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TUMORS OF THE JUGULAR FORAMEN

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DEFINITION

Jugular foramen tumors are one of the biggest challenges in skull base surgery due to the importance of the structures that pass through the jugular foramen and the adjacent structures. The jugular foramen is formed anteriorly by the petrous portion of the temporal bone and posteriorly by the occipital bone. The glossopharyngeal (IX), vagus (X) and accessory (XI) nerves are located anteriorly and the jugular bulb (junction between the sigmoid sinus and the internal jugular vein) - responsible for most of the blood drainage of the central nervous system - is located posteriorly (Fig. 1). Anterior to the jugular foramen is the internal carotid artery, the facial nerve (VII) laterally, the hypoglossal nerve (XII) medially and inferiorly the vertebral [artery1]. All these important structures, plus the fact that the jugular foramen varies in size and shape according to each skull, make the foramen a region of difficult surgical access and complex anatomy. Tumors are the pathologies that most often affect the jugular foramen, with paragangliomas being the most common, accounting for 60-80% of cases, followed by schwannomas and meningiomas[2,3]. Other tumors can also affect the jugular foramen, but there are few cases reported in the literature: squamous cell carcinoma, plasmacytoma or multiple myeloma, chondrosarcoma, neurofibroma, chordoma, fibrosarcoma and [hemangiopericytoma4].

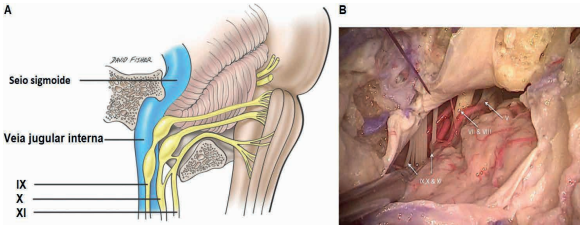

Figure 1: A - Anterolateral section of the jugular foramen. B - Anatomical relationships of the nerves of the jugular foramen, internal auditory canal, vertebral artery and its branches. A - Gutierrez Santiago, et al. Lower cranial nerve syndromes: a review. Neurosirgial Review. 2020. B - Constanzo F, Coelho Neto M, Nogueira GF, Ramina R.Front Surg. 2020 May 15.

Paragangliomas can also be called glomus tumors and originate from the paraganglia or glomus bodies, which are normally occurring structures of the temporal bone. A paraganglion is defined as a group of non-neuronal cells derived from the neural crest. This tissue has neuroendocrine, non-chromaffin characteristics and resembles the cells of the adrenal medullary layer as it has the same embryonic origin. In the skull, the glomus bodies are located, more specifically, in the adventitial layer of the jugular bulb, in the bony wall of the tympanic canaliculus, in the promontory of the middle ear, in the ciliary ganglion, in the auricular ramus and in the auricular ramus. ciliary ganglion, in the auricular branch of the vagus nerve (Arnold's nerve), in the glossopharyngeal nerve and its tympanic branch (Jacobson's nerve) and in the walls of the great arteries.

Schwannomas are tumors originating from Schwann cells and commonly originate from the glossopharyngeal, vagus or accessory nerve. Characterized by being rare, jugular foramen schwannomas represent 2.9 to 4% of cranial schwannomas and are responsible for only 4% of temporal bone lesions, being the second most common jugular foramen [neoplasm5]. The Kaye *et al.* classification is used

to stratify the lesion, so four groups are proposed: A - lesions of intracranial origin with expansion of the jugular foramen; B - tumors originating in the jugular foramen; C - tumors of extracranial origin with involvement of the jugular foramen or posterior fossa; D - "hourglass" tumors, with involvement of the jugular foramen and extra and intracranial components2,[5].

Primary jugular foramen meningiomas are extremely rare and there are few case reports in the [literature6]. They originate from meningothelial cells and the genetic components of tumor development and transformation are not yet fully understood. This tumor, although rare in the jugular foramen, is considered the most common primary neoplasm of the central nervous [system7]. In addition, there is an association between meningiomas and [neurofibromatosis8].

NATURAL HISTORY OF THE DISEASE

The natural history of jugular foramen tumors depends on the histological classification of the neoplasms. Initial symptoms usually occur due to compression of structures in or near the foramen.

Pulsatile tinnitus, of venous and primary origin, is the most common symptom of glomus [tumors9]. Aquino's sign - interruption of tinnitus after compression of the internal carotid artery ipsilateral to the tumor - may be present in these cases. At otoscopy, it is possible to find a bulging behind the tympanic membrane (Fig. 2). Other symptoms can be found as the disease progresses, such as: reduced hearing acuity, hoarseness, paralysis of the cranial pairs (X, IX, XII and XI), dysphasia, facial paralysis and, in rarer cases, a mass in the cervical region and atrophy of the [tongue10]. The neoplasm is highly vascularized and has benign histological characteristics, but invasion of bone tissue, cranial nerves, blood vessels and the dura mater adjacent to the tumor can occur if the disease is left [untreated11]. Although most paragangliomas behave like benign tumours, around 4% have become malignant with metastasis, usually to the cervical lymph nodes, liver, lungs, spleen and bones4,[12].

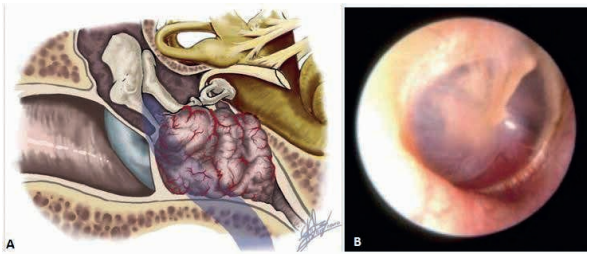

Figure 2: A- Anatomical relationships of a jugular foramen paraganglioma tumor. B- Otoscopy showing a tumor in the right lower field. Nery Breno, et al. Jugular Foramen Paragangliomas. IntechOpen: Brain and Spinal Tumors. 2019.

Schawannomas are benign, encapsulated, slow-growing lesions with a smooth surface and well-defined borders. Unlike paragangliomas and meningiomas, there is little involvement of skull base structures, as schwannomas do not infiltrate adjacent bone structures. As these tumors can originate from different cranial nerves (XII, XI, X, IX and the sympathetic chain), symptoms can vary. The main clinical manifestations are decreased hearing acuity, tinnitus, ataxia and vertigo. Depending on the stage of the disease, dysphasia, hoarseness, dysphonia, shoulder muscle weakness and tongue atrophy may be found. Some patients have had headaches, blurred vision and nystagmus due to increased intracranial [pressure13]. Classic neurological syndromes may be present in these patients, such as: Vernet's Syndrome; Collet-Sicard's Syndrome; Villaret's Syndrome; Jackson's Syndrome (Tab. 1). The literature on the natural history of this neoplasm is scarce, so vestibular schwannomas, which are much more common, end up being

the parameter for studying the evolution of jugular foramen schwannomas. It has been shown that the majority of schwannomas have an average growth rate of 1mm/year. This growth is reduced in elderly patients and the spontaneous regression of this neoplasm has already been reported in the literature[10,14].

Neurological syndromes and the respective nerves affected	Vernet	Collet-Sicard	Vilaret	Jackson
IX	+	+	+	-
X	+	+	+	+
XI	+	+	+	+
XII	-	+	+	+
Sympathetic fibers	-	-	+	-

Table 1: Syndromes related to the jugular foramen

In terms of the natural history of tumors in the topography of the jugular foramen, meningiomas present symptoms earlier than paragangliomas. The most common symptoms are: hearing loss, a mass in the middle ear, dysphasia, tinnitus, ataxia, cranial nerve palsy, headache, and in more advanced cases, hydrocephalus and hemiparesis are relevant findings[10]. Meningiomas are sometimes incidental findings on imaging tests. In these cases, tumor growth was slow and most patients remained asymptomatic during follow-up. Male patients, as well as those with a tumor larger than 25mm in diameter, hyperintense signal on MRI T2 sequences, symptomatic patients or those with edema were more likely to have fast-growing tumors[15].

Among the less common neoplasms, chordomas and chondrosarcomas stand out. Their common manifestation is headache (due to paralysis of the 6th nerve) and diplopia, while other symptoms such as numbness in the topography of the 5th nerve, dysphagia, hoarseness, lingual paralysis and tinnitus may also be present[10]. It is worth noting that dysfunction of multiple cranial nerves is more common in chondrosarcomas, but the prognosis of patients is better compared to chordoma. Chordomas have a survival rate of approximately 18 months, with post-treatment recurrence in 35 to 70% of cases[16].

TREATMENT

Paragangliomas can be treated surgically, with radiotherapy, radiosurgery or even expectantly, depending on the characteristics of the tumor and the patient's preferences. It's worth noting that surgery is currently the only approach that can be used. promotes the cure of the disease. In this approach, the main focus is on preserving the patient's neurological capacity at the same time as total tumor resection is desired. Surgical mortality is low and is directly linked to the size of the tumor, and tumor recurrence rates are low, ranging from 0.7 to 11.8%[10]. For older patients or those with small tumors, treatment with radiosurgery may be feasible, however, this method does not promote a cure. Radiotherapy can also be used as an adjuvant when total resection is not possible[17].

Since schwannomas are benign lesions that do not infiltrate structures, they have a high cure rate with treatment. Among the options are surgical resection, clinical observation or radiation modalities. As with glomus tumors, surgical treatment is curative, especially in small volume schwannomas[13]. Neurological deficits associated with surgical complications are directly linked to the size of the tumor. The indications and benefits of treatment with radiosurgery and radiotherapy for schwannomas are similar to those for paragangliomas.

Radical resection of meningiomas, involving the cranial nerves, is a more complex process; sometimes total resection is not possible. Tumor recurrence is possible and some symptoms may remain or worsen due to surgical complications. *Sanna et al.* reported 11

cases in which the patients did not improve their neurological deficits after surgery, and more than 50% developed new deficits[18]. The efficacy of radiosurgery has already been proven, especially in small tumors, but there are no studies comparing the results of conventional surgical treatment and radiosurgery[10].

The treatment of chordomas involves radical surgical resection of the tumor followed by adjuvant radiotherapy or radiosurgery. If there is infiltration of adjacent structures, total resection is unfeasible. Among the pathologies discussed in this chapter, chordomas are among those with the worst prognosis.

For chondrosarcomas, total surgical resection is the first-line treatment, with radiotherapy indicated in cases of subtotal resection or high-grade lesions[19].

The main complication of surgical treatment for jugular foramen tumors is cerebrospinal fluid fistula, since bone involvement and surgical manipulation sometimes do not promote good sealing of the dura. Ramina *et al.*[20] developed a skull base reconstruction technique for jugular foramen tumors, which showed lower fistula rates, as well as better aesthetic results. It is worth noting that surgical complications vary little according to the type of neoplasm. Other complications include cranial nerve damage, peri- and post-operative hemorrhage, infection, neurological deficit (temporary or not) and hydrocephalus.

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