Hamartomas of the Cerebellopontine Angle and Internal Auditory Canal

Report of Two Cases

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Most mass lesions of the internal auditory canal and cerebellopontine angle are neoplastic, expansile, and eventually symptomatic. We report two cases of neurogenic hamartomas at these anatomic sites. In a patient with unrelated auditory or vestibular symptoms, such a developmental lesion might result in false-positive posterior fossa contrast results. (Arch Otolaryngol 106:500-502, 1980)

With the rapid advances in neurotologic techniques in recent years, the anatomy and pathologic features of the internal auditory canal (IAC) and cerebellopontine angle (CPA) have taken on considerable practical significance. Diagnostic techniques have been refined to the point that mass lesions around 1 cm in diameter may be delineated at these sites with fair regularity. Surgical excision of these neoplasms, when they are still of such small dimensions, greatly enhances the chance of maintaining auditory and facial nerve function. Since the clinician is aware that these tumors are best diagnosed when symptoms are subtle or even ambiguous, the number of posterior fossa myelograms performed in the absence of an acoustic neuroma has increased, justifiably. Recent attention has been drawn to the 2% incidence of false-positive tomographic findings in the diagnosis of tumors of the IAC. Posterior fossa myelography is still the single most reliable technique of demonstrating all but perhaps very large tumors at this site. We report a potential cause of a false-positive posterior fossa myelogram, a coexistent neurogenic hamartoma of the IAC or CPA.

REPORT OF CASES

Case 1.—A 43-year-old man had primary biliary cirrhosis, hypersplenism, and secondary intestinal malabsorption. He experienced multiple right ear infections throughout his life and hearing was.

Fig 1.—Fusiform, unencapsulated mass (H) lies between intracanalicular portions of auditory and vestibular nerves (hematoxylin-eosin, original magnification \( \times 20 \)).
decreased on this side. There was no history of vertigo. At his initial visit, a right tympanic perforation and an aural polyp were seen. The polyp was removed and an audiogram showed a moderate conductive hearing loss on the right. Mastoid series at this time demonstrated a poorly pneumatized right mastoid without bony erosion. For the following five years, he had increasing problems with malabsorption and coagulopathies. During this period, his ear remained dry. He eventually died of upper gastrointestinal tract bleeding. The temporal bones were received ten hours after death, fixed in buffered formaldehyde solution, and decaledified in nitric acid. After embedding in celloidin, they were sectioned at 20-µm intervals, and every tenth slide was stained with hematoxylin-eosin. Subsequent sections were stained with PAS, with diastase controls.

Microscopic sections of the right tympanic bone revealed posterior-inferior perforation of the tympanic membrane and submucosal fibrosis with mineralization consistent with myringosclerosis. The mastoid was poorly pneumatized, and the middle ear and mastoid mucosa were thickened with multiple adhesions between ossicles and the otic capsule. The organ of Corti was present in all turns, but autolysis prevented adequate hair cell evaluation. Cystic degeneration of the stria vascularis was noted, and ganglion cells were reduced to 25% of normal in the apical regions and 50% in basilar regions. In the left temporal bone, there was similar cystic degeneration of the stria vascularis, but the reduction of ganglion cells was less marked.

In the right IAC, a compact, fusiform, unencapsulated mass was opposed to the intracanalicular portions of the vestibular, auditory, and facial nerves (Fig 1). The mass consisted of normal-appearing neurons and astrocytes; atypical or binucleate forms were absent. There was no evidence of inflammation, glial reaction, or compression of the adjacent structures (Fig 2). The IAC was of normal size and equal in dimensions to the opposite ear. Beyond the porus acusticus, the mass was closely applied to the glial sheaths of the cochlear nerve. The PAS staining of Schwann's cell basement membrane emphasized the distinction between peripheral nerve elements and the mass, which was of CNS origin. The normal-appearing neurons and glial elements and the absence of invasion or compression are characteristic of a hamartoma. No similar structure was seen associated with the pons or CPA in microscopic sections of the brainstem. The cochlear changes noted in the right ear are attributed to long-standing otitis media.

**Case 2.—** A 36-year-old woman was admitted to the hospital with jaundice, fever, and gastric ulcers. She also had Horner's syndrome on the right, which was unrelated to her gastrointestinal tract disease. Exploratory laparotomy established a diagnosis of postnecrotic cirrhosis with portal hypertension. During the next year and a half, she was treated for pulmonary edema, oliguria, proteinuria, and ascites. She experienced paresis of the right arm after a right subclavian hematomata involved the brachial plexus. Prior to death, there were signs of diffuse intravascular coagulopathy, hepatic encephalopathy, and sepsis.

Findings at autopsy included macronodular cirrhosis, varices, and chronic duodenal ulcer with candidiasis. Diffuse bilateral pneumonitis and generalized atherosclerosis were noted. In the CNS, there was generalized cerebral swelling, and a 10-mm nodule intimately associated with the eighth nerve in the right CPA (Fig 3).

Microscopic examination showed Alz-
heimer's type 2 gliosis in the basal ganglia and dentate nucleus. A section of the lesion in the CPA contained mature ganglion cells, astrocytes, and Rosenthal's fibers. These elements were arranged in a pattern more consistent with a neuronal and glial hamartoma than a neoplasm such as ganglioglioma (Fig 4).

COMMENT
Most mass lesions described in the IAC and CPA are neoplastic, expansile, and eventually symptomatic. When small, they cause subtle dysfunction of the auditory and vestibular systems. When large, they may produce a CPA syndrome, with involvement of cranial nerves V through IX, and pontine, cerebellar, or medullary dysfunction. Acoustic neoplasms make up 90% of the lesions at these sites. Meningiomas are second most common, followed by gliomas and primary cholesteatomas. Complete differential diagnoses are as follows: acoustic neuroma; meningioma; glioma—ependymoma and astrocytoma (pontine or cerebellar); primary cholesteatoma; vascular neoplasm; vascular malformation—aneurysm of anterior inferior cerebellar artery and tortuous basilar artery; metastatic carcinoma; arachnoid cyst or hyperplasia; ceruminoma; lipoma; chordoma; gamma; choroid plexus papilloma; and hamartoma.

Hamartomas... n the other hand, are not neoplastic, but represent an abnormal arrangement or grouping of tissue normally found in the organ. Choristomas are related malformations, but consist of elements normally foreign to the locale. The most common neurogenic hamartoma encountered by the otolaryngologist is the nasal glioma. Mesenchymal hamartomas have been described in the middle ear cleft, causing Eustachian tube obstruction. A vascular hamartoma of the pinna has also been reported.

To our knowledge, these are the first reported cases of neural hamartoma involving the IAC and CPA. Their significance lies in the potential danger they pose for an individual with coexistent, unrelated auditory or vestibular symptoms.

References