A Limited Approach to the Jugular Bulb: Anatomical Considerations in a Rare Jugular Foramen Tumour

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Abstract We report a case of a rare schwannoma of the jugular foramen and removed it by a limited transjugular approach. A limited surgical approach was performed for removal of the tumour, with preservation of surrounding structures. A thorough understanding of the anatomy of the region is necessary for our limited transjugular approach. The schwannoma was successfully removed. Through a detailed understanding of the anatomy of the region and adequate classification of the tumour, a limited surgical approach is not only possible, but preferable.

Keywords Jugular foramen · Schwannomas · Cranial nerve tumour

Introduction

Schwannomas of the jugular foramen are rare tumours that represent between two and four percent of all intracranial schwannomas [1]. They are Schwann cell tumours that arise from the lower cranial nerves and are of anatomical interest due to their rarity. They constitute a special entity due to their location as the jugular bulb is one of the most surgically inaccessible sites in the body.

A thorough knowledge of the anatomy and relations of the jugular foramen is essential to a successful surgical outcome. The aim of surgery is to preserve the function of the lower cranial nerves whilst completely resecting the

tumour to avoid recurrence. It is the goal of the anatomist and surgeon to plan an approach that optimally preserves the integrity of surrounding anatomical structures and prevents further neurological deficits.

With a growing trend towards endoscopic approaches in head and neck surgery; a limited approach through a small incision is of great value when faced with surgery that traditionally involves wide exposure and dissection.

Schwannomas that are located at the jugular foramen arise from the ninth, tenth or eleventh cranial nerves, the glossopharyngeal, vagus or accessory nerves, respectively. Some authors classify the hypoglossal nerve in this category due to its proximity. Schwannomas of the jugular foramen may extend intracranially within the cerebello-pontine angle anterior to the pons and into the clivus, inferiorly below the foramen magnum [2], extracranially or both intra and extracranially simultaneously. Generally, the degree of intracranial or extra-cranial extension is dependant on the point of origin of the tumour on the lower cranial nerve.

Case Report

A 63-year-old female patient attended the ENT Clinic at the University Hospital in Durban, presenting with dysphagia, dysarthria, pain in the left shoulder, weakness of the left shoulder and a dropped shoulder on the left side. The symptoms had been present for some years and had progressively become worse.

Examination: A 63-year-old Caucasian female patient was found to have IXth, Xth, XIth and XIIth nerve palsies. She had loss of sensation in the oropharynx on the left, hoarseness due to a left vocal cord palsy and the palate was deviated to the right. This is in keeping with ninth and tenth

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Fig. 1 Large jugular bulb tumour in left jugular bulb

nerve palsies. She had a dropped shoulder on the left with wasting of the sternomastoid and trapezius muscles due to involvement of the eleventh nerve. Her tongue deviated to the left on protrusion in keeping with twelfth nerve involvement.

An MRI scan was performed which demonstrated a tumour of the jugular bulb on the left side consistent with a schwannoma (Fig. 1). CT Angiogram confirmed a tumour mass whose vascularity was consistent with a schwannoma eroding the base of skull which expanded and filled the jugular foramen eroding the bar between the carotid artery and the jugular foramen (Fig. 2). It effaced the jugular vein which was compressed but not occluded. The bony margins around the foramen were sclerotic, there was no intracranial extension.

In view of the progressive deterioration of symptoms, a decision was made to remove the lesion surgically.

Operative Procedure

A post auricular incision was performed and extended for an additional 1 cm along the anterior border of the sternomastoid muscle. The mastoid bone was exposed and a mastoidectomy was performed. The sternomastoid and digastric muscles were dissected off the mastoid tip and the tip was removed. The accessory nerve was isolated and dissected out and followed into the jugular foramen, as were the internal carotid artery and the internal jugular vein. The vagus nerve was exposed between the internal carotid artery and jugular vein and followed up into the jugular bulb. The hypoglossal nerve was similarly traced superiorly. The glossopharyngeal nerve was isolated and traced superiorly to the anterior pars nervosa of the jugular bulb. The facial nerve was exposed throughout its vertical portion in the mastoid cavity. The lateral and sigmoid sinuses were then exposed all the way into the jugular foramen. The lateral sinus was then mobilised, dural flaps

Fig. 2 CT Angiogram showing tumour occupying left jugular bulb

were created on either side of the sinus in the posterior fossa and the lateral sinus clamped with large ligacclips. The jugular vein was tied off high in the neck, the jugular bulb was opened and the inferior petrosal sinus was found medially and packed with muscle internally to secure haemostasis.

The tumour was then approached through the medial wall of the jugular bulb and removed completely without damage to the nervous structures. Total operating time was under 2 h as a result of the limited approach.

Post Operative Course was Uneventful

Six months after the operation there was a marked improvement in her symptoms, there was no pain and while there are residual pareses of the nerves, there has been an improvement in speech and swallowing and evidence of return of function. Repeat scan shows no residual tumour present.

Discussion

No consensus exists on the surgical management of Jugular Foramen Tumours. This is due to their rarity and differences in presentation. The anatomy of the jugular foramen and its surrounding structures becomes highly important in planning the correct surgical approach in order to achieve maximum exposure and minimal damage to surrounding nerves.

The radiographic methods of choice were MRI and CT Angiography. The tumour showed marked enhancement but was not a vascular tumour thus eliminating the differential diagnosis of a glomus jugulare tumour.

As confirmed by Kadri and Al Mefty [1], “The choice of surgical approach is of vital importance for success. A safe exposure demands that the surgeon understand the micro-anatomy of the region to preserve the neurovascular structures.” [1].

Anatomical considerations firstly involve identifying the location of the jugular foramen which is located at the junction of the base of the temporal and occipital bones. Two compartments are identified; the posterolateral pars venosa containing the jugular bulb plus the X and XI cranial nerves and the anteromedial pars nervosa containing the inferior petrosal sinus and the IXth cranial nerve [3]. The next anatomical consideration is that the lower cranial nerves pass medially to the jugular bulb and are further displaced medially when a tumour is present in the jugular bulb. Importantly, the inferior petrosal sinus courses from the cavernous sinus and empties into the medial aspect of the jugular bulb [3]. The inferior petrosal sinus has a variable relationship with the location of the lower cranial nerves. describe the inferior petrosal sinus commonly passing between the IXth nerve anteriorly and the Xth and XIth nerves posteriorly. Surgically it is important to be aware that packing the inferior petrosal sinus or cautery can damage one of the cranial nerves in this region [4].

Schwannomas of the jugular foramen are rare tumours and classification can assist with management. Few units have large series and tumours often mimic an acoustic neuroma or a glomus tumour. Variability of presentation is common in the literature.

Kaye et al. [5], classified these tumours according to their pattern of growth. Type A was primarily intracranial. Type B mainly involved bone in the jugular foramen with or without extension and Type C were primarily extracranial with extension into the jugular foramen posterior fossa [5]. Pellet et al. [6] added in an extra Type D that were dumbbell tumours with intracranial and extracranial extension [6].

Franklin et al. [7], had a classification similar to that for glomus tumours. Tumours confined to the neck were Type A. Type B tumours were primary neck tumours with extension into the jugular foramen. Type C tumours filled the jugular foramen and were further divided according to involvement of the carotid artery, and Type D was for intracranial extension with i and i.e. referring to intradural or extradural; and 1 or 2 refer to 2 cm or less or 2 cm or more, respectively, in diameter. Mazzoni et al. [8] had a similar classification that left out Franklin Type B. Class A is tumour present in neck <2 cm in diameter; A+ is tumour present in neck but >2 cm in diameter; Class C is tumour in jugular foramen without involvement of the petrous internal carotid artery. Class C+ is tumour in the jugular foramen with involvement of the petrous internal carotid artery. Class D are intradural <2 cm; D+ are intradural tumours >2 cm in diameter [8].

With such rare tumours no consensus exists on their management.

Our case was a Kaye Type B or Franklin Type C1 or Mazzoni Class C.

The patients tumour was confined to the jugular foramen but also had all of the lower cranial nerves involved as part of a jugular foramen syndrome.

We have described a limited transjugular approach with no morbidity. This approach preserves hearing and facial nerve function and gives adequate tumour visualisation and control of vasculature and yet centres on the jugular foramen. Modern MRI techniques give an accurate definition of the tumour position and extent and allow for accurate positioning of the surgical approach. Total removal of the tumour in a single stage operation is the goal and preservation of as much nerve function as possible.

Through a detailed understanding of the anatomy of the region and adequate classification of the tumour, a limited surgical approach is not only possible, but preferable.

Summary

- Schwannomas of the jugular bulb are rare tumours that are difficult to resect due to their inaccessible position.
- Excellent knowledge of Anatomical landmarks and anatomical planning of the approach give best surgical results with optimal preservation of surrounding structures.
- A limited approach, whilst ensuring complete removal, has less morbidity and mortality than an open approach.

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