TUMOR OF THE GLOMUS JUGULARE WITH EXTENSION INTO THE MIDDLE EAR
(NONCHROMAFFIN PARAGANGLIOMA OR CAROTID-BODY-TYPE TUMOR)*

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(Received for publication May 28, 1951)

PRIMARY tumors of the middle ear and petrous portion of the temporal bone occur rarely and are usually observed by the otologist. Occasionally an invading tumor in this region will cause sufficient neurological disorder to bring the case to the attention of a neurologist or neurosurgeon. One of the authors encountered such a case with neurological signs and symptoms of primary importance, in which a carotid angiogram was helpful in delineating the tumor and determining future therapy.

HISTORICAL REVIEW

In 1840 Valentine\(^5\) originally described a slight swelling of the tympanic nerve; Krause later studied this structure more completely (1878) and noted its resemblance to the carotid body at the bifurcation of the common carotid artery.

Apparently this problem received little attention until 1941, when Guild\(^6\) examined serial sections of human temporal bone and found small bodies (0.45–0.25 mm.) in the adventitia of the dome of the jugular bulb. Some of these bodies were situated along the tympanic branch of the glosso-pharyngeal nerve and others were below the tympanic cavity. They varied in size and position, and frequently two or more were found on each side. Histologically, these bodies resembled the carotid bodies at the bifurcations of the common carotid arteries. Guild believed the blood supply of these tumors was derived from the external carotid artery, through the tympanic branch of the ascending pharyngeal artery; he described the nerve supply arising from the glosso-pharyngeal nerve, through the *ramus tympanicum*. The location of these small bodies and their histological structure were confirmed by more recent work of Lattes and Waltner.\(^8\)

In 1945 Rosenwasser\(^13\) performed subtotal removal of a tumor in the middle ear, and Dr. Otani, who studied the histologic preparations, was impressed by the structural resemblance of this tumor to the normal carotid body. It was postulated that the tumor arose from the glomus jugulare, as described by Guild,\(^5\) and this was the first time a tumor of this type was reported. Subsequently similar tumors were described by LeCompte, Som-

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mers and Lathrop\textsuperscript{6} and Kipkie.\textsuperscript{6} Winship, Klopp and Jenkins\textsuperscript{14} reported 2 additional cases and reviewed histologic material from several cases which were reported previously. It would seem that several tumors in and about the middle ear and petrous portion of the temporal bone had been classified as endotheliomas and hemangioendotheliomas, although they were probably unrecognized tumors of the glomus jugulare. They found at least 11 such cases and 13 others which they suspected were probably tumors of the glo-

Lundgren\textsuperscript{11} reported 4 similar cases in which the lesions were termed “tympanic body tumors of the middle ear of the carotid body type,” and the microscopic features of these tumors were elucidated, also, by Berg.\textsuperscript{2} More recently De Lisa,\textsuperscript{8} Dockerty, Love and Patton,\textsuperscript{4} Poppen and Riemenschneider,\textsuperscript{12} and Lewis and Grant\textsuperscript{10} have reported additional cases in which tumors of the glomus jugulare were encountered.

In addition to confirming the presence of nests of cells resembling the carotid body in the region of the jugular bulb and middle ear, Lattes\textsuperscript{7}, and Lattes and Waltner\textsuperscript{8} point out that similar structures are found also (1) in the ganglion nodosum of the vagus nerve, (2) in the adventitial portions of the ascending aorta, aortic arch, innominate artery and probably the pulmonary artery and (3) possibly in the orbit near the ciliary ganglion. They suggested that these structures be considered as homologous parts of a system composed of nests of nonchromaffin cells, of “epithelioid” type, situated in a perivascular stroma and resembling the carotid body histologically. Generally these structures are innervated by afferent fibers from a cranial nerve, and these authors believe the cell-nests probably function as chemoreceptors. However, these structures probably do not secrete epinephrine or other known hormones. The name “paraganglion” was suggested because this term had been used to designate the carotid body, which these structures resemble so closely. The term “nonchromaffin” was applied to differentiate such structures from the chromaffin paraganglion located in the adrenal medulla and the nests of chromaffin cells associated with the visceral sympathetic ganglia. Inasmuch as Lattes\textsuperscript{7} reported tumors of this type in the ganglion nodosum and adventitia of the aortic arch, as well as in the region of the carotid body and glomus jugulare, the authors postulated that such tumors probably arise from nests of “epithelioid” cells normally situated in these locations.

Lattes and Waltner\textsuperscript{8} studied their own histologic preparations and found a total of 8 cases that they classified as tumors of the glomus jugulare. Some of these had been classified in other categories prior to the above concept of derivation. Judging from their paper and the review of Winship, Klopp and Jenkins,\textsuperscript{14} approximately 28 proved cases of tumors of the glomus jugulare have been reported in the literature, 22 of them in females.

To date only two reports concern instances in which patients with tumors of the glomus jugulare required neurosurgical exploration.\textsuperscript{4,12} Angiography was used in studying the location and extent of the tumor in the case of