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REVIEW

An overview of definition and approaches to jugular fossa tumors

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Abstract
Jugular foramen has a complex anatomy composed by occipital and temporal bone. It is divided in two segments: (1) Pars nervousa (2) Pars vascularis. Jugular foramen tumors are the rare group of brain lesions which their management and surgical approaches are the challenging debate in skull base neurosurgery. Although the most prevalent tumor arising in this area is paraganglioma, the range of tumor diversity can be seen this region. Surgery of jugular foramen tumors is a complicated procedure due to the complex regional anatomy and presence of crucial structures. We explain the jugular fossa lesions classification, their related syndromes and detailed aspects of available surgical techniques at this paper. The main aim of the surgery is maximal safe resection along with preserving the highest level of neurological function. Multidisciplinary approach that involves teamwork cooperation of neurosurgeons, otorhinolaryngologist and endovascular interventionist is the novel concept in surgical planning can improve surgical outcome and increase patient consent. The main aim of this manuscript is to explain a variety of approaches for successful resection of the tumors in this complex anatomical region.

Keywords Jugular fossa tumors · Retrosigmoid approach · Suboccipital approach · Post auricular transtemporal approach

Introduction

Jugular foramen is a large skull base foramen which is located in posterior cranial fossa. It is characterized by its complex anatomy composed by occipital and temporal bone. It has been divided by a fibrous ligament or sometimes a bony septum called jugular spine in two segments: (1) Pars nervousa (2) Pars vascularis. Pars nervousa is the anteromedial part contains inferior petrosal sinus, glossopharyngeal nerve and its tympanic branch, Jacobson nerve. The greater posterolateral part, pars vascularis consist of jugular bulb and inferior jugular vein, vagus nerve and its auricular nerve, Arnold branch and accessory nerve [1, 2]. Jugular foramen tumors are the rare group of brain lesions which their management and surgical approaches are the challenging debate in skull base neurosurgery. At this manuscript we spend on the jugular fossa lesions classification, their related syndromes and detailed aspects of available surgical techniques.

Jugular foramen lesions are asymptomatic in primary stages but they can represent various symptoms based on involved neural or vascular components until they become enlarged. The common symptoms includes:

- Loss of taste from the posterior one-third of the tongue.
- Glossopharyngeal nerve palsy.
- Vocal cord paralysis and absent gag reflex.
- Vagus nerve palsy.
- Weakness of the trapezius and sternocleidomastoid muscles.
- Accessory nerve palsy.

The most common symptoms tend to arise from expansion to middle ear that cause:

- Conductive hearing loss.
- Pulsatile tinnitus due to tumor vascularity.
- Vertigo and sensorineural hearing loss as a result of involvement of the inner ear.

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Symptoms of raised intracranial compression can develop if there is significant intracranial extension of the underlying tumor [3].

The main goal of this paper is to elucidate different surgical techniques for efficacious resection of the tumors in this intricate anatomical region. For this propose we collected the data from Pub Med, Medline data bases.

Types of tumors

The range of tumor diversity can be seen in region of jugular foramen although the most prevalent tumor arising in this area is paraganglioma. Schwannoma and meningioma are the next culprits of this regional neoplasms. Most of the masses are intrinsic and arise from the structures that travers the jugular fossa like paraganglioma, schwannoma, meningiomas, primitive neuroectodermal tumor, metastatic malignant tumors or lymphoma while extrinsic masses arise from surrounding tissues around the jugular fossa and extend in to the foramen. Meningiomas and glial cell tumors are the samples of these tumors [4, 5].

Jugular foramen paragangliomas are often benign and slow growing tumors originated from glomus cells of neural crest located along temporal bone in the jugular bulb and middle ear. They are the most prevalent tumor of this region. These lesions are the rare type of brain tumor encompass about 1 in 1 million people per year and women are more susceptible to be affected. Chemodectomas is the other name considered for these tumors. They also have been divided in two sub types: Sympathetic and parasympathetic. Most of the glomus jugular tumors are parasympathetic type. Although most of them are slow growing and malignant forms are rare, they can cause considerable complications due to compression effect and local structure destruction. On MR Scans they appear hypointense onT1 and isointense on T2 WI with specific characteristic called salt and paper [6].

Jugular foramen is mostly a common site for development of nerve sheath tumors like schwannomas which are responsible for about 10 − 30% of tumors represented in this area. Other pathologic lesions like neurofibroma, meningioma, and metastatic tumors have lower incidence rate in general population [4].

Method

In this paper we aim to overview the available approaches to the jugular foramen lesions and propound conceptual surgical techniques to help maximal safe tumor resection of this complex anatomic site. In general, the approaches can be divided in to: Anterolateral and posterolateral perspectives [7].

Posterolateral approaches

They are accessed via the dissection of structures behind and lateral to the sigmoid sinus and jugular foramen and include:

Retrosigmoid approach

Retrosigmoid is the most common and preferable approach for posterior fossa surgery among neurosurgeons. It gives a good corridor for tackling with various types of lesion located in posterior fossa especially for cerebellopontine angle. Also it can be used for jugular foramen tumors which are intradural and have no extension to extradural compartments. During procedure the skin is incised in C-shaped format posterior to the auricle. Suboccipital craniotomy or craniectomy is done, then dura is also opened C or U shaped inferior and posterior to the transvers and sigmoid sinuses. The cerebellum is retracted blandly from its posterior surface. The lateral aspect of brain stem and intracranial parts of lower cranial nerves through jugular foramen and CN XI-VIII existing through IAC will appear [8, 10] (Figs. 1 and 2).

Far-Lateral approach

The far lateral approach is a modified form of suboccipital approach defined as an inferolateral extension of this technique. It has been designed for surgical resection of lesions located in ventral aspect brain stem between lower third of clivus and superior to the body of C2. Lateral, sitting, three-quarter prone, supine and prone position can be applied for surgery based on surgeon preference. Skin incision may be reverse hockey stick which starts 5 cm below inion and continued to 5 cm below mastoid tip or C-shaped starts 4 cm above auricle curves 4 cm behind mastoid and terminates anteriorly to the border of sternocliodomastoid muscle. Drilling the posterolateral part of foramen magnum and C1 hemilaminectomy enhance surgical corridor view, diminish the demand for retraction and give a great arterial access to vertebral artery, vertebrobasilar junction and posterior inferior cerebellar artery. The muscles attached to the occipital bone shaved inferiorly. Posterior hemiarch of C1 and distal occipital bone inferior to transvers and sigmoid sinuses removed through by craniotome, rongeur and Kerrison punch. Protection of the vertebral artery is one the main concern during this procedure, reflecting the vertebral artery inferiorly after opening the C1 foramen transversarium can prevent damaging to vertebral artery [9, 10].

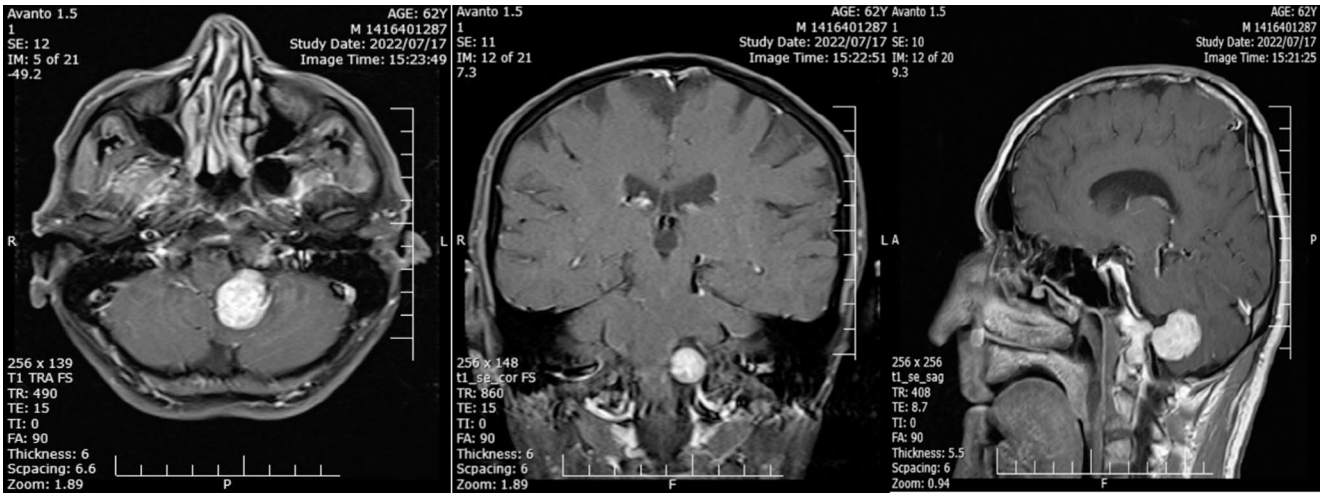

Fig. 1 Pre op MRI of left solitary fibrous tumor of jugular foramen resected by retrosigmoid approach

Fig. 2 Post op MRI of left solitary fibrous tumor of jugular foramen resected by retrosigmoid approach

Transcondylar

In this way the jugular foramen opened by partial resection of occipital condyle and used when tumors extend in to occipital condyle. Superior facet of C1 and parts of occipital condyle below the hypoglossal canal may be removed [9].

Supracondylar

This approach is used for access to the jugular tubercle by drilling the occipital bone above and behind occipital condyle and condylar fossa [9].

Paracondylar

Paracondylar approach used for lesions enfolded posterior portion of jugular foramen like paraganglioma or schwannomas of lower cranial nerves. In this method the hypoglossal canal opened and lateral portion of occipital condyle and mastoid tip partially drilled [9].

Anterolateral approaches

These approaches embrace the dissection of the structures located in front and lateral to the sigmoid sinus and jugular foramen and include:

Post auricular transtemporal approach

This approach can provide an excellent exposure to the jugular foramen and lateral skull base.It is accessed from lateral approach through the mastoid and the neck, in the other hand mastoidectomy and neck dissection are the main component of this approach. At the first the retroauricular skin incised in the C-shaped manner then sternocleiodomastoid muscle and posterior belly of digasrtric muscle retracted and neck dissection continued till internal and external carotid artery, internal jugular vein and lower cranial nerves become appeared. Mastoidectomy involves the infralabyrinthine region with fully exposure to sigmoid sinus, jugular bulb and mastoid portion of the facial nerve, for this purpose the styloid process is resected, mastoid part of facial nerve moved anteriorly which may lead to nerve palsy and rectus capitis lateralis muscle dissected from jugular process, finally the Trautman triangle (the triangular shaped dura matter on the posterior side of temporal bone which consists from jugular bulb inferiorly, posterior semicircular canal anteriorly and superior petrosal sinus superiorly) will be exposed and accessing to the jugular foramen obtained [7, 10, 11, 15].

Preauricular subtemporal-infratemporal approach

This approach exposes the jugular foramen anteriorly and it may be suitable for selected tumors that extend along the petrous portion of the internal carotid artery through the eustachian tube or the cancellous portion of the petrous apex. A periauricular skin incision continues till zygomatic process. The temporalis muscle is reflected inferiorly follow that frontotemporal craniectomy with or without superolateral orbital rim preservation will be done. The middle cranial fossa floor removed until accessing to the carotid canal be achieved. Kawase triangle is drilled for resection of petrosal apex and reaching to the posterior fossa [7, 10, 15].

Combined approach

The main purpose of combined approach is total exposure of the jugular foramen in a single stage surgery. This approach allows radical resection of tumors around the jugular foramen, the lower clivus and the high cervical region, generally as a result of combination of transmastoid, infralabyrinthin, transjugular and transcondylar transtubercular high cervical approach [7, 10].

Result

Choosing the appropriate surgical approaches to the jugular foramen lesions depend on the location, size and background pathology. The main applicable approaches categorized as lateral, posterior and combined. The privilege of each method correlates with tumor characteristics. For instance, infratemporal method of lateral approach used for large tumors like glomus jugular and schwannomas which provides an extensive corridor but requires facial nerve rerouting. Posterior approaches which applied for lesions extended to the posterior fossa, divided to Retrosigmoid and Far-Lateral approaches. Retrosigmoid approach is suitable for tumors with intracranial extension that needs minimal bone drilling and preserves hearing and lower cranial nerves function. Far-Lateral approach used for meningioma, schwannoma or other tumors extended to lateral medulla and vertebral artery. Although these approaches have higher risk for CSF (Cerebro spinal fluid) leakage. The combination of available approaches can obtain better access to jugular fossa and neurovascular preservation. Among these surgical techniques, surgeon experience plays the most important role for achieving the best surgical accomplishments.

Discussion

The cardinal concerns about choosing the best surgical techniques in jugular tumor surgery are facial nerve preservation and mobilization. CN IX-XI protection, vascular management of internal jugular vein (IJV) and carotid artery and finally proper skull base reconstruction for prevention of cerebrospinal fluid leakage. Facial nerve is often under the compression of the jugular foramen lesions particularly in paragangliomas and schwannomas, Intraoperative neuromonitoring with electromyography, translocation technique include rerouting of the facial nerve and neural grafting (using sural or greater auricular nerve) are the technical samples for preserving neurological function. Jugular foramen tumors which encase lower cranial nerves often caused dysphagia and voice hoarseness, surgeons can apply

fine surgical strategies like sharp and subcapsular dissection for avoiding direct nerve injury or preventive tracheostomy or gastrostomy in high risk patients with severe CN IX-XI involvement. Despite the fact that vascular injury is a probable complication specially in case of large tumors encase IJV and sigmoid sinus, pre op and peri op vascular intervention can be a technical principle for safety resection include preoperative vascular embolization especially in paragangliomas for reducing intraoperative hemorrhage, selective internal jugular vein ligation in case of patent contralateral side and intraoperative doppler for continuous vascular monitoring and prevent complications like jugular bulb thrombosis [12, 13]. Resection of the large tumors and petrosectomy approaches almost always create skull base defect which require rubust duraplasty and applying the reconstruction methods like using fat graft, fibrin sealant for small defect or utilization of the vascularized flap (temporalis fascia, nasoseptal flap or pericranial flap) in case of larger defect. Although for high risk patients with rising in intracranial pressure (ICP), lumbar drain or intermittent lumbar puncture can be used for reducing the CSF pressure [13, 14, 16].

Conclusion

Although surgery of jugular foramen tumors is a challenging problem among neurosurgeon society, paying attention to the tumor characteristics and neurovascular involvement, cost -benefit assessment between extent of resection and resultant morbidity are the key factors for attaining the best result. For example preoperative vascular embolization in case of paragangliomas or subtotal resection followed by stereotactic radiosurgery are the preferable strategies for reducing the surgical complications. Among approaches that have been explained above suboccipital rettrosigmoid, transcondylar, supracondylar, approaches are preferred for tumors with intracranial extension [17]. We conclude that the approaches mentioned above are useful in efficacious resection of jugular fossa tumors. These approaches would have better clinical outcome if combined with multidisciplinary approach that involves teamwork cooperation of neurosurgeons, otorhinolaryngologist and endovascular interventionist. This notion is a novel concept in surgical planning that can improve surgical outcome and increase patient consent.

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