

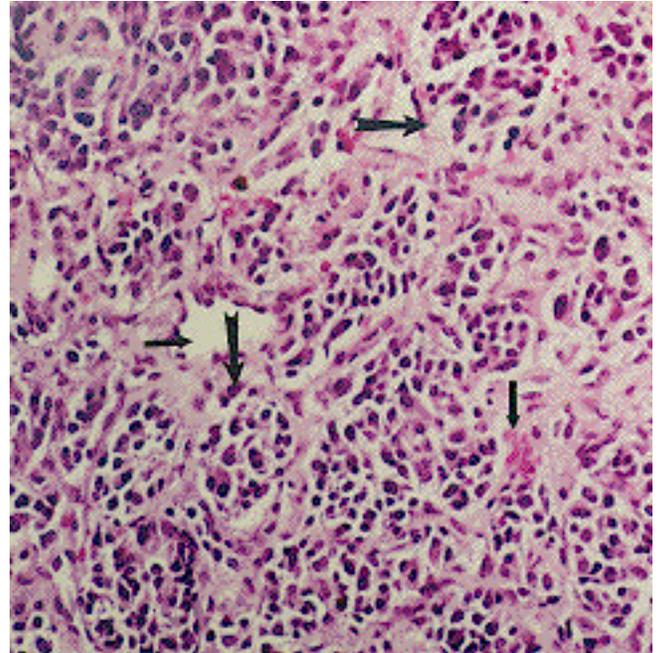
## **GLOMUS TUMOR (NONCHROMAFFIN PARAGANGLIOMA, CHEMODECTOMA)**

These neoplasms, the most common tumors of the middle ear, occur in the temporal bone as 1) glomus tympanicus tumors in which a small neoplasm arises from the promontory (medial wall of cochlea) or 2) glomus jugulare tumor which arises from glomular cells on the apex of the bulb of the internal jugular vein and grows upward into the middle ear and mastoid and other bones of the skull base. All are extremely vascular. Their origin is from chemoreceptor organs of the parasympathetic nervous system. The glomus bodies of a glomus tympanicus tumor are located along the tympanic branches of the vagus and glossopharyngeal nerves.

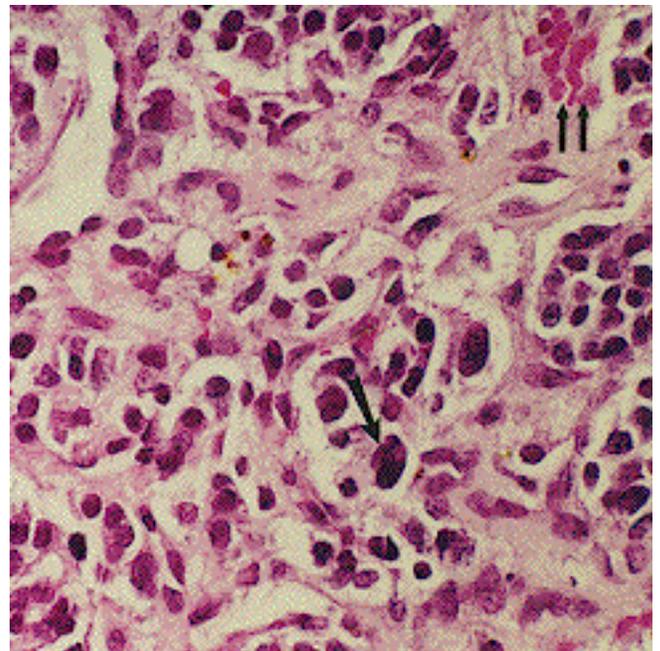
The carotid body tumor is also a paraganglioma and is found at the carotid bifurcation. It is histologically identical to the glomus tumor of the temporal bone. Other paraganglion cells are in the laryngeal, orbital and nasal areas and tumors, are reported in association with these sites.

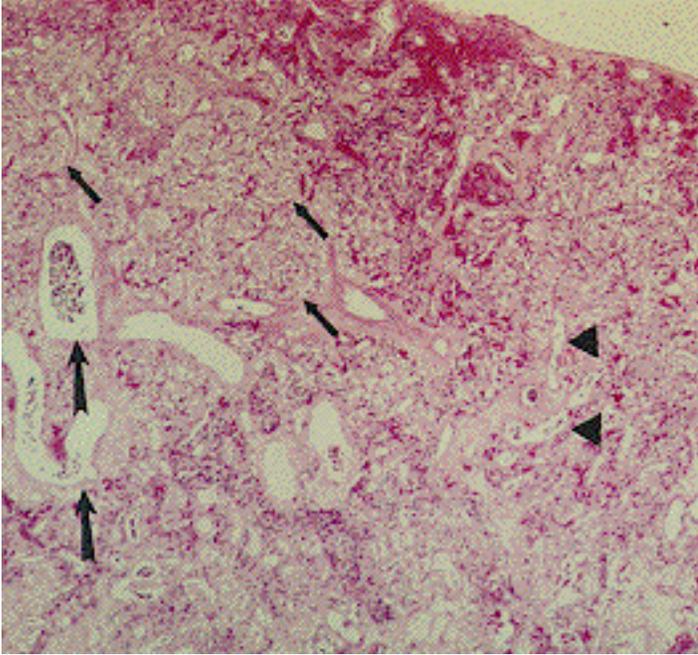
Histologically, a glomus tumor demonstrates “Zellballen” — alveolar-like nests of tumor cells. Surrounding these cell nests is a dense network of thin walled sinusoidal capillaries. Thick bands of collagenous tissue divide the tumor. There are 5-20 epithelial cells comprising a nest or group of cells.

Glomus jugulare tumor showing "Zellballen" (large arrows), cell nests of 5 to 30 cells with eosinophilic granular cytoplasm sometimes with considerable nuclear pleomorphism as seen here. Small arrows indicate blood vessels.

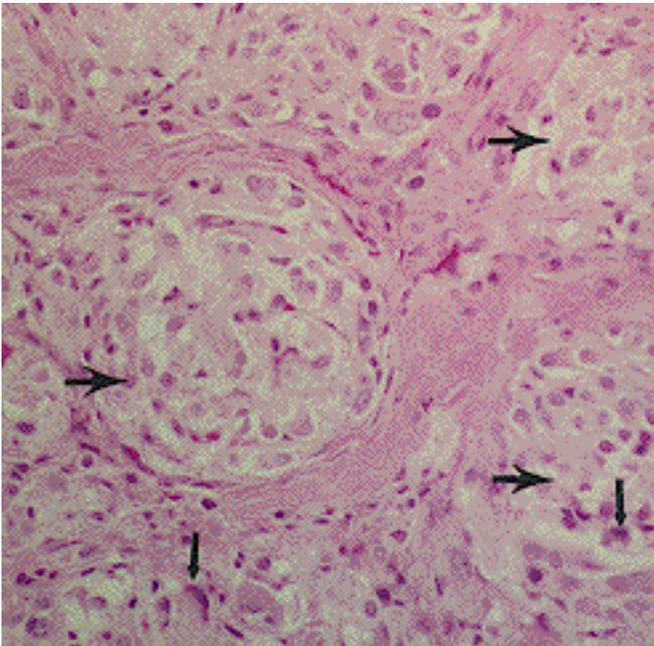


High power photo of tumor to show pleomorphic nuclei (large arrow), not an uncommon feature of these tumors. Mitoses are rare. The double arrows point to a small vessel.



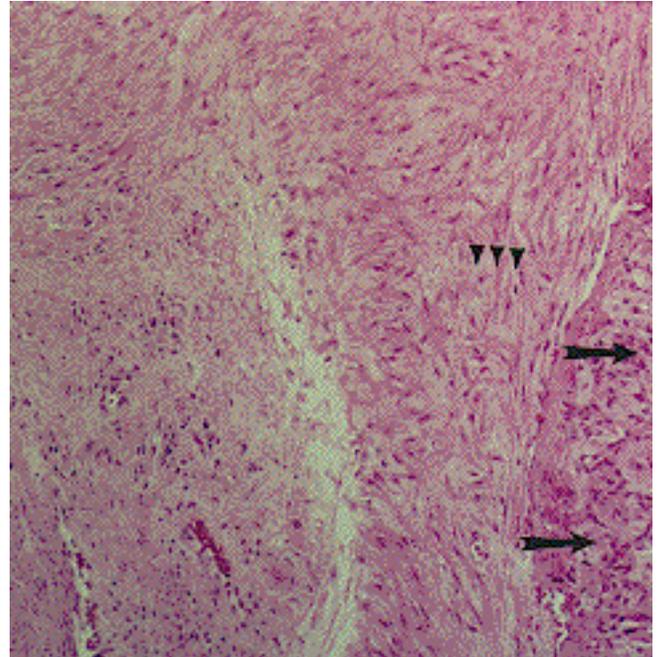


Recurrent glomus jugulare shows blood spaces (large arrows) with endothelial lining. Small sinusoids indicated by triangles. Cell nests are present (small arrows) but they are not as well seen as in other examples.

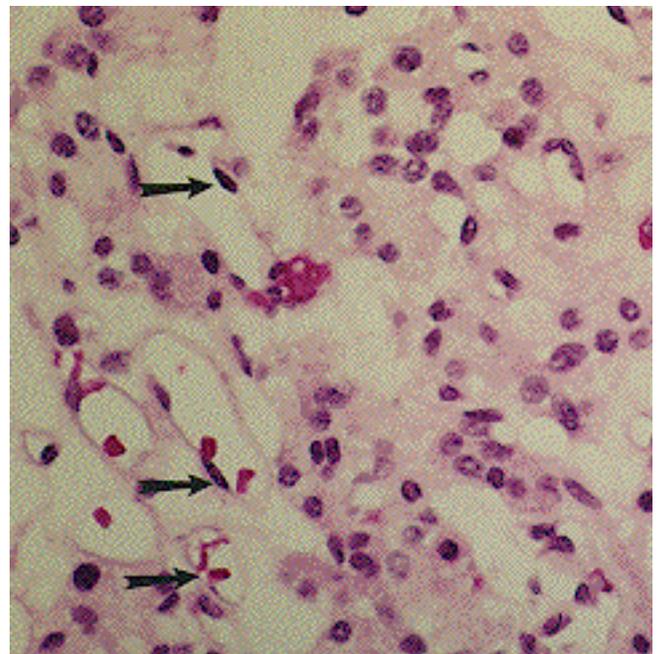


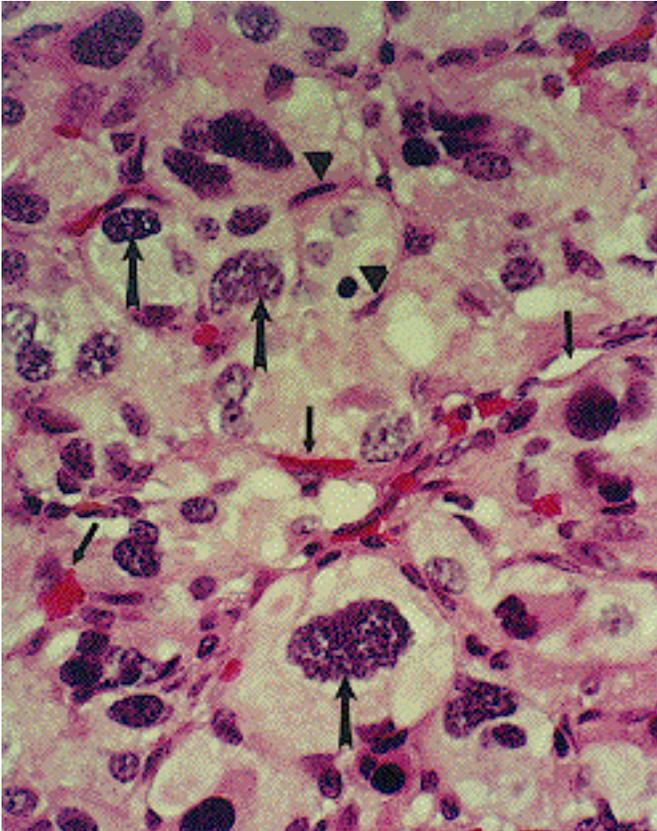
Glomus tumor, carotid, high power to show "Zellballen" (large arrows). These cells are of neural crest origin. Thick bands of collagen separate the nests. Some nuclear pleomorphism is seen (small arrows).

Glomus tumor (arrows), rests against adventitia (triangles) of carotid artery.

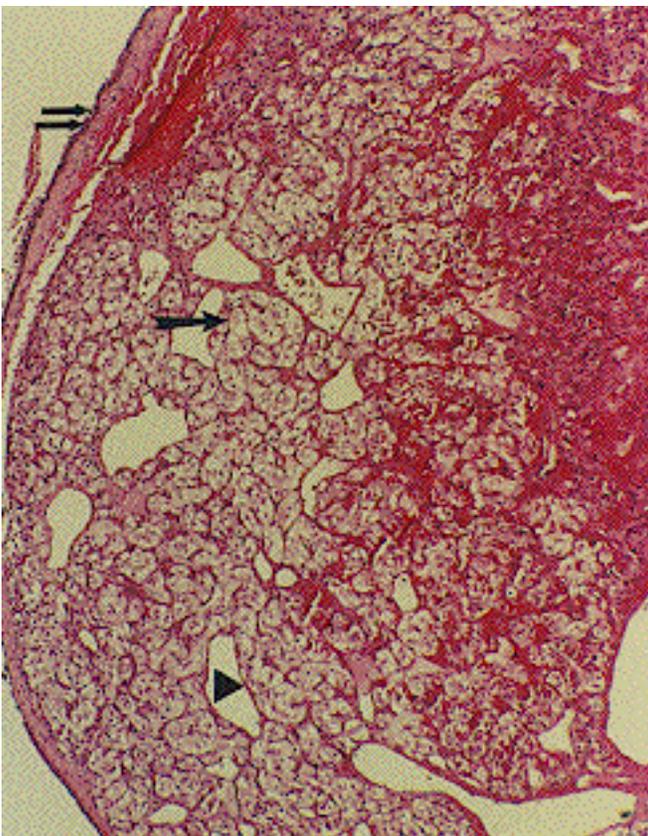


Glomus tympanicus, with demonstration of sinusoids lined by endothelium (arrows).





Glomus tumor, carotid, showing unusually large nuclei (large arrows) in large vacuolated cells. These rather spectacular hyperchromatic nuclei do not indicate malignancy but reflect the neuro-endocrine nature of the tumor. Note the blood-filled sinusoids (small arrows). Sustentacular cells (triangles) are modified Schwann cells, spindle-shaped and basophilic, and are seen at the margins of cell nests.



Glomus tympanicus. Zellballen pattern (arrow) is very definite and there are large vascular spaces (triangle). Also note the covering of the globular middle ear tumor with thin layer of epithelial cells over fibrous tissue (double arrows).

**CLINICAL ASPECTS:**

Parangliomas or glomus tumors are generally benign but some metastasize. They are more common in women than in men and generally appear in the latter half of life.

The glomus tympanicum arises from the promontory of the middle ear and can be seen (6 or 10 power magnification) as a small red or pink mass that pulsates. Symptoms are a loud tinnitus and, depending on the size of the tumor, a conductive hearing loss.

The glomus jugulare tumor arises from the dome of the jugular bulb or walls of the jugular fossa and grows much larger than the glomus tympanicus tumor. There are no early symptoms but eventually neural deficits appear as cranial nerves VII, VIII, IX, X, and XII become involved. The tumor grows upward through the floor of the middle ear and may be seen as a mass filling the middle ear or even growing into the floor of the ear canal. Sometimes the tumor presents as an aural polyp filling the ear canal. Then bleeding is common and may be profuse.

The carotid body paraganglioma presents in the neck at the carotid bifurcation and can be moved sidewise but not vertically. Bilateral carotid body tumors are known, especially in the familial form. This tumor, too, can be locally invasive with involvement of the vagus nerve and obliteration of the internal jugular vein. It may metastasize.

Diagnosis of any of these tumors is made by an arteriogram or CAT scan with contrast.

Treatment of all of these tumors is preferably by surgery, but irradiation therapy is effective in limiting further growth and may even shrink some tumors.