PETROUS RIDGE CHEMODECTOMA (NONCHROMAFFIN PARAGANGLIOMA) SIMULATING MENINGIOMA

CASE REPORT

FRANCIS KRUSE, JR., M.D.*

Armed Forces Institute of Pathology, Washington, D. C.

(Received for publication December 10, 1959)

This report concerns a chemodectoma (nonchromaffin paraganglioma) that grew into the cranial cavity and, for a time, masqueraded as a meningioma. The object in reporting this unusual case is to emphasize that tumors of this type may arise not only from the carotid body or glomus jugulare, but also in the region of the petrous ridge of the temporal bone.

CASE REPORT†

A 63-year-old white male was admitted to hospital because of marked impairment of concentration and memory, of several months’ duration, and complaints of severe headache and episodes of vomiting during the week before admission.

Inquiry into the past history revealed that for a period of 30 years the patient had been subject to attacks, which occurred at least once a month, during which he would run about the house, scream, curse his wife, and occasionally strike her. He apparently had no subsequent recollection of these episodes, and during one 5-year period was entirely free of them while taking phenobarbital. Some 10 months before admission he began to have episodes of transient aphasia and, 2 months later, brief losses of consciousness with cyanosis and twitching of the right side of his face. At about this time his family noted that he had marked impairment of memory for recent and remote events, an inability to concentrate on his business, and periods of depression. Moreover, he complained of pains in the left side of his head and of sensing peculiar odors.

Examination. The patient was obviously confused and disoriented, and both recent and remote memory were poor. Nominal aphasia was detected. Pupilledema of 2 diopters was noted in both eyes. Right hemiparesis, most marked in the lower limb, was evident. The deep muscle reflexes were hyperactive on the right, and Babinski’s sign was elicited on this side. There was a positive Romberg sign, with a tendency to fall backward and to the right, and coordination of both upper and lower limbs was found moderately impaired bilaterally.

Laboratory Studies. Routine studies of blood and urine were not remarkable. On spinal tap the initial pressure was 400 mm. of water. The total proteins were 84 mg. per cent. Films of the skull showed slight demineralization of the sella turcica. Neither erosion nor eburnation was detected in the petrous ridge. A ventriculogram revealed a marked shift of the ventricular system to the right, suggesting a temporal-lobe mass on the left. The left lateral ventricle was not visualized.

Operation. A left frontotemporoparietal craniotomy was performed under local anesthesia. After opening the dura mater, which was tight, the temporal lobe was cannulated and a hard mass was located. Transcortical incision and removal of a portion of the inferior temporal gyrus revealed a firm, lobulated, encapsulated mass, which “appeared to be growing from the petrous ridge and passed backward and medially on to the tentorium, hollowing out the entire temporal lobe.” Despite severe bleeding, removal of what appeared to be the entire tumor (Fig. 1) was accomplished. An area of about 3 sq. cm. of the dura mater of the petrous ridge

* Present address: USAF Hospital, Lackland Air Force Base, Texas.
† From the Frazier-Grant Brain Tumor Collection, Armed Forces Institute of Pathology Accession #630218.
was removed with the tumor. The dura mater was free-grafted and a rubber-tissue drain was left in the bed of the tumor. The bone flap was replaced and the wound was closed.

Microscopic Examination. Microscopically the tumor consisted of nests of "epithelioid" cells, with the component cells of uniform appearance and size (Fig. 2A). Between the cell masses lay thin fibrous septa, which contained capillaries. A Gomori silver stain revealed strands of reticulin limited to the septa (Fig. 2B). In several parts of the tumor there were extensive areas of apparently recent necrosis. The architectural features were those of a chemoreceptor organ.

Postoperative Course. The patient recovered slowly. For a time he continued to be emotionally unstable, and his memory was poor. However, at the time of discharge from hospital these disturbances were less pronounced, and he was able to walk with some assistance and he could feed himself. About 3 months later, he fell while in the bathroom and was believed to have suffered a cerebral hemorrhage. He died in another hospital, where no autopsy was performed.

COMMENT

Most of the tumors of the chemoreceptor organ that have been reported have been readily related to the known sites of chemoreceptor-cell groups in the head and neck, i.e., the carotid body, glomus jugulare, paraganglion tympanicum, and the ganglion nodosum of the vagus. Depending on the sites of origin of the tumor, the clinical features have been fairly distinctive.

Tumors originating in the carotid body, located at the carotid bifurcation or a few millimeters higher along the internal carotid artery, usually present themselves as nontender masses at the angle of the jaw, are of slow growth, and show no signs or symptoms referable to the central nervous system. Chemodectomas arising from cells of the glomus jugulare, located in the adventitia of the jugular bulb, or similar groups of cells located along the course of the tympanic branch of the glossopharyngeal nerve (paraganglion tympanicum), usually induce otological disturbances, principally deafness and chronic discharge from the ear. Examination reveals red, polyp-
like tissue in the external auditory canal, which may hemorrhage freely when removal is attempted. Growth of these tumors within the middle ear often causes pronounced destruction of bone, and the tumor may extend into the posterior cranial fossa and give rise to neurological disturbances. Occasionally these disturbances antedate or arise coincident with the auricular disorders. In a review of 33 cases of tumor of the glomus jugulare, Siekert reported significant neurological abnormalities in 14. Ipsilateral cranial nerves were affected in all 14, nystagmus was observed in 2, and ataxic gait and incoordination of the ipsilateral limbs in 1. Disturbances re-

Fig. 2. (A) Representative field of the tumor showing masses of "epithelioid" cells and thin septa. Hematoxylin-eosin stain, X130. (B) Strands of reticulin, limited to the septa and outlining capillaries, are to be seen. Gomori silver stain, X165.
ported by other authors have included signs referable to the pyramidal tract,\(^3\)\(^7\) ipsilateral cerebellar disturbances,\(^2\)\(^3\) and papilledema.\(^4\)

Apparently only 3 authenticated cases of tumor arising from chemoreceptor cells in the ganglion nodosum of the vagus have been reported.\(^6\) In 2 of these cases, neurological symptoms were noted that were brought about by compression of cranial nerves and by encroachment upon the brain substance within the posterior fossa.

In the case here presented, the absence of roentgenographically demonstrable bone destruction and extension of the tumor into the middle cranial fossa, where it compressed the temporal lobe in the manner of a meningioma, suggests origin from chemoreceptor-cell groups of unusual location. LeCompte\(^6\) has stated that groups of chemoreceptor cells are probably present in several locations in the temporal bone. Guild,\(^2\) in a study of 44 pairs of temporal bones, noted an extreme diversity in the location of chemoreceptor-cell groups. In addition to the previously recognized locations, Guild noted these cell groups (1) along the tympanic branch of the glossopharyngeal nerve as far distally as the region at which the lesser superficial petrosal nerve is given off, and (2) along the auricular branch of the vagus as far distally as the descending part of the facial canal in the temporal bone. Lattes and Waltner\(^5\) have also shown that marked diversity exists in the location of the chemoreceptor-cell groups from which chemodectomas could conceivably arise. The unique location of the tumor in the present case, namely, along the surface of the petrous ridge, is attributed to origin from aberrant chemoreceptor cells related to the complex of chemoreceptor-cell groups present within the temporal bone. In the present case, the histological appearance and 30-year history of disturbances of temporal-lobe origin would indicate, as is true of chemodectomas in general, that the tumor was biologically benign.

**SUMMARY**

This report concerns a chemodectoma (nonchromaffin paraganglioma) that simulated a meningioma of the petrous ridge at operation. This case is unusual in that growth of the tumor apparently was limited to the cranial vault and caused compression of the temporal lobe, producing temporal-lobe-seizure phenomena. The probable relationship of this tumor to the tumors of glomus jugulare and paraganglion tympanicum is emphasized.

**REFERENCES**

1. Alexander, E., Jr., Beamer, P. R., and Williams, J. O. Tumor of the glomus jugulare with extension into the middle ear (nonchromaffin paraganglioma or carotid-body-type tumor). *J. Neurosurg.*, 1951, 8: 513–522.