IX

MULTIPLICITY AND FAMILIAL INCIDENCE OF CAROTID BODY AND GLOMUS JUGULARE TUMORS

DONALD R. RESLER, M.D.
JAMES B. SNOW, JR., M.D.  G. RAINNEY WILLIAMS, M.D.
OKLAHOMA CITY, OKLA.

The purpose of this paper is to report an unusual case of bilateral carotid body tumors and a glomus jugulare tumor occurring in the same patient and to discuss the multiplicity and hereditary-familial tendencies of chemodectomas.

Carotid body tumors and glomus jugulare tumors are considered to be chemodectomas, a term introduced by Mulligan\(^{30}\) to describe a neoplasm arising in the chemoreceptor system. The chemoreceptor bodies are located at the carotid bifurcation, jugular bulb, aortic arch, auricular branch of the vagus nerve (Arnold's nerve), tympanic branch of the glossopharyngeal nerve (Jacobson's nerve), inferior ganglion (ganglion nodosum) of the vagus nerve known as the vagal body, and the bifurcation of the pulmonary arteries. Chemodectomas, according to Berk,\(^{9}\) have also been reported in the eye, mandible, lung, stomach, the retroperitoneal tissues, mesentery of the ileum, and along the femoral artery. The tumors of the carotid body and glomus jugulare do not have epinephrine or glycogen in their cytoplasm. Although they are nonchromaffin their origin as paragangliomas remains in doubt. Their origin from tissue other than sympathetic tissue is suggested by the inability to demonstrate epinephrine in them.\(^{47}\)

CAROTID BODY TUMORS

In the eighteenth century (1757-1763) Von Haller\(^{16}\) was the first to dissect and describe the carotid body. The original description of a case of carotid body tumor was by Marchand\(^{30}\) in 1891. DeCas-

From the Department of Otorhinolaryngology and Department of Surgery, University of Oklahoma Medical Center.
INCIDENCE OF JUGULARE TUMORS

M.D.

W. WILLIAMS, M.D.

Oklahoma City

an unusual case of bilateral jugulare tumors occurring in the tympanic and hereditary-familial

ulare tumors are considered by Mulligan[30] to describe a stem. The chemoreceptor nerve, jugular bulb, aortic arch, Arnold's nerve, tympanic Jacobson's nerve), inferior s nerve known as the vagal arteries. Chemodectomas, in the eye, mandible, lung, xery of the ileum, and along carotid body and glomus cogen in their cytoplasm.

origin as parangangiomas sue other than sympathetic lemmonestrate epinephrine in

MORS

63) Von Halle[10] was the y. The original description archand[30] in 1891. DeCas-

sy and Department of Surgery,

tro[30] first suggested in 1926 that the carotid body might be a chemo-

receptor.

The carotid body is a 5 x 5 mm, ovoid mass usually located medially and behind the carotid bifurcation. The blood supply is mainly external carotid artery and possibly vertebral artery. The nerve supply is predominantly sensory via the glossopharyngeal nerve. The bodies are responsive to changes in pH, temperature, carbon dioxide tension, and oxygen[38,81]

Grossly the typical tumor is ovoid, firm, encapsulated, and trabeculated. Microscopically, the tumor is composed of large nests of cuboid or polyhedral cells arranged in alveoli separated by thick, vascular, fibrous trabeculae. The cells have a pale, eosinophilic, finely granular cytoplasm, and the nuclei are large and ovoid or round. There is an abundance of blood or lymphatic vessels, and the cells may group around the vessels. Mitosis is rare. The tumor shows a remarkable tendency to reproduce the architecture of the normal carotid body. The tumor is histologically considered benign.

There is an excellent review of the incidence of malignancy in carotid body tumors by Reese[30]. He relates a view by Monro that almost all carotid body tumors show some degree of local infiltration, and that the commonest overt malignant manifestation is local recurrence following local excision.[82] Harrington, Clagett, and Dockerty[37] found that half of their twenty tumors show histologic malignancy, that is active mitosis, cellular variation with giant cells, and capsular invasion. Other authors feel malignancy is very rare. Reese[30] reported a case of lymph node metastasis which was the thirteenth such case recorded in the literature by 1963.

The patient characteristically complains of a non-tender lump in the neck, slowly increasing in size over a period of years. Dysphagia may occur due to enlargement medially with displacement of the tonsillar fossa and pharyngeal wall. There may rarely be involvement of the cervical sympathetic chain, the vagus nerve, or the hypoglossal nerve by the tumor mass. Free lateral and little or no vertical mobility is a characteristic feature as is the presence of the external carotid artery stretched over the lateral aspect of the tumor. A carotid arteriogram is frequently important in the diagnosis and evaluation of carotid body tumors.

Surgical removal in the past has carried a mortality rate of 30 to 50 per cent, along with a high incidence of morbidity. Common
cartoid ligation is a frequent sequella to surgical intervention. Those who believe that carotid body tumors are benign suggest surgical intervention be limited to a diagnostic biopsy. This procedure is certainly not without hazard. Carotid body tumors are not considered radio-sensitive.

GLOMUS JUGULARE TUMORS

The first documentation of the histological appearance and anatomical location of the glomus jugulare was published by Guild in 1941. In 1945 Rosenwasser reported the first case of a glomus jugulare tumor. Guild suggested that the innervation of the glomus jugulare was from non-medullated fibers of the adjacent Arnold’s and Jacobson’s nerves. The blood supply is via the tympanic branch of the ascending pharyngeal artery. There are small anastomoses with the carotid system within the middle ear. The exact function of the glomus jugulare is unknown, but the presence of chemoreceptors is generally accepted.

The histological structure of the glomus jugulare tumor is indistinguishable from that of the carotid body tumor. There are large groups of epithelial cells with eosinophilic, granular cytoplasm and hyperchromatic nuclei. Tumor cells again are adjacent to thin walled capillary vessels. Mitosis is rare. Glomus jugulare tumors are locally invasive. There have been reports of metastasis to regional lymph nodes and to distant sites.

Usually the tumor grows slowly, and significant symptomatology may not be evident until the tumor is quite large. Occasionally the tumor is aggressive and results in death in two or three years. Alford and Guilford in an excellent review of glomus jugulare tumors and Rice point out the greater incidence of these tumors in women. The tumor is typically found in the middle-aged patient. Alford and Guilford found the most frequent presenting complaint in reviewing 277 patients to be hearing loss (91 per cent). Tinnitus of a pulsating nature, facial nerve paralysis, aural discharge, vertigo, hemorrhage, and palsy of the IX, X, XI, and XII cranial nerves were other prominent presenting and accompanying symptoms. Characteristically a red mass in the external auditory canal or a dark red or blue mass behind a tympanic membrane can be seen.

Pulsation and Blanching of the mass with compression with the pneumatic otoscope described by Brown may be of help in diagnosis. There may be x-ray evidence of petrous pyramid erosion.

R.B., a 47-year-old patient with a three month history of hearing loss, vertigo and mastication of the right ear was referred to Otolaryngology Section. Examination revealed right facial weakness and a right tympanic membrane mass 3 cm in diameter. A mass 3 cm in diameter was found in the external auditory canal with involvement of the tympanic membrane and a resultant conductive hearing loss. The patient was referred to Otolaryngology Section for further evaluation.

Treatment without a radical neck dissection and biopsy is the treatment of choice in this case. The patient received radiation therapy to the right side of the head and neck. After two months of radiation therapy, the patient noted significant improvement in hearing and facial function. The mass in the external auditory canal was reduced in size, and a new tympanic membrane was formed.

The patient reported no further episodes of vertigo and continued to have normal hearing and facial function. The mass in the external auditory canal was reduced in size and a new tympanic membrane was formed. Both tumors were removed with a minimal surgical procedure.
CAROTID BODY AND GLOMUS JUGULARE TUMORS

Treatment consists of excision of the smaller tumors with or without a radical mastoidectomy as described by Shambaugh.42 When the lesion becomes very large, radiation therapy to a dose of 5000 rads is the treatment of choice.

REPORT OF A CASE

R.B., a 47-year-old white female, was first seen by the Department of Otorhinolaryngology of the University of Oklahoma Medical Center on October 25, 1955. She gave a two year history of vertigo, twitching of the right side of the face, tinnitus and hearing loss in the right ear, and considerable pain about the right ear. There are no members of the family with ear or neck tumors. On physical examination she had a small red mass in the right ear canal. There was right VII and VIII cranial nerve paralysis and a three week history of drooping of the right eyelid. A biopsy of this lesion of the right ear on November 4, 1955, was consistent with a glomus jugulare tumor. During biopsy considerable bleeding was encountered. A right carotid arteriogram on November 14, 1955, was normal. X-rays of the mastoid region did not exhibit bone destruction.

Radiation therapy was begun on December 16, 1955. The patient received 2400 rads over a period of 15 days.

In November, 1957, the patient was re-admitted with an acute episode of right-sided headache, pain about the right ear, continued tinnitus, and hearing loss. There was erythema and swelling of the right tympanic membrane, accompanied by postauricular erythema. A mass 3 cm in diameter with a bruit was found in the left neck. On November 27, 1957, a modified radical mastoidectomy was performed.

The patient was re-admitted in November of 1962, complaining of severe vertigo, blurring of vision, headache, and tingling of the face, hands, and feet. She stated that a mass had appeared in her left neck six years ago and a smaller mass had appeared in her right neck four years ago. Both masses had a slow growth pattern. On physical examination she had a 3 cm in diameter mass in the left neck anterior to the sternocleidomastoid muscle and inferior to the mandible. There was a 2 cm in diameter mass in a similar position of the right neck. Both tumors moved laterally, but not vertically. Both had a bruit. A bilateral carotid arteriogram revealed an increased separation of the internal and external carotid arteries. There was also an unusual
collection of dye material about the internal carotid arteries which was thought to represent carotid body tumors. On December 14, 1962, a 4½ x 3 cm mass which was attached to the left common and internal carotid arteries, and adjacent to the vagus nerve trunk and internal jugular vein was excised by the surgical service. The gross specimen was a gray-pink, encapsulated, rubbery mass. Microscopically, there were large nests of cuboid or polyhedral cells, disbursted within thick vascular septa. The cells had an eosinophilic, granular cytoplasm and contained large, hyperchromatic nuclei.

In March, 1963, she was again complaining of excruciating pain and tenderness about the right ear without drainage. On physical examination a bluish red mass was present at the external auditory meatus. There was marked pre- and postauricular pain and tenderness and postauricular hyperemia. The mastoid x-rays exhibited destruction of the bone of the petrous pyramid. Because of the very large size of this tumor, she was started on a course of radiation therapy on March 13, 1963, and received a total of 4574 rads over the next seven weeks. Her pain has subsided, but the bluish red mass in the external auditory meatus persists.

MULTIPlicity

Since the first description of the carotid body by Von Haller and the glomus jugulare by Guild there have been numerous reports of tumors of these structures. Reports of multiple chemodeactomas occurring within the same patient are rare. Certainly the coexistence of tumors represented by this patient is extremely uncommon. James and Saleeby state the first reported case of bilateral carotid body tumors was by Middleton and Bierring in 1897. There have been several reported cases since that time of bilateral carotid body tumors: Chase, DeTarnowsky, Harrington, Lahey, Lewison and Weinsberg, Lund, Phelps, Rankin, James and Saleeby, Desai, McNealy, Rush, and Kroll.

Rosenwasser reported the first glomus jugulare tumor in 1945. Recorded cases of bilateral glomus jugulare tumors includes only that of McNeill. The incidence of multiple glomus jugulare tumors is low.

Lubbers in 1937 reported a patient having a carotid body tumor on one side and a tumor of the ear on the opposite side. The histological configuration of the two tumors was identical. Lubbers felt this case represented to the opposite the tumor of the tumor was a ca and report a co tumor in 194 Zacks, Hawk erer, Dibble, combinations er the patient had retroperitoneal ham reported ear, one of the

Chase in body tumors, a sister of the familial occurrence, in a review of the tumors were published and James and Warren publ Desai and Pate of the cases be of bilateral tumor occurring in published nine bilateral tumor brother to a pnow a total of logically conf individuals be twelve had bil

In 1963 F body tumors be more common with sporadic being carotid b
carotid arteries which ated. On December 14, o the vagus nerve tracheal surgical service. The d, rubbery mass. Micro-

ing of excruciating pain drainage. On physical at the external auditory articual pain and tender-

FAMILIAL-HEREDITARY TENDENCY

Chase in 1933 was the first to note familial occurrence of carotid body tumors. He described the pathology of a case of bilateral carotid body tumors, along with the histology of a carotid body tumor from a sister of the first patient. Rush points out that several additional familial occurrences of carotid body tumor have been reported, and a review of these cases discloses that in each family reported, bilateral tumors were present in at least one member of the family. McNealy and James and Saleeby reported similar familial cases. Lahey and Warren published cases of tumors in three siblings in one family. Desai and Patel reported five cases occurring in one family with two of the cases being bilateral. Lewison and Weinberg reported a case of bilateral carotid body tumors with three verified unilateral tumors occurring in three first cousins of the patient. Sprong and Kirby published nine cases of carotid body tumor in eleven siblings with one bilateral tumor. Rush added a case of bilateral tumors which is a brother to a patient reported by Lewison and Weinberg. There are now a total of eight tumors in six members of that family, all histologically confirmed. Kroll reported carotid body tumors in twelve individuals belonging to two generations of a family and five of the twelve had bilateral tumors.

In 1963 Rush concluded in his study of familial bilateral carotid body tumors that the occurrence of bilateral carotid body tumors is more common among patients with familial tumors than in patients with sporadic tumors. We would concur with this conclusion regarding carotid body tumors, but have been unable to find a similar
relationship in glomus jugulare tumors which are multiple or familial. The only reported case of familial glomus jugulare tumors is that of Goekoop in 1937 in which he described glomus jugulare tumors in three sisters. The incidence of familial glomus jugulare tumors is low.

SUMMARY

An unusual case of three chemodectomas occurring in the same patient is reported. The incidence of multiple chemodectomas occurring in the literature is reviewed. Evidence from the literature of the familial nature of chemodectomas is presented. The evidence from the literature for the apparent higher incidence of multiple carotid body tumors in patients with demonstrated familial tendency toward development of carotid body tumor is presented and discussed. Multiple or familial glomus jugulare tumors appear to be quite rare.

800 NORTHEAST 13TH ST.

Acknowledgement—Jerry Milton Ingalls, M.D., Resident in Surgery, performed the excision of the carotid body tumor.

REFERENCES


are multiple or familial, jugulare tumors is that of mus jugulare tumors in glomus jugulare tumors is occurring in the same chemodectomas occur. The evidence from the multiple carotid body tendency toward development and discussed. Multiple be quite rare.


A female patient, aged 25, presented with a mass in the neck. She was referred for further evaluation. On examination, a firm, nontender, palpable mass was palpated. Radiographs revealed an osteolytic lesion in the mandible. A fine-needle aspirate was performed, and the results were consistent with a metastatic lesion. A PET scan showed avid uptake in the neck mass.

The mass was biopsied, and the histopathological examination revealed a metastatic deposit of squamous cell carcinoma. Further imaging and laboratory tests were performed to assess the primary site of the malignancy. The patient was referred to a multidisciplinary team for further management.

Dr. Rebecca Smith, in consultation with the patient's oncologist, recommended a course of radiation therapy and systemic chemotherapy. The patient tolerated the treatment well and showed clinical improvement.

The patient was discharged with close follow-up appointments for continued monitoring of the disease. Regular check-ups included physical examinations, blood work, and imaging studies to assess the response to treatment.