CASE REPORTS AND TECHNICAL NOTES

INTRACRANIAL CERUMINOUS ADENOMA

LOUIS BERLIN, M.D.*

Neuropathology Laboratory of the Neuropsychiatric Institute,
University Hospital, Ann Arbor, Michigan

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Of the intracranial tumor masses responsible for the syndrome of progressive unilaterial deafness, the best known is the acoustic neurinoma. However, other tumors, either primary or metastatic, may occur in the same region and give rise to a similar syndrome. The most common primary tumors to be differentiated from an acoustic neurinoma are the meningiomas of the cerebellopontine angle, gliomas of the pons, and cholesteatomas. The metastatic tumors that invade that area are the carcinomas and abscesses, especially those arising from the external and middle ear, lymphoepitheliomas, angiomomas of the middle ear, carotid body tumors, malignant melanomas, and sarcomas of the temporal bone. To this list of tumors involving the 8th nerve should now be added the ceruminous adenoma.

The ceruminous adenoma is a rare tumor that usually arises from the ceruminous glands of the external ear. It is made up of tubules or alveoli of various dimensions which may become distended into cystic cavities. These may then present the appearance of cystadenomata. The epithelium of the tumor is generally composed of two layers of cells, an inner layer of cuboidal to cylindrical cells, and an outer layer of flat cells. The epithelium may be thrown into papillary folds within the cystic areas. The tumor is surrounded by a fibrous capsule and there are also numerous connective-tissue septa between the glandular elements.

Since the first description of this tumor by Haug in 1894, a total of 6 cases have been described. In every instance the tumor was present only in the external ear. It is the purpose of this paper to report for the first time the intracranial occurrence of a ceruminous adenoma and discuss the clinical and pathologic diagnostic problems which this presents.

CASE REPORT

J. S. (631642), a 26-year-old white farmer, was admitted to the University Hospital on Feb. 16, 1948, because of deafness in the left ear, and left facial paralysis. The first symptoms were noted 9 years previously, when he suddenly discovered during a telephone conversation that he was deaf in his left ear. One and a half years prior to admission there was noted also some flattening of the left side of the face and this progressed to a complete left facial paresis 8 months before admission. There was no associated headache, dizziness, tinnitus, ataxia or weakness. The rest of the past history and family history was non-contributory.

Examination. There was an almost complete, peripheral type of facial paresis which permitted the slow closure of the eyelid, and a complete nerve deafness on the left. The left tympanic membrane was hyperemic and the light reflex was absent. There was no other evidence of impairment of sensory or motor function anywhere, no ataxia, cerebellar dysfunction or increased intracranial pressure.

X-rays of the skull showed a destructive process involving the petrous portion of the left

* Now at Winter Veterans Administration Hospital, Topeka, Kansas.
temporal bone (Fig. 1) and obscuring the internal auditory meatus. There was a minimal irregular calcific density within the region of the lesion. This was interpreted as representing a malignant neoplasm arising at the base of the skull and invading the cerebellopontine angle. It was not considered to be the x-ray appearance of an acoustic neurinoma.

Operation. On Feb. 20, 1948, a left suboccipital craniotomy was performed by Dr. Max M. Peet. After the dura was opened, a tough gray circumscribed tumor was encountered which extended from the petrous ridge of the left temporal bone posteriorly to the tentorium cerebelli and superiorly for a small, but undetermined distance above the tentorium and inferiorly down to the pons. The tumor was seen to displace the left hemisphere of the cere-

![Fig. 1. Destruction of the petrous portion of the left temporal bone by the tumor. Note also the calcific density within the tumor.](image)

bellum downward and to the right. It was located entirely extradurally. Exploration of neighboring subdural and subarachnoid spaces failed to reveal any extension of the tumor. Because of the size of the mass, it was necessary to perforate it and thereby decompress it in order to facilitate removal. It was estimated that the tumor weighed 50 grams.

Postoperative Course. The immediate course was marked by some difficulty in swallowing and paralysis of the uvula. Bronchoscopy had to be performed on the 3rd postoperative day in order to aspirate the accumulated secretions. However, by the 8th day the patient was able to swallow and the weakness of the uvula had diminished. At the time of his discharge, on Mar. 8, 1948, there were residual 6th nerve weakness, complete facial paresis, nerve deafness, and partial paralysis of the uvula, all on the left side. The gag reflex was present and the patient was able to swallow liquids and soft foods. There was no other sensory or motor impairment and no ataxia.

Pathologic Report. The specimen consisted of a firm, fibrous, spherical mass, 2½ cm. in diameter, with numerous irregular knobs on the surface. Upon section the tumor was seen to contain numerous cystic areas filled with a brown, hemorrhagic fluid undoubtedly introduced during the operation. The tumor was basically an adenoma, composed of tubules which were often distended into cystic cavities. The epithelium of the tubules was composed of two layers...
of cells, an inner layer of low cuboidal to columnar cells and a surrounding layer of flattened elongated cells (Fig. 2). The larger cells contained a large, oval nucleus with a prominent nucleolus and a small amount of pale-staining vacuolated cytoplasm. The external layer was made up of poorly defined elongated cells with large elliptical nuclei. This epithelium rested upon fibrous connective tissue which formed the capsule of the tumor. Numerous septa intervened between the glandular epithelium. Many areas of the glandular tissue were distended into cystic cavities. Here the epithelium was thrown into papillary folds which had a central connective-tissue core (Fig. 2). The lumina of the tubules and cysts were filled with fresh blood and amorphous debris, but the exact contents could not be determined because of the perforation of the tumor during the operative procedure.

**Fig. 2. Left:** Demonstration of the cystic nature of the tumor and the epithelium thrown into papillary folds. Haemotoxylin and eosin, Zeiss planar 35 mm. **Right:** Cuboidal cells and surrounding flattened, elongated cells constituting the epithelium of the adenoma. \( \times 185 \).

**DISCUSSION**

The clinical findings of unilateral deafness and facial paresis combined with roentgenologic evidence of severe erosion of the petrous bone suggested the possibility of a malignant neoplasm at the cerebellopontine angle. However, the chief interest in this case is derived from the unusual tumor that was removed.

From the pathoanatomic point of view, the tumor could easily be confused with a papilloma of the choroid.\(^3\) The cuboidal epithelium regularly arranged on papillae of connective tissue suggested this diagnosis. However, the extradural location made such a diagnosis unlikely. Furthermore, closer examination revealed that the epithelium was not merely composed of a single layer of cuboidal cells as found in choroid. Instead, there was an inner layer of cuboidal to columnar cells and an outer layer of flattened cells analogous to the myoepithelial cells of sweat glands. In some areas the epithelium seemed piled up and appeared to be stratified. The depth of the connective tissue in the capsule and interalveolar septa was much greater than that usually encountered in papillomas of the choroid.

The mechanism by which this tumor arose within the cranial cavity can only be surmised. It has been suggested that the tumor may arise from aberrant glandular
tissues usually found in the external or middle ear. Further investigation of these areas will ultimately have to be done to confirm this hypothesis. Another mode of invasion into the cranial cavity possibly may be malignant degeneration of the tumor and subsequent erosion of the bone. However, there was no definite histologic evidence of malignancy.

SUMMARY

A case is reported of the intracranial occurrence of a ceruminous adenoma which heretofore has been reported as occurring only in the external or middle ear. The tumor was found extradurally in the region of the left internal auditory meatus where it produced a destruction of the petrous portion of the temporal bone, and a paralysis of the 7th and 8th nerves.

REFERENCES

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PAPILLOMA OF THE CHOROID PLEXUS

REPORT OF CASE

Everett O. Jeffreys, M.D., and Richard H. Ames, M.D.

Division of Neurological Surgery, Bowman Gray School of Medicine, Winston-Salem, North Carolina

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Papilloma of the choroid plexus is a relatively rare intracranial tumor. Less than 100 of these tumors have been reported, and the majority of these were disclosed at autopsy. As far as we have been able to determine from the literature, only 24 papillomas have been diagnosed surgically and of this group less than half the patients survived the immediate postoperative period.