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Anatomy, physiology, and genetics of paragangliomas

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Paragangliomas arise from an extra-adrenal paraganglionic cells, derived from the neural crest of the autonomic nervous system and make up the most common class of benign vascular neoplasms of the neck. PGLs of the head and neck originate most commonly from the paraganglia within the carotid body, the jugular foramen, the middle ear and the vagus nerve. Knowledge of the embryology, anatomy and genetics of these rare tumors is paramount to successful diagnosis and treatment of head and neck paragangliomas.

Paragangliomas (PGLs) arise from an extra-adrenal paraganglionic cells, derived from the neural crest of the autonomic nervous system and make up the most common class of benign vascular neoplasms of the neck.[1] PGLs can be divided into the following 2 groups: sympathetic-derived and parasympathetic-derived tumors.[2] Tumors in the thorax, abdomen, and pelvis are usually derived from sympathetic paraganglia, and are more often associated with catecholamine production.[3] Pheochromocytomas are histologically similar to sympathetic PGLs, and arise in the adrenal gland.[3] On the other hand, head and neck region PGLs (HNPGLs) are most often benign, slowly progressing, derived from parasympathetic paraganglia and rarely secrete catecholamines.[3,4] PGLs represent an estimated 0.01% of human tumors.[5] Pheochromocytomas make up approximately 90% of tumors arising from the paraganglion system and the remaining 10% arise from extra-adrenal sites, with 85% arising in the abdomen, 12% in the thorax, and the remaining 3% in the head and neck area.[1] PGLs of the head and neck originate most commonly from the paraganglia within the carotid body, the jugular foramen, the middle ear (tympanicum), and the vagus nerve (Figure 1).

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Carotid body

The carotid body is the most common location for HNPGLs.[1] The carotid body, located in the adventitia of the posteromedial aspect of the carotid artery bifurcation, is a chemoreceptor that modulates respiratory and cardiovascular function in response to pH, oxygen, carbon dioxide tension changes.[1] Low pH and low oxygen stimulate the carotid body to initiate an autonomic response to increase respiratory rate, heart rate, blood pressure, and cerebral cortical activity [1] (Figure 2). Most carotid body tumors (CBTs) are nonfunctional, nonsymptomatic, and slow growing.[6,7] CBTs usually present as a lateral cervical mass, which is mobile laterally but less mobile cranio-caudally owing to its adherence to carotid arteries.[1] As they enlarge, 10% of patients may present with symptoms of dysphagia, odynophagia, hoarseness, and other cranial nerve (CN) deficits.[1] Moreover, carotid sinus syndrome syncope, which is a loss of consciousness and reflex bradycardia and hypertension, has been described to be associated with CBTs.[1] However, some CBTs may be functional, which

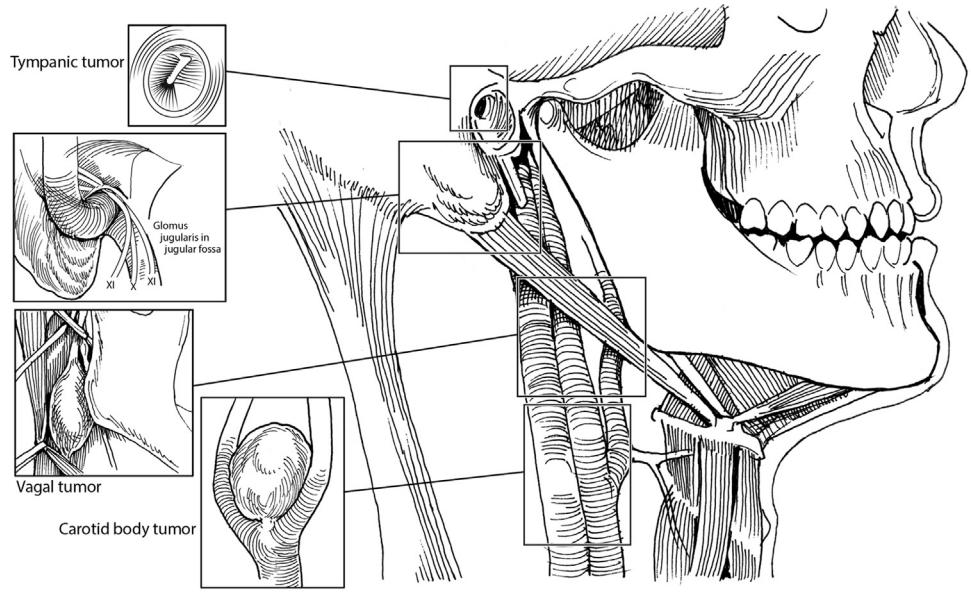

Figure 1 Common locations of paragangliomas of the head and neck.

means they are able to synthesize catecholamines, and therefore may lead to symptoms such as heart palpitations, dizziness, headache, and hypertension.[7]

The right common carotid artery originates from the brachiocephalic trunk immediately posterior to the right sternoclavicular joint, whereas the left common carotid artery begins in the thorax as a direct branch of the arch of the aorta and passes superiorly to enter the neck near the left sternoclavicular joint.[8] The right and left common carotid arteries ascend lateral to the trachea and esophagus within the carotid sheath.[8] Each common carotid artery then divides into its 2 terminal branches, the external and internal carotid arteries.[8] At the bifurcation, the origin of the internal carotid artery is dilated. This dilation is the carotid sinus, which has receptors that monitor blood pressure.[8] The carotid body is another accumulation of receptors in the area of the bifurcation and is responsible for detecting changes in blood chemistry, particularly the oxygen content.[8] The carotid body is innervated by branches from both the glossopharyngeal (IX) and vagus (X) nerves.[8]

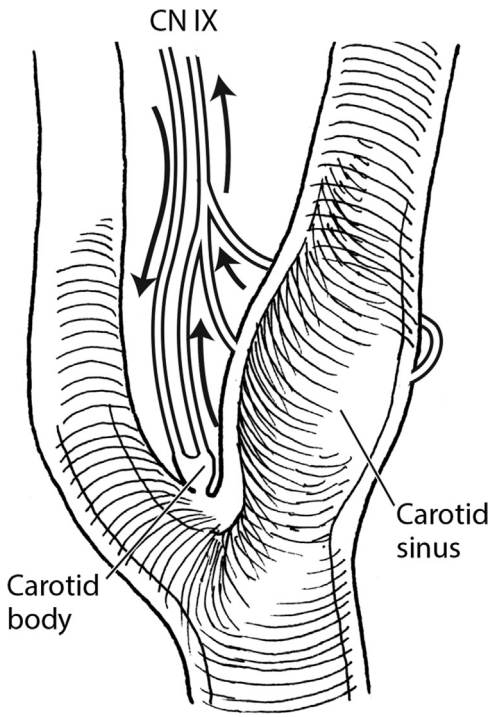

Figure 2 The bifurcation of the common carotid artery, demonstrating baroreceptors in the wall of the carotid sinus and chemoreceptors within the carotid body.

Jugular foramen

Jugular PGLs, the second most common HNPGL, arise from paraganglia in or around the jugular bulb along Jacobson nerve (CN IX) or Arnold nerve (X).[9,10] Pulsatile tinnitus is the most frequent symptom, and conductive hearing loss is seen with the progression of the tumor, which either causes impairment of ossicles vibration or invades bones behind the eardrum.[11] When the tumor invades the inner ear, sensorineural hearing loss and dizziness is reported.[11] Moreover, jugular PGLs can cause deficits of other cranial nerves and dysfunctional swallowing and hoarseness.[11] Upon further growth of these tumors, they can also invade the facial nerve, which can lead to facial paralysis or invade the hypoglossal nerve, leading to paralysis of the tongue [11] (Figure 3).

The jugular foramen is located in the floor of the posterior fossa, posterolateral to the carotid canal, and between the petrous temporal bone and occipital bone.[12] A complex canal of neurovascular structures in the skull base, the jugular foramen is divided into the pars nervosa (anteromedial) and the pars vascularis (posterolateral).[12] The pars nervosa contains the glossopharyngeal (IX) with Jacobson nerve and the pars vascularis contains the internal jugular vein, vagus nerve (X), and spinal accessory (XI).[12]