

CASE REPORT

JUGULAR FORAMEN SCHWANNOMA—A VERY RARE ENTITY

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Jugular foramen schwannoma is a very rare tumour and very few cases have been reported in the literature. Usually it is misdiagnosed as acoustic neuroma and the diagnosis is made peroperatively. It has significant neurological morbidity and mortality. In addition to the experience and expertise of the surgeon, introduction of better anaesthesia and microsurgical techniques have significantly reduced the morbidity and mortality. We present a case of jugular foramen schwannoma who was operated by the senior author. The outcome was very good and the patient recovered without any permanent deficits. The epidemiology, pathophysiology, clinical features, radiographic findings and treatment of this rare entity are reviewed.

Keywords: Jugular foramen, schwannoma, surgical options, outcome

INTRODUCTION

Schwannoma (neuroma) represents approximately 7–10% of all primary intracranial tumours. It can arise from any cranial or spinal nerve but most intracranial neuromas occur on the acoustic nerve. Rarely, the trigeminal, facial, glossopharyngeal, vagus and hypoglossal nerves are the sites of origin.^{1–3}

The majority of jugular foramen schwannomas arise from the IX cranial nerve but they may also arise from the X or XI cranial nerve. Because of this uncertainty, most authors prefer the term 'jugular foramen schwannoma'.²

CASE SUMMARY

A 45-year-old female presented with history of tinnitus, vertigo and gradually increasing right-sided deafness for the last 6 months and ataxia, headache and vomiting for 2 months. On examination, higher mental functions were intact. She had reduced hearing on right side and fundoscopy showed gross papilloedema. There were no signs of lower cranial nerve involvement.

MRI brain showed a large space occupying lesion in the right cerebellopontine angle. It was isointense to the brain both on T1 and T2 weighted images and showed homogeneous enhancement with gadolinium. There was obstructive hydrocephalus due to compression and distortion of the fourth ventricle. A preoperative diagnosis of acoustic neuroma was made.

After informed consent and preparation, the patient was operated. In the lateral (park bench) position, right suboccipital craniectomy was done and the tumour was approached. It was found to be a jugular foramen schwannoma arising from the IX–X cranial nerve complex. Under microscope, the tumour was excised completely.

Postoperatively, the patient was found to have partial lower cranial nerves palsy and had some difficulty in swallowing. Naso-gastric tube was passed and retained for about 2 weeks. It was removed when

the patient was checked and found to have normal swallowing reflex. There were no significant postoperative complications. Postoperative CT scan did not show any residual tumour or hydrocephalus. She was discharged from the hospital in a stable condition.

DISCUSSION

Incidence

Schwannomas involving the jugular foramen are very uncommon¹. Maniglia *et al.* reported a case, bringing the total in the literature at that time to 56 cases. Pluchino *et al.* and Kaye *et al.* have reported their personal experiences of a total of 25 cases.⁴ Till now only nearly 100 cases have been reported in the literature.^{2,3} It is the first case during the 30 years experience of the senior author of this article, working in the busiest centre of the country during these three decades.

Pathophysiology

Kaye *et al.* (1984) observed three main patterns of tumour growth. More distal lesions expand inferiorly from the base of the skull, whereas more proximal lesions will enlarge into the posterior fossa and the tumours arising in the mid region will tend to expand primarily into the bone.^{2,3,5,6}

Jugular foramen schwannoma becomes symptomatic either from dysfunction of the parent or neighbouring cranial nerves or from progressive distortion of the brain stem.⁷

Clinical Presentation

The clinical features of jugular foramen schwannomas closely resemble acoustic neurinoma (tinnitus, dizziness, deafness and ataxia) and there may be only minimal deficits of the jugular foramen nerves. Deafness is the most common initial presenting feature. Compression of the lower cranial nerves may cause hoarseness and swallowing difficulties. Occasionally, with large tumours, there may be involvement of the V or VII cranial nerve, along with signs of raised intracranial pressure. The

tumours growing within bone or extracranially are often misdiagnosed pre-operatively as glomus jugulare tumours, epidermoids or chordomas.^{2,3,8}

Diagnosis

The diagnosis of glossopharyngeal schwannoma is usually made once the tumour attachment to the IX nerve is seen at surgery.³

Plain radiographs and tomography may demonstrate erosion and widening of the jugular foramen with smooth distinct margins. This is in contrast to glomus jugulare tumours which typically cause enlargement of the jugular foramen with irregular indistinct margins.^{6,9,10}

On plain CT scan the tumour is hypodense or isodense with brain while it shows moderate enhancement on contrast administration and internal auditory canal is normal. The jugular foramen is enlarged in the majority of patients.^{2,3}

On MRI, the tumour is hypointense on T1 and hyperintense on T2, and shows moderate contrast enhancement. Angiography generally demonstrates an avascular or only slightly vascular mass whereas glomus jugulare tumour is usually highly vascular.^{6,10}

Treatment

Standard treatment is surgical resection and the target is total excision, keeping in view the safety of the patient.⁹ A retrosigmoid suboccipital craniotomy or craniectomy is performed. The anaesthetist has to be aware of the risk of excessive vagus nerve activity (bradycardia) and air embolism.^{2,3}

Removal of the intraforaminal portion of the jugular foramen schwannoma under direct vision requires resection of the adjacent bone which has a potential danger of CSF leakage and damage to the adjacent cranial nerves as well as venous outflow. The use of endoscope can be very useful for removal of this portion of the tumour.⁹

Stereotactic radiosurgery (with gamma knife or linear accelerator) is an option used in patients with residual or recurrent tumours, patients medically unfit for surgery, or old age. Local control rates of up to 100% have been reported but it has risk of lower cranial nerves dysfunction, although the morbidity appears lower than in surgical series.^{2,10}

Postoperative Care

In the ICU, the function of the lower cranial nerves should be carefully checked. To avoid aspiration due to dysfunction of lower cranial nerves, extubation is performed only after the patient is completely awake.

The nasogastric tube is left in place until swallowing reflex is found to be normal. Tracheostomy should be performed when the patient presents with swallowing difficulty and recurrent aspiration.¹

Outcome

Injury to the lower cranial nerves is the main cause of poor outcome. Pluchino *et al.* reported an operative mortality of 16% and their review of literature showed an overall mortality of 9%. Kaye *et al.* reported no mortality in their series and attributed this to modern improvements in diagnosis, microsurgical techniques and combined suboccipital-otological surgical approaches.^{1,4}

CONCLUSION

Jugular foramen schwannomas are very rare tumours. These may not always produce lower cranial nerve deficits, clinical presentation may mimic an acoustic neurinoma, and are diagnosed only preoperatively. Surgical morbidity and mortality are quite high but the experience and expertise of the surgeon, better anaesthesia and microsurgical techniques can contribute to a favourable outcome.

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