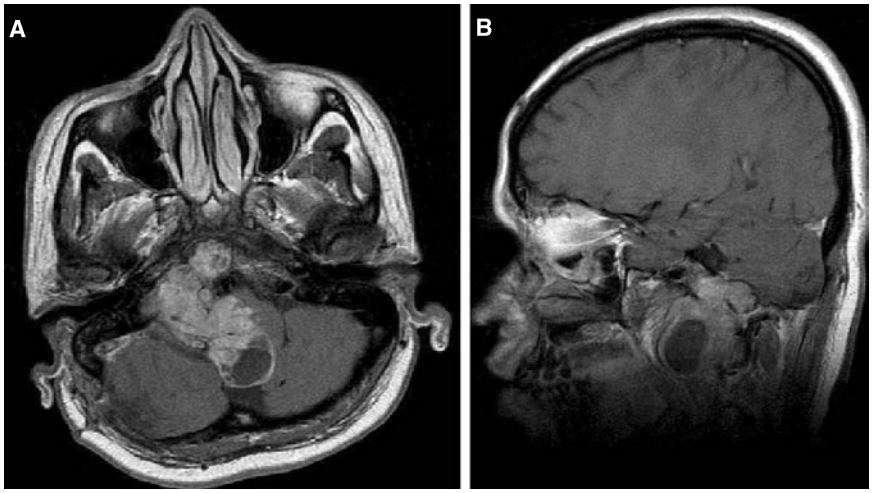

Fig. 1. Pre-operative MRI (T1W with Gadolinium) with intracranial and extracranial tumour extent. (A) Axial, (B) sagittal

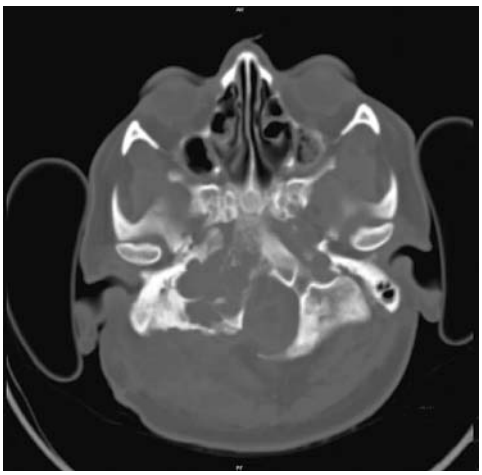

Fig. 2. Pre-operative CT scan (bone window) with destruction of the posterior fossa skull base (occipital, petrous and clivus)

the medulla and cerebellar hemisphere. It had eroded the posterior cranial fossa base on the right side (Fig. 2) and was extending extracranially into the parapharyngeal space almost upto the C3 vertebra. The tumour was also extending into the spinal canal to the level of C2.

An elective pre-operative tracheostomy was done for better pulmonary care. A combined surgical approach was utilised for the procedure. The intracranial portion was decompressed by a lateral suboccipital retromastoid route that had been used during the previous operations. There were significant adhesions between the tumour and the brain stem and as a result only partial tumour decompression was possible. The extracranial portion was approached through an extended incision into the neck. Here too there were significant adhesions making the dissection difficult. There was a small rent in the

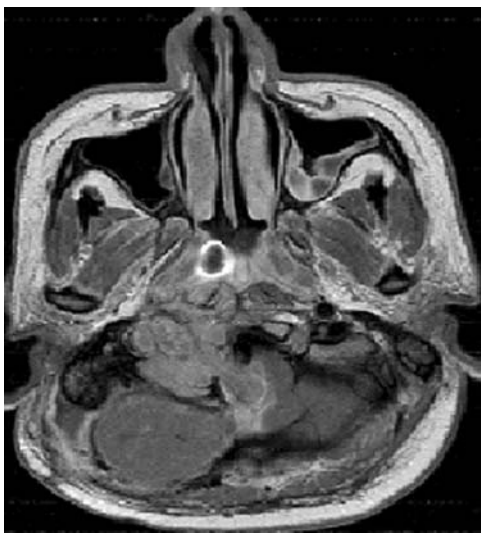

Fig. 3. Post-operative MR demonstrating residual tumour with persistent compression at cervico-medullary junction

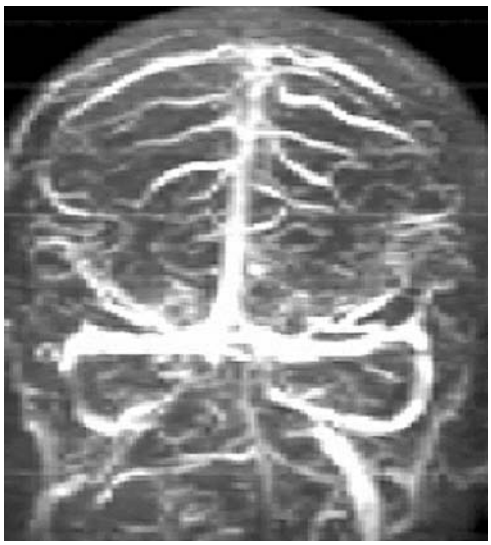

Fig. 4. Post-operative MR venogram demonstrating no flow in the right internal jugular vein

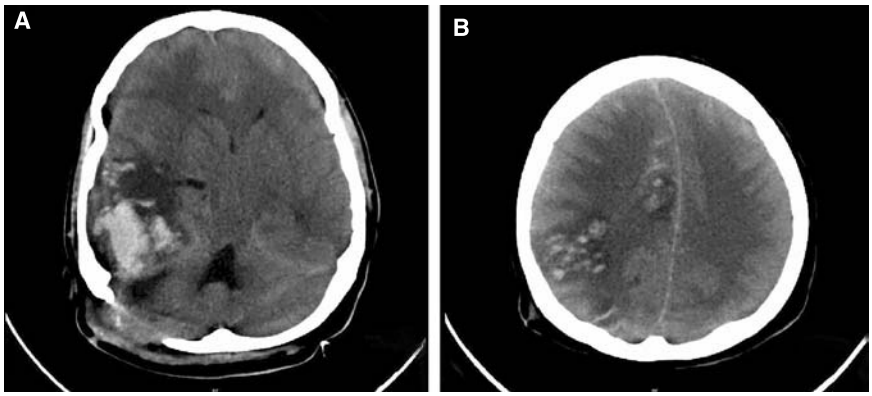

Fig. 5. Haemorrhage in the right temporal (A), parietal and frontal parasagittal regions (B)

internal jugular vein, which was repaired primarily. There was profuse bleeding from the internal carotid artery while dissecting the tumour and the artery had to be ligated. An indwelling lumbar drain was instituted for better wound healing. The patient recovered well from surgery. On the same day, she had one episode of generalised tonic clonic seizure. Computed tomography (CT) scan of the brain did not reveal any abnormality. She was continued on ventilatory support electively and gradually weaned off from the ventilator. The patient was neurologically stable at this time. She was conscious, alert, and had grade 4/5 power in all four limbs. The lumbar drain was removed on the 5th post-operative day. The CSF samples sent from the lumbar drain were sterile. On the 14th post-operative day, the patient had sudden neurological deterioration with a right total ophthalmoplegia and right hemiplegia. She developed left upper and lower limb weakness over the next half hour. She then lapsed into altered sensorium and had to be put back on the ventilator due to poor respiratory effort and altered sensorium. A repeat MRI scan at this stage showed significant tumour residue causing severe distortion and compression, of the medulla though less as compared to the pre-operative scan (Fig. 3). The MR venogram revealed absent flow in the right internal jugular vein upto the jugular fossa. The ipsilateral sigmoid, transverse and other sinuses were seen well (Fig. 4). There were no features of brain stem infarction. There was no abnormality detected in the supratentorial compartment. At this point we were unable to explain the cause of the right sided ophthalmoplegia. The significant compression of the cervico-medullary region could explain the quadriparesis as also the respiratory depression. The patient partially recovered from this event. She regained her sensorium but had persisting neurological deficits. Swelling was also noticed over the right calf. Duplex scan done revealed partial thrombosis of the right popliteal vein. Unfractionated heparin (5000 U Subcutaneously 6 hourly) was started. A repeat coagulation profile revealed a low platelet count of 76000/mm^3 and therefore heparin was discontinued. She was haemodynamically unstable requiring inotropic support for maintaining her blood pressure. She was conscious, maintaining eye contact with her left eye and was obeying commands. The power in her left upper and lower limb was grade 2/5 but grade 0 in the right upper and lower limbs. About 20 h after the previous episode, there was another episode of sudden neurological deterioration when she became unresponsive to a painful stimulus with no eye opening. Her pupils were dilated and fixed. A repeat CT scan revealed multiple areas of haemorrhagic infarction in the right temporal lobe and the right middle third parasagittal area. These were confined to the drainage areas of the vein of Labbe and vein of Trolard (Fig. 5A, B). There was severe diffuse cerebral oedema with mass effect. There were no direct features of dural sinus thrombosis such as the cord sign, delta sign or dense vein sign. The patient did not improve and expired.

Discussion

Thrombosis of the internal jugular vein (IJV) is an underdiagnosed condition that may occur as a complication of head and neck infections, surgery, central venous access and intravenous drug abuse [1, 3–5, 19, 20]. In the earlier part of this century, IJV thrombosis was a feared complication of acute oropharyngeal infection. In 1936, Lemierre described the first series of septic thrombophlebitis of the IJV [2, 15, 21]. The mortality was upto 50% at that time. Nowadays, central venous catheters are the most common underlying cause of IJV thrombosis. Recent studies found that 41–66% of the patients who had IJV catheter in place at some time had either ultrasonographic or autopsy evidence of IJV thrombosis [1, 3, 23]. Recent reported series describe IJV thrombosis rates of 25–30% following functional

neck dissection [4, 16, 20]. Doppler ultrasound is a very sensitive non-invasive investigation to detect intraluminal thrombus. Magnetic resonance angiography is the other non-invasive investigation which can be used [25]. A study by Quraishi [20] of 100 internal jugular veins by duplex scanning following functional neck dissections revealed that 25% had thrombus in the IJV on the 1st day and 26% on the 7th post-operative day. Majority underwent recanalisation with thrombus in only 5% at the end of three months. In most cases collateral venous flow will allow adequate venous drainage and prevent increased intracranial pressure [25].

The clinical features of IJV thrombosis are often very subtle, making it easy to overlook the diagnosis. Duplex scanning in these situations would help to diagnose the condition early when the patients are asymptomatic [4]. However, once infection sets in, significant objective findings may be found. It may propagate into the intracranial sinuses or veins causing increased intracranial pressure, into the superior vena cava, may cause pulmonary embolism, fever and sepsis. The patient presented here had injury to the internal jugular vein which was repaired primarily. This injury could have predisposed to thrombosis of the IJV. The patient however had a delayed deterioration after 14 days due to intracranial extension of the thrombus. In the majority of patients, this is asymptomatic. In the study by Wusternberg [25], of the 17 patients who had their IJV ligated, post-operative imaging had revealed thrombosis of the sigmoid sinus in 4 and transverse sinus in 3. All these 7 patients were asymptomatic. However, there have been few case reports of such extensions of the thrombus with associated elevations of intracranial pressure [7, 14, 22]. The clinical picture in such conditions can be varied. This can be attributed to the anatomical variations in the intracranial venous system that comprises the collateral circulation [10, 11]. In the presence of an atretic or hypoplastic transverse sinus, ipsilateral occlusion of the sigmoid sinus causes congestion of the vein of Labbe and contralateral occlusion causes circulation failure in both the hemispheres. The clinical picture varies from no symptoms to raised intracranial pressure (pseudotumour cerebri) causing headache, visual disturbances (including decreased visual acuity and diplopia) and alterations in the level of sensorium [9, 13] and development of cerebral venous thrombosis. Brain imaging may show variable findings from normal to the presence of an intracranial venous sinus thrombosis [18]. In our patient, the CT scan showed haemorrhagic infarct involving the territory of vein of Labbe and also patchy areas in the territory of the vein of Trolard. There was significant diffuse cerebral oedema with mass effect and midline shift. These findings were suggestive of cortical venous thrombosis. This radiological picture was in marked contrast to the MRI scan which was done just 24 h prior to this deterioration which did not reveal any supratentorial pathology. The MR venogram too did not reveal thrombosis of any of the dural sinuses or the cortical veins. With the repeat scan showing features of cortical vein thrombosis, the patient's right sided ophthalmoplegia could have been due to cavernous sinus thrombosis by extension through inferior petrosal sinus. The cause for this delayed progression remains unknown but retrograde spread of the thrombus from the IJV could be a probability. The possibility that the patient was having any pre-existing prothrombotic state is less likely as the previous two operations were uneventful. One possibility is of heparin induced thrombocytopenia (Type II) producing a prothrombotic state. This occurs due to the antibody binding to the platelet-heparin complex. This is independent of the amount of heparin given, the type of heparin given and the route of administration. Most of the time, it manifests 3–5 days after initiating heparin. But in patients who have been exposed to heparin previously due to prior surgery this can occur early [24] and the progression of the thrombus could have occurred in this short time. Non-infective conditions such as dehydration, anaemia with inadequate replacement of blood, leukemias and lymphomas [8], following air embolism in the sitting position [17] have been reported. These patients can be managed with anticoagulants most of the time with good recanalisation rates. However, thrombolysis can be considered in the case of rapid progression of the thrombus or in the progressively symptomatic patient [7]. In our patient thrombolytic therapy was not considered due to the sudden deterioration and poor neurological status.

This report has been presented to highlight the risks involved in skull base surgery in this region. Injury to the IJV can occur during the intervention and can be associated with a high incidence of thrombosis of the IJV. Progression of this thrombosis can occur with a disastrous outcome. Duplex scanning is a very sensitive non-invasive investigation to detect intraluminal thrombus even in asymptomatic patients. Early institution of anticoagulant therapy can prevent extension of the thrombus with good recanalisation rates. Awareness about this fact can help us to suspect this condition and deal with this condition more aggressively at a relatively early stage thereby reducing morbidity and mortality.

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Comment

This is a well-written manuscript presenting a 35-year old female who had recurrent hypoglossal schwanomma with both intracranial and extracranial components. The patient was operated on by using a combined suboccipital and a lateral cervical approach with partial removal of the tumour. Intraoperatively, vascular damage occurred, both to the jugular vein and to the internal carotid artery (ICA), both of which had to be ligated. On the 14th post-operative day the patient presented a sudden ophthalmoplegia, a right hemiplegia and a decrease in the level of consciousness. The patient finally died. The CT scan disclosed the presence of a venous hemorrhagic infarction, a diffuse hemispheric swelling and multiple hemorrhages in the parietal and frontal parasagital areas. The authors considered this presentation compatible with a thrombosis of the internal jugular vein with retrograde venous thrombosis of the Labbe vein.

I think the authors have shown courage in presenting a patient with a bad outcome due to complications directly related to surgery. The management of such a case would be difficult for any neurosurgeon. The lesson that can be learnt from this case is that postoperative monitoring of difficult patients such as the one presented here should probably be more proactive. Doppler ultrasound is a widely used non-invasive method of monitoring that can be repeated as many times as needed to diagnose subclinical jugular vein thrombosis. Ultrasound equipment has developed rapidly over the past decade and is now used routinely for assessment of arteries and veins. Duplex Doppler scanning is a powerful tool with a high sensitivity and specificity to diagnose jugular vein thrombosis.

In difficult skull-base surgery cases with potential or actual damage to the jugular vein, serial Doppler ultrasonographic examinations should probably be scheduled post-operatively. In the study of 65 patients who underwent functional neck dissections Qureshi *et al*. found a relatively high incidence of thrombosed jugular veins in the immediate post-operative period of neck dissection [1]. However, a significant number of these veins underwent spontaneous recanalization. Whether or not neurosurgical patients with a significant risk of jugular venous thrombosis should receive anticoagulants or antiplatelet therapy and the risk/benefit of such management is not known and could be a matter of considerable debate.

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