Case Report

Micro-neurosurgical excision of dumbbell shaped very large jugular foramen schwannoma

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Abstract:

Introduction: Jugular foramen tumor is a rare tumor. Jugular foramen schwannoma is rarer. This type of tumor may present in combination of different cranial nerve palsies in the form of different syndromes or may also be diagnosed incidentally. Management of such tumor is not straightforward. Case reports: Two young male presented with headache, vomiting, gait instability, right sided hearing loss. Clinically they had different cranial nerves dysfunction. Imaging showed jugular foramen tumor extending from posterior fossa to almost common carotid bifurcation at neck in both cases. Near total microsurgical excisions of the tumor were done through retrosigmoid suboccipital plus transmastoid high cervical approach with facial nerve mobilization in one case and without mobilization in other case. In case 1 combination of lower cranial nerve palsies is unique with inclusion of VIII nerve and that does not belong to any of the jugular foramen syndromes (i.e. Vernet, Collet-Sicard, Villaret, Tapia, Schmidt, and Jackson). Here IX, X, XI, XII and VIII cranial palsies was present (i.e. Collet-Sicard syndrome plus VIII nerve syndrome!). In the second case there was IX & X dysfunction with VIII dysfunction. We also went through the short review of the literature here.

Key words: Jugular foramen tumor; jugular foramen schwannoma; jugular foramen syndrome

Introduction:
The jugular foramen tumors are very rare, and the paragangliomas are the most common tumors of this region1. Schwannomas with origin in the jugular foramen are extremely rare, and there are about two hundred cases described in the literature. These represent 2.9 to 4% of the intracranial schwannomas and less than 4% of all lesions of the temporal bone2.

Jugular foramen tumors may be diagnosed incidentally. But most of the time, the patients with jugular foramen schwannomas present with headache, ataxia and visual disturbance, and combination of cranial nerves palsy. In one case combination of lower cranial nerves palsy in this case is unique with inclusion of VIII nerve and that does not belong to any of the jugular foramen syndromes (i.e. Vernet, Collet-Sicard, Villaret, Tapia, Schmidt, and

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Jackson). Here IX, X, XI, XII and VIII cranial nerve palsies were present (i.e. Collet-Sicard syndrome plus VIII syndrome!). In the other case there was IX & X dysfunction with VIII nerve dysfunction and here we closed the Jugular foramen after tumor removal with two sheets of fascia lata (one inside and another outside, connected centrally with a stitch). We also went through the short review of the literature here.

**Case report:**

**Case 1:**

A 28-year-old male presented with gradually increasing headache, vomiting, visual disturbance, vertigo and instability during walking with tendency to fall to right for one and half years. He also complained for hearing loss on right for last six months. He had no complaints regarding swallowing difficulty, nasal regurgitation, and hoarseness of voice with weakness of limbs, tongue, neck or shoulder. On examination he had hearing loss on right side. There was right sided IX, X, XI and XII nerve palsies. There was atrophy of sternomastoid muscle and trapezius muscle on right side (Figure 1A). Right sided cerebellar signs were positive with bilateral papilleodema. Limbs muscle power was normal. Long tract signs were negative. MRI of brain and neck showed isointense tumor in posterior fossa (compressing cerebellum and brain stem), jugular foramen and neck that enhanced with contrast (Figure 1B, C & D).

CT scan showed enlargement of jugular foramen with above mentioned extension of tumor. Cerebral DSA showed normal arterial, capillary and venous phase including bilateral symmetrical normal sigmoid and internal jugular vein. Right sided external carotid artery shoot showed prominent occipital and posterior auricular artery. Pure Tone Auidiometry (PTA) showed neural hearing loss on right side. The tumor was removed near totally through combined retrosigmoid suboccipital craniectomy and transmastoid high cervical jugular foramen approach with anterior-superio-medial mobilization of facial nerve, preservation of labyrinth and obliteration of middle ear cavity (Figure 2A, B, C, D, E & F).

Jugular vein kept undisturbed. All cranial nerves was tried to preserve by intracapsular removal of tumor in the neck. Jugular foramen, middle ear cavity and mastoido-petorsal areas were packed with thigh fat and mobilized vascularized posterior part of temporalis muscle. External auditory meatus was obliterated. Post operatively patient recovered.
uneventfully except mild paresis of right sided facial nerve (Figure 3) that recovered by twelve weeks.

Case 2:
A 32-year-old male presented with gradually increasing headache, vomiting, visual disturbance, vertigo, gradual right sided hearing loss and instability during walking with tendency to fall to right for one year. He had no complaints regarding swallowing difficulty, nasal regurgitation, hoarseness of voice and weakness of limbs, tongue, neck or shoulder. On examination he had hearing loss on right side. There was uvular deviation to left with less movement on the right side of palate. Right sided cerebellar signs were positive with bilateral papilleodema. Limbs muscle power

Histopathology confirmed jugular foramen schwannoma. MRI of brain and neck on three months after operation showed small residual tumor at posterior fossa and fat in jugular foramen, middle ear cavity and neck (Figure 4 A, B, C& D).


Figure 3: (After having written permission of patient to publish his photographs). Post operative picture of patient. A: post operative right sided facial paresis. B: incision line after stitches removal.

Figure 4: Post operative MRI (case 1) of brain and neck. A&B: T2W axial images, C: T1W sagittal and D: T2W coronal images showing no residual tumor in posterior fossa with fat graft at jugular foramen, middle ear cavity and tumor dead space at neck.
was normal. Long tract signs were negative. MRI of brain and neck showed isointense tumor in posterior fossa (compressing cerebellum and brain stem), jugular foramen and neck that enhanced with contrast (Figure 5 A, B & C). CT scan showed enlargement of jugular foramen with above mentioned extension of tumor. Cerebral DSA was not done. Pure Tone Audiometry (PTA) showed neural hearing loss on right side. The tumor was removed near totally through combined retrosigmoid suboccipital craniectomy and transmastoid high cervical jugular foramen approach without mobilization of facial nerve with preservation of labirynth and middle ear cavity. External ear canal and internal jugular vein kept undisturbed. All cranial nerves was tried to preserve by intracapsular removal of tumor in neck. Jugular foramen was closed by putting two sheets of fascia lata; one intracranially and other extracranially. A stitch was given connecting the center of both sheets to prevent migration. Then Jugular foramen, mastoido-petorsal areas and tumor dead space at neck were packed with thigh fat and mobilized vascularized posterior part of temporalis muscle. Post operatively patient mild dysphgia that recovered within four weeks. Histopathology confirmed jugular foramen schwannoma. CT scan of brain and neck three weeks after operation showed no residual tumor at posterior fossa and jugular foramen (Figure 5 C, D & F).

Discussion:
The fifth decade of life is mostly affected. The jugular foramen lesions are very rare, the paragangliomas are the most common tumors in this region, followed by schwannomas, meningiomas, metastases, giant cells tumors, chondrosarcomas, plasmocytomas, epidermoid cysts, amyloidomas and endolymphatic sac tumors that are also described in this location with primary and expansive lesions. The schwannomas arising from the glossopharyngeal nerve are most common in this region, followed by the vagus, accessory, and, more rarely, hypoglossal nerve tumors. Jugular foramen important neurovascular structures, cranial base, upper cervical region and areas of the posterior cavity may be involved by these lesions.

The tumors at jugular foramen area are classified by Kaye et al into three types. Type A regards the intracranial origin lesions, with a minimal osseous extension. Type B lesions are the primary ones of the bone with or without intracranial component. Type C lesions are primarily extra-cranial with minimal extension for cranial bone or posterior cavity. Pellet et al added the so-called type D, which regards the hour-glass-shaped tumor that involves the jugular foramen, with intracranial and extra-cranial components. These patients are in Type D. The schwannomas are usually solid, circumscribed and well-delimited, and may cause cystic degeneration or primary cysts.

The clinical presentation of the jugular foramen

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**Figure 5:** Preoperative contrast MRI of brain and neck (case 1); A: saggital, B-axial and C: coronal images showing right sided jugular foramen tumor extending posterior fossa to neck through jugular foramen. D & E: post operative CT scan showing no residual tumor at posterior fossa and jugular foramen. F: Bone window showing extent of bone removal.
tumors depend on their site of origin. The tumor origin, size and site may be responsible for delayed presentation\textsuperscript{1, 6}. The early symptoms of tumors originated in the jugular foramen are, in most cases, conductive hearing loss and tinnitus. Other lesions may cause paralysis of the lower cranial pairs as a first symptom\textsuperscript{4}. The tumors that expand to the posterior fossa usually manifest primarily with deafness, vertigo and ataxia that may mimic the clinical presentation of tumor originating from vestibular-cochlear nerve. For the jugular foramen lesions that expand to extra-cranial proportions, may present with hoarseness, diminishing of the coughing reflex and reduction of the motor strength of the shoulder girdle, symptoms representing the typical jugular foramen syndrome\textsuperscript{1, 2, 5, 8}. It is believed that the diagnosis and nerve origin can be determined by the initial symptoms and the grade of dysfunction of the nerve, especially when compared with adjacent nerves. This connection gives rise to the classic descriptions of the different syndromes affecting the lower cranial nerves\textsuperscript{2-5, 8, 9} including descriptions by Vernet (involvement of the ninth, 10th, and 11th cranial nerves), Schmidt (the 10th and 11th nerve), Collet-Sicard (9th, 10th, 11th and 12th nerves), Jackson (the 10th, 11th, and 12th nerves), and Tapia (the 10th and 11th, 12th nerves and sympathetic), Villeret (9th, 10th, 12th and sympathetic) as well as a combination of these syndromes. Cases in the study can be named as Collet-Sicard plus VIII nerve syndrome!

Rather than corresponding to the nerve of origin, however, the symptoms are marked by their variability in presentation, and some patients have no dysfunction of the lower cranial nerves. Symptoms secondary to increased cranial pressure (blurred vision and headaches) are actually the most common complaints, and the most common and earliest sign is compromise of the eighth cranial nerve complex. This is what makes the differential diagnosis of schwannomas and acoustic tumors difficult\textsuperscript{10}. The presence of nystagmus, ataxia, vertigo, pyramidal tract involvement, and other cranial nerve deficits (fifth, sixth, and seventh nerves) has been reported\textsuperscript{8, 11-21}. Hakuba, et al.\textsuperscript{9} divided the clinical symptoms into four categories: cranial nerve involvement, unilateral cerebellar involvement, nystagmus, and signs or symptoms secondary to intracranial pressure.

Neuroimaging indicates the tumor extension, as well as help differentiate between the schwannomas and other jugular foramen tumors, such as meningiomas and paragangliomas. On CT scan there may be enlargement of the jugular foramen with well defined osseous margins. The scan without contrast describes hypo or isodense tumor with a reduced capturing of contrast, features which differ it from meningiomas, that present an important contrast capturing, as well as sclerosis and erosion of the subjacent bone\textsuperscript{6}. On MRI images there is hyperintensity in T2 and hypointensity in T1, with a strong enhancement after gadolinium administration. It may present as a heterogeneous tumor due to presence of necrosis or cystic formations\textsuperscript{6, 8, 22}. Cerebral digital subtraction angiography shows moderate vascularization that differ the schwannomas from the paragangliomas of such region, which are intensely vascularized\textsuperscript{5}.

Successful treatment of lesions involving the jugular foramen includes complete tumor removal without creating additional neurological deficits. Although in early surgical series\textsuperscript{23} a mortality rate as high as 16% had been reported, recent advances in surgical microneuroanatomy, neuroradiology, and microsurgical and skull base techniques have drastically decreased this rate, and no deaths directly related to the surgical
procedure have been reported in the latest series\textsuperscript{9, 11-17, 20-25}. The lower cranial nerves must be carefully evaluated, because these are the source of major and life-threatening postoperative complications, especially if the patient had no deficits preoperatively. The acute development of postoperative deficits in such patients allows no chance for compensatory mechanisms to develop\textsuperscript{25}. Speech pathology and otolaryngological evaluations with pre and postoperative swallowing, as well as audiological studies, are essential in the treatment of these patients. Appropriate measures, such as the nothing-by-mouth regimen with parenteral nutrition, swallowing exercises, and soft mechanical diets with swallowing precautions, are taken if the patient exhibits a risk of aspiration on postoperative swallowing studies. Postoperative deficits might improve, but if there is risk of aspiration persists or the patient has marked dysphonia, vocal cord medialization is advised.

Complete surgical resection is still a challenge, in spite of the development of new advanced operative microsurgical techniques applied to the cranial base. The tumor extension to the cavernous sinus, clivus or brain stem is sometime a limiting factor to the total removal of the lesion. Multidisciplinary approach is sometime needed where neurosurgeons, otorhinolaryngologists and sometimes neuroradiologists work together for a better diagnostic and pre-operative assessment, in addition to a better management of these patients\textsuperscript{1, 3, 4}. Radical removal of benign jugular foramen tumors is the treatment of choice, may be curative, and is achieved with low mortality and morbidity rates. Larger lesions can be radically excised in one surgical procedure by using a multidisciplinary approach. Reconstruction of the skull base with vascularized myofascial flaps reduces postoperative CSF leaks. Postoperative lower cranial nerves deficits are the most dangerous complications\textsuperscript{26}.

The surgical approach should be planned to achieve complete resection, as repeated operation drastically increases the chance of injury to the lower cranial nerves. Intra-operative electrophysiological monitoring of these nerves as well as brainstem auditory evoked potentials are important adjuvants for preventing injury to the lower cranial nerves, the facial nerve, and the vestibule-cochlear complex. The choice of surgical approach is of vital importance for success. A safe exposure demands that the surgeon understand the microanatomy of the region to preserve the neurovascular structures. The tumor’s location and extension defines the surgical approach. For tumors confined intracranially (Type A), a retrosigmoid approach provides adequate exposure\textsuperscript{25}. For tumors involving the bone window, a wider exposure is required.

The choice of the surgical approach implies the surgeon knowledge and understanding of the micro-anatomy of the region to preserve as much as possible the neurovascular structures. Katsuta et al.\textsuperscript{27} split up the JF region in three portions: one nervous (intra-jugular) and two venous; the cranial nerves IXth, Xth and XIth are in the nervous portion localized between the two venous. Finally, it should be considered that there are many anatomical variations in the course of neurovascular structures in this region and that they do not always respect and conform to the compartmentalization into different portions (pars nervosa and pars venosa)\textsuperscript{28}. The complexity and the large range of anatomical variations of such region require careful clinical and neurological examination should be realized; CT, MRI and MRA are highly recommended. In cases of suspected much vascularized lesions, digital subtraction
angiography should be done to explore the vascular network and contemplate embolization. Sometimes, internal carotid artery is involved and invaded so that a balloon occlusion test is useful. Even if many and different approaches exist to manage such lesions either with intracranial and/or extra-cranial extensions, some believes that the juxtacondylar approach provides an optimal and safe exposure to the Jugular foramen (JF) with a limited drilling of petrous bone and mastoid thus significantly reducing the risk of auditory loss, facial nerve palsy and CSF leak. Actually, this approach provides an inferior and posterior access to the JF being mainly based on the vertebral artery control so giving also the chance of dividing the tumour blood supply.

The tumour location and extension generally defines the best surgical approach; for tumours confined intra-cranially (type I), a retrosigmoid approach may provide an adequate exposure; for foraminal and extra-cranial tumours, a more extensive exposure is necessary but preferably the labyrinth should not be sacrificed as hearing improvements have been already reported in many occasions. Kadri and Al-Mefty describing a series of six dumbbell jugular foramen schwannomas reported that the tumor was completely resected in all patients with no additional cranial nerve deficits and that with a mean follow up of 32.8 months, two of the six patients with pre-operative IXth and Xth cranial nerve palsy attained significant improvement and two of four patients with pre-operative hypoglossal nerve palsy completely recovered mobility of the tongue; in the same series, one of the two patients with hearing loss had significant improvement of that deficit, and a patient with facial palsy completely recovered nerve function. A patient with a sixth cranial nerve deficit completely recovered ocular motion. A study reporting on a series of 16 JF schwannomas of whom five were type IE described that in the immediate post-operative period, a temporary cranial nerve morbidity rate was 38% three patients experiencing facial palsy, one abducent palsy, one swallowing problems and one tongue weakness as new deficits; in the other patients, the cranial nerve dysfunction remained as it was pre-operatively and with a mean follow-up of 22 months, all cranial nerve dysfunction improved considerably. Another series reported a total resection in six of 12 patients, a near total in three and a subtotal in three with a tumor progression in two of the three patients having sub-total excision. In one patient, a tumor re-growth was recorded 12 months after surgery and had radiosurgery; in another re-growth, it occurred within 8 months and a malignant peripheral nerve sheath tumour was identified. No tumor recurrence was observed in totally and near totally resected patients during a mean 27-month follow-up period.

The experience with radiosurgical treatment of jugular foramen schwannomas is still small. In the largest reported series, concluded that this treatment should be reserved for patients with small tumors and intact lower cranial nerves.

In our second case we did not go for extensive facial nerve mobilization, external or middle ear disruption as tumor itself gave the space between the sigmoid sinus and facial nerve for removal of tumor below the jugular foramen and rest of the tumor removed at neck below the facial nerve. For effective closure of jugular foramen and tumor dead space we close the jugular foramen by two sheets of facial lata and fat in case 2.

References:
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