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Clinicopathologic Features of Jugular Foramen Tumors

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The jugular foramen (JF) is an anatomically complex region where important bony, neural, and vascular structures converge. Tumors are the primary cause of JF pathology and often present therapeutic challenges. In this article we discuss the clinical and pathologic aspects of JF tumors. These aspects include the classic JF clinical syndromes, imaging characteristics of specific JF tumors, and gross pathologic and histopathologic features. The three most common and important JF tumors, glomus jugulare tumors, meningiomas, and schwannomas, are discussed in depth. A wide variety of tumors can occur in the JF. Nonetheless, our current clinical and radiographic knowledge usually allows a clinician to ascertain the pathology of a JF tumor preoperatively with a high degree of certainty. Doing so allows the clinician not only to formulate the most effective treatment plan but also to provide accurate and thorough preoperative counseling to patients harboring these serious lesions.

KEYWORDS jugular foramen, meningioma, schwannoma, glomus jugulare

The jugular foramen (JF) region is a complex anatomical area that is an important though relatively uncommon location for skull base tumors. Pathology in the JF may manifest with dysfunction of only the traversing cranial nerves (CN IX, X, XI). More commonly, however, surrounding structures such as the middle ear, cerebellopontine angle, or high retropharyngeal space are also involved. In the surgical literature the term, JF tumor, usually refers not only to tumors that arise in the foramen itself (eg, glomus jugulare tumors) but also to tumors that arise in the jugular fossa adjacent to the foramen such as jugular fossa meningiomas.

The differential involvement of the lower cranial nerves by disease processes in and near the JF led to the characterization of a number of JF syndromes.[1] These syndromes must be understood in context because the most common JF tumor, glomus jugulare, rarely ever leads to one of them. When seen, JF syndromes usually result from less common lesions like schwannomas or metastases. Though termed JF syndromes, these clinical pictures may result from lesions from the brainstem nuclei to the extracranial retropharyngeal space (Table 1).

JF (Vernet's) syndrome: Unilateral involvement of CNs IX, X, XI leads to loss of taste in the posterior third of the tongue; hemianesthesia of the palate, pharynx, and larynx; and weakness or paralysis of the vocal cords, palate, trapezius, and sternocleidomastoid muscle.

Posterior lacerocondylar (Collet-Sicard) syndrome: Involvement of the hypoglossal (CN XII) nerve in addition to CNs IX, X, and XI leads to tongue atrophy and weakness or paralysis.

Posterior retropharyngeal (Villaret's) syndrome: This syndrome has the same clinical picture as the Collet-Sicard syndrome with the addition of Horner's syndrome, usually related involvement of the sympathetic nerves near the high cervical or petrous internal carotid artery.

Allied syndromes: Several other syndromes involving lower cranial nerve dysfunction also have been described. They are usually the result of brainstem ischemic injury and rarely because of tumor. In *Avellis's syndrome*, vocal cord/palate paralysis (CN X) is associated with contralateral dissociative anesthesia (spinothalamic tract). *Schmidt's syndrome* involves ipsilateral anesthesia of the pharynx and larynx (CN X), paralysis of the soft palate and larynx (CN X), and weakness or paralysis of the trapezius and sternocleidomastoid muscles (spinal CN XI). *Tapia's syndrome* involves paralysis of the ipsilateral tongue, pharynx, and larynx because of a lesion affecting the motor nuclei of CNs X and XII. Finally, *Jackson's syndrome* involves ischemic nuclear or radicular injury to CNs X, XI, and XII.

Many etiologies other than tumor and brainstem ischemia must be considered in patients presenting with one of the JF syndromes or a single lower cranial nerve deficit. Historically, foremost among these are leptomeningeal processes such as tuberculosis and syphilis, which used to account for almost half of the cases of JF or related syndromes.[1] Other

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Table 1 JF and Allied Syndromes

Syndrome	Neurologic involvement
JF (Vernet's) syndrome	CN IX, X, XI
Posterior lacerocondylar (Collet-Sicard) syndrome	CN IX, X, XI, XII
Posterior retropharyngeal (Villaret's) syndrome	CN IX, X, XI, XII, sympathetic chain
Avellis's syndrome	CN X, spinothalamic tract
Schmidt's syndrome	CN X, spinal XI
Tapia's syndrome	CN X, XII
Jackson's syndrome	CN X, XI, XII

leptomeningeal processes include neurosarcoidosis and carcinomatous meningitis. Basilar skull fractures, retropharyngeal abscesses, jugular vein thrombosis, and aneurysms are a few of the other rare causes of JF syndromes.

Imaging of JF Lesions

Advances in neuroimaging have greatly simplified the diagnosis of pathology involving the JF. With respect to tumors, assessment with magnetic resonance imaging (MRI), computed tomography (CT), and cerebral angiography can usually suggest the pathologic entity with a high degree of certainty. This has obvious advantages for preoperative decision making, including the choice of approach, surgical goals, need for preoperative embolization, and patient counseling.

The most common tumors involving the JF are glomus tumors, meningiomas, and schwannomas.[2] Less commonly found are metastatic lesions, primary bone tumors, chordomas, chondrosarcomas, and epidermoid tumors. Among the three most common tumors of the JF, glomus tumors are by far the most numerous. A comprehensive review of JF lesions revealed 509 glomus jugulare tumors in comparison to 104 schwannomas and 34 meningiomas.[3] Because glomus tumors, meningiomas, and nerve sheath tumors compose most JF lesions, the surgeon must have an intimate knowledge of their imaging characteristics.[4]

This will be reviewed in the next article, but the salient features for these tumors are listed in Table 2.

Clinicopathologic Aspects of Individual Tumors

Meningiomas, schwannomas, and glomus jugulare tumors compose more than 80% of tumors involving the JF in one series[2] and probably compose a higher proportion of tumors whose primary involvement is the JF. Considering them individually is useful.

JF Meningiomas

Primary meningiomas of the jugular fossa and foramen are rare tumors; fewer than 50 cases have been reported. Their rarity is attested to by their absence from the authoritative meningioma treatises by both Cushing and Eisenhardt[5] and Castellano and Ruggiero.[6] The largest published series have contained only eight patients,[7,8] and in a series of 161 poste-

Table 2 Imaging Characteristics of Common JF Area Tumors

Tumor	MRI	CT	Angiography
Meningioma	Isointense to cortex on T1- and T2-weighted images, homogeneous enhancement with gadolinium, enhancing dural tail, absence of vascular flow voids	Jugular foramen is not enlarged, occasional calcifications, hyperostosis	Faint tumor blush
Glomus jugulare	Salt and pepper appearance, intense enhancement with gadolinium	Erosive enlargement of JF with irregular bony margins	Extreme vascularity with obvious tumor blush, AV shunting, JF filling defect, occasionally tumor extends intraluminally up the sigmoid sinus or down the jugular vein
Nerve sheath tumor: schwannoma or neurofibroma	Irregular enhancement with gadolinium, high signal intensity on T2-weighted images	Smooth, scalloped enlargement of the JF, low signal density	Minimal if any tumor blush
Chondrosarcoma	Slight enhancement with gadolinium, high signal intensity on T2-weighted images	Erosive enlargement of JF with irregular bony margins	Avascular mass without tumor blush