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

EBEN ALEXANDER, JR., M.D., PARKER R. BEAMER, M.D.,
AND JEROME O. WILLIAMS, M.D.

Departments of Neurosurgery and Pathology, Bowman Gray School of Medicine
of Wake Forest College, Winston-Salem, North Carolina

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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 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 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 glosopharyngeal 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 glosopharyngeal 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.

In 1945 Rosenwasser 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, and this was the first time a tumor of this type was reported. Subsequently similar tumors were described by LeCompte, Som-

mers and Lathrop and Kipkie. Winship, Klopp and Jenkins 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 glomus jugulare, but exact identification was not possible.

Lundgren reported 4 similar cases in which the lesions were termed "typanic body tumors of the middle ear of the carotid body type," and the microscopic features of these tumors were elucidated, also, by Berg. More recently De Lisa, Dockerty, Love and Patton, Poppen and Riemenschneider, and Lewis and Grant 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, and Lattes and Waltner 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 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 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, 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. Angiography was used in studying the location and extent of the tumor in the case of
Poppen and Riemenschneider. They reported a vertebral angiogram showed slight pooling of the thorotrast in the region of the tumor.

CASE REPORT

A 35-year-old negro woman was admitted to the Kate Bitting Reynolds Memorial Hospital in Winston-Salem, June 24, 1950. Her chief complaint was that of hiccoughs for 1 month.

She had not been in good health for approximately 3 years and complained initially only of generalized weakness, for which she had been given thyroid extract without much benefit. One year prior to admission, she first noted deafness in her left ear and tinnitus associated with some ataxia. She had had some headache previously, but began to have headaches of greater severity. She denied having had any drainage in either ear or any bleeding at any time.

Four months before admission, she noticed one morning a sudden onset of paralysis of the left side of her face associated with numbness of the left face as well as the left side of her palate and tongue. There was increased ataxia and she felt that her voice took on a different tone. There was possibly slight numbness of the entire left side of her body but during the 3 months before admission, this had disappeared as well as the numbness of her face, tongue and palate.

One month before admission, hiccoughs began and were almost constantly present. She had been admitted to another hospital where a spinal puncture had been done; the CSF pressure and protein were reported as normal.

Examination. Temperature 98°, pulse 80, respirations 16 and B.P. 110/80. The patient was thin but fairly well developed. She hiccuped during the entire examination. There was a "nasal twang" to her voice. Except for loss in weight, the general physical findings were normal, and there was no evidence of lymphadenopathy. When she was sound asleep, secretions collected in the posterior pharynx and were removed with some difficulty by the patient.

Palpation of the head revealed no abnormality. On auscultation, behind the left ear, a continuous murmur with a loud systolic component and a very faint diastolic component was heard. The first 6 cranial nerves showed no abnormalities. There
was no papilledema and no residual paralysis of the 5th cranial nerve, either motor or sensory. There was no nystagmus.

There was almost complete peripheral paralysis of the left 7th nerve with only faint motion of the orbicular oris on the left. There was complete paralysis of the 8th, 9th, 10th, 11th and 12th cranial nerves on the left. The vocal cords were not examined. The right ear drum was normal. The left ear drum showed, protruding through it, a smooth, red mass with very slight pulsations synchronous with the heart beat.

There was ataxia of the left arm and leg. There were hyperactive deep tendon reflexes on the left, an unsustained ankle clonus of the left foot and a questionably positive Babinski response on the left.

*Laboratory Data.* RBC, WBC and Hb. were normal. Urine was normal. Blood Kahn was negative.

Roentgenograms of the chest were normal. The skull showed extensive destruction of the petrous portion of the left temporal bone with cloudiness of the left mastoid (Fig. 1).

*Biopsy* of the tumor mass in the left external auditory meatus was made by Dr. George Bradford. There was extensive bleeding from this which was controlled by packing the external meatus.
Histological Study. This is a relatively cellular tumor in which the cells are closely packed in an alveolus-like arrangement surrounded by fibrous-tissue stroma. The connective-tissue supportive stroma is generally variable in character, being dense in some areas while in others it is less prominent. The Masson’s trichrome stain confirms the presence of an abundance of fibrous connective-tissue stroma between the alveolus-like arrangements of the cells. Within the stroma there are many large thin-walled vessels. The cells making up the alveolus-like structures are all equal in size, shape and staining property, and are uniformly hyperchromatic. Although the cells have a distinct basophilic nucleus, the cell borders are not well defined. The cytoplasm is palely eosinophilic and contains a few fine granules. No mitotic figures are seen (Fig. 2).

Course. Subsequently left common carotid angiography was performed by the open technique, using 35 per cent diodrast. The films (Fig. 3) show a normal vascular pattern for the anterior and middle cerebral vessels. In addition they disclosed a very vascular mass which filled the floor of the posterior fossa on the left side, extending slightly across the midline. It extended posteriorly as far as the foramen magnum and seemed to extend into the middle fossa as well. It was obvious that the petrous portion of the temporal bone was grossly involved by this very vascular tumor.

The left vertebral artery was also injected under direct vision with 35 per cent diodrast but no dye was evident in any of the films. Why the vessels associated with the basilar artery and the posterior cerebral artery failed to fill is not apparent. Possibly there was improper timing in exposure to the films.

Because of the extreme vascularity of this tumor and its very extensive nature with invasion of bone and obvious growth across the midline, it was felt that this tumor was not operable. The patient was, therefore, subjected to radiation therapy, receiving 2080 roentgen units (in air) through each of two 10×10 cm. occipital portals crossfiring to the tumor. The total dosage was administered over a 19-day period. About 6 weeks later she received further roentgen therapy in another city. On this occasion approximately 900 roentgen units were given through the left occipital portal, and 1900 R. through the right occipital portal. She noticed some
subjective improvement, particularly relief of hiccoughs, but there was no change in the objective findings. The tumor mass in the left ear remained approximately the same size, and the murmur over the left posterior fossa was still apparent when she was discharged. A letter from her family physician in April 1951 indicated the patient has shown definite improvement. She is now able to walk and to do her housework. Efforts to see the patient by the authors and to repeat the carotid angio-
gram have so far not been successful.

**DISCUSSION**

Most tumors of the glomus jugulare now recorded in the literature were discovered because of signs and symptoms related to the ear; usually these were deafness and chronic otorrhea and, occasionally, complaints of dizziness, tinnitus and pain. Consequently, most of the cases were encountered by otologists and, in some instances, the tumors were small enough to permit complete or almost complete removal. In such cases, there was no apparent invasion of bone beyond the tympanic region, and it would appear that a cure could be expected. As mentioned before, certain tumors of this histo-
logical type occurred in more than one location; namely, the regions of the middle ear, the carotid body and the aortic arch. The tumor was regarded as malignant in only 3 cases. In the first, a case reported by Winship, Klopp and Jenkins\(^4\) the tumor was regarded as malignant on the basis of histologic observations; there was extensive destruction of the left mastoid and petrous portion of the temporal bone and apparently there were metastases to regional lymph nodes in the neck. In the second instance (Case 3 of Lattes and Waltner\(^5\)), autopsy studies revealed extensive invasion of the temporal bone with extension into the middle cerebral fossa. In addition to this, the liver contained a large focus which, although poorly preserved in the post-
mortem specimen, was regarded as tumor histologically similar to that in the petrous portion of the temporal bone. The third case, that of Poppen and Riemenschneider,\(^12\) showed metastases to five local lymph nodes.

Tumors of the glomus jugulare are encountered most frequently in the age group from 30 to 50 years, but they have been found in patients as young as 23 and as old as 80 years. In the relatively small number of cases (28) thus far recorded in the literature, there is a higher incidence (22) in women. The medical history is usually of relatively long duration, and in at least 2 cases the patients suffered symptoms for 20 years or more.

There have been at least 7 deaths among the recorded cases of tumors of the glomus jugulare: 2 patients died following the attempted removal of the tumor; 4 died apparently of gross extension of the tumor into the intra-
cranial cavity; and 1 died of meningitis and a cerebellar abscess. The outlook for our patient is obviously not good in view of the involvement of so many of the cranial nerves and the extension of the tumor to the middle and pos-
terior cranial fossae. If roentgen therapy fails to give any substantial and per-
manent relief, and if the patient begins to show signs of increased intracranial pressure, it is possible that some relief of symptoms might be obtained by splitting of the tentorium, although this would be a very temporary measure.
Roentgen therapy has not been found to be particularly effective in tumors of this histologic type, occurring in the region of the carotid body, and it is likely, as has been indicated previously, this tumor will not be favorably affected by such treatment. Bevan and McCarthy in 1929 reported striking diminution in the size of a carotid-body tumor 13 months after local roentgen therapy. A continued trial of roentgen therapy in tumors of this size invading the petrous portion of the temporal bone and with intracranial extension is indicated. Possibly Poppen’s sugestion of ligating the arterial supply to these tumors will be of benefit, although it is likely they receive some blood from both carotid and vertebral arteries when they attain large size. Guild’s studies would point to the external carotid artery as the principal source of blood for the glomus jugulare.

It was found that the carotid angiogram was of valuable assistance in determining whether the particular tumor was operable, but it is well to point out that since there is great variation in the vascularity and some variation in the actual cellular structure of these tumors, all tumors of the glomus jugulare may not be demonstrated by carotid angiography so dramatically. In many patients coming to the neurologist or to the neurosurgeon because of involvement of the intracranial cavity, carotid angiography would appear to be a very valuable part of the neurological study.

SUMMARY

1. A tumor of the glomus jugulare has been reported. Approximately 28 proved cases have been previously reported under the terms “glomus jugulare tumors,” “tympanic body tumors” and “nonchromaffin paraganglioma of the middle ear.”

2. These tumors are histologically similar to the carotid body tumors and to the rare tumor occurring in the region of the aortic arch.

3. Tumors of this type are locally invasive but usually benign and very slow in their growth. Occasionally such tumors can be removed completely when discovered in the early stages.

4. The extent and vascularity of the tumor in this case was demonstrated by carotid angiography.

5. Because of the x-ray appearance of this tumor in the plain skull films and in the angiograms, it has been considered inoperable and is being subjected to extensive radiation therapy.

6. It remains to be seen whether roentgen therapy will be effective in shrinking tumors of this type.

REFERENCES

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DISCUSSION

Dr. R. E. SEMMES, Memphis, Tennessee: A housewife, aged 32, had had an intermittent blowing sound in the right ear, synchronous with the heart beat, for 3½ years, pain in the right ear for 1 year, and weakness in elevating the right shoulder for 1 year. A red discoloration across the right ear drum had been present for 1 year.

Because of these symptoms, a spinal fusion for spondylolisthesis was deferred. After several months the pain in the ear subsided, and fusion was carried out on Jan. 4, 1951.

Shortly thereafter, the pain returned, the blowing noise increased, and there developed weakness of the right side of the tongue, paralysis of the sternocleidomastoid muscle, and ringing and diminished hearing in the right ear. Compression of the jugular vein resulted in no changes but compression of the common carotid artery stopped the bruit and the pain—which returned promptly on release of the vessel.

Roentgenograms of the skull showed questionable enlargement of the jugular foramen.

An arteriogram of the right internal carotid was negative. The carotid was then exposed and it was found that compression of the internal carotid stopped the bruit and the pain, but compression of the external carotid, which was small, had no effect. An arteriogram of the vertebral was negative. Weakness of the 9th, 10th and 12th nerves was observed, pointing to the jugular foramen.

On Jan. 29, 1951, a right suboccipital exploration was done through a straight incision and a firm, tough, pinkish tumor, about 2 cm. in diameter, was found blocking the jugular foramen. It was moderately vascular and incorporated the 9th, 10th and 11th nerves. The 11th nerve could not be saved and damage was done to the 9th and 10th. The tumor was adherent to the 8th. It was removed completely as far as could be determined and the dural attachment of the foramen was lightly cauterized.

Postoperatively there was no pain in the ear, the bruit was gone, the 11th, 10th, and 9th nerves were out of commission, there was increased deafness, and 3 days later paralysis of the 7th, which later cleared.

Dr. Exum Walker, Atlanta, Georgia: A 32-year-old male complained of pounding noise in the right ear of 5 years' duration. There was slight deafness in the right
ear and a definite tenseness and bluish discoloration of the right ear drum. Carotid compression stopped the pounding noise.

On June 22, 1950, right arteriography was done. No abnormality was seen in any of these films. Ordinary x-rays of the skull showed clouding of the right mastoid region, interpreted as thickening of the mucosa.

In January 1951, a right facial paralysis developed. The patient was operated upon by an otologist, Dr. Leslie Brown, who found a tumor filling the region of the middle ear and pressing the carotid artery in the carotid canal. The tumor was said to have been completely removed but the facial nerve could not be spared. Histological diagnosis was nonchromaffin paraganglioma.

Dr. Brown stated that he had removed 1 other such tumor and had seen 3 additional cases of this type.

Dr. Jess D. Herrmann, Oklahoma City, Oklahoma: A 59-year-old housewife complained of excruciating pain in the left side of her face over the trigeminal area. Some 20 years before, a "blood tumor" had been removed from the left external canal, and about that time she noted loss of hearing on the left. About 7 years later paroxysmal attacks of pain developed in the left cheek. Two years before we saw her, she had had an alcohol injection of the infraorbital nerve which had relieved the sharp, stabbing pain but the "drawing pain" persisted. A year and a half before, a mass was seen in her external auditory canal and a biopsy was attempted, but bleeding was so profuse that no tissue was obtained.

There was involvement of the 5th nerve on the left with hypesthesia of the 1st and 2nd divisions. The motor root was not involved but there was corneal hypesthesia. There was a peripheral 7th nerve weakness which, from the history, had been present for some 5 years. There was total deafness with a dead labyrinth and there was also left 9th and 10th paralysis. Roentgenograms of the skull showed erosion of the left petrous bone.

At operation through the temporal route, a tumor mass was found extending through the anterior portion of the petrous bone. It had eroded the dura. It was quite vascular and we felt that it must be some kind of carcinoma. Our pathologist informed us of the true nature of this tumor. It was a tumor of the glomus jugulare.