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Acoustic neuromas and meningiomas. Histopathological aspect
A post mortem study on temporal bones

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Abstract. – **The histopathological changes in the temporal bones of three deceased donours individuals, two with concomitant acoustic tumors and one with concomitant petroclival meningioma have been studied. The differences between neuromas and meningiomas are examined. The different clinical relevance and the different therapeutic guidelines are discussed.**

Key Words:

Temporal bone, Acoustic tumor, Surgery, histology, Cerebello pontine angle, Magnetic resonance.

Introduction

Neoplasms arising primarily in the inner ear are unusual. More usually they reach it either by direct invasion from adjacent structures or as blood-borne metastases by the bloodstream.

We have studied the histopathological changes in the temporal bones of two deceased individuals with concomitant acoustic neuroma, one on the right and the other on the left side.

Materials and Methods

These patients were donors and agreed during their life to donate *post mortem* their temporal bones to the House Ear Institute Los Angeles, CA USA as a contribution to a better knowledge of temporal bone diseases.

We have removed the temporal bones in our usual way[1].

Results and Discussion

Acoustic neuromas

The cellular constituents of the inner ear, apart from bone are, for the most part, fully differentiated non-mitotic structures -nerve cells and sensory epithelia- so that neoplasms would not be expected to arise in them. Primary neoplasms are indeed rare except for acoustic neuroma (schwannoma).

Acoustic neuroma is the most frequent neoplasm in the temporal bone. It is a schwannoma of the eighth cranial nerve, but the term neuroma is in most frequent usage. Other synonyms are also encountered in the literature, such as acoustic neurilemmoma, acoustic neuronoma and acoustic neurofibroma.

Site. The neoplasm is stated to arise most commonly at the glial-neurilemmal junction of the eighth nerve, which is usually within the internal auditory meatus. When seen at surgery or autopsy, however, acoustic neuroma is found to occupy a much greater part of the nerve. In most cases it is the vestibular division of the nerve which is affected whereas in a few the cochlear division is the source of the neoplasm. Growth takes place from origin, both centrally onto the cerebello-pontine angle, and distally along the canal. A case has been described in which the acoustic neuroma arose from the intravestibular portion of the nerve[2]. Nager[3] reported a small schwannoma arising in the cochlea of a case of Paget's disease affecting the temporal bone and it has been noted a similar lesion in the same condition. Acoustic neuroma is usually unilateral but may be bilateral. In the large series reported by Erickson et al.[4] 129 cases were unilateral and 11 bilateral.

Incidence. The neoplasm may grow for years without causing symptoms and may be first diagnosed at *post-mortem*. Hardy and Crowe's classical finding[5] of six acoustic neuromas in 250 temporal bones was corrected on review to four in 883 *post-mortem* by Leonard and Talbot[6]. Females represent 64% of cases of the neoplasm. It may appear from between 13 and 72 years of age with a mean of 45 years[4].

Clinical features. Although the neoplasm usually grows from the vestibular division of the eighth nerve, most patients have hearing loss and tinnitus at presentation, while only a few complain of vertigo. Defective function of both the cochlea and the labyrinth are, however, elicited more often by the sophisticated procedures of audiometry and caloric testing.

Appearances. The neoplasm is of variable size and of round or oval shape. The larger tumors often have a mushroom shape with the two components, the stalk – an elongated intratemporal part – and an expanded extratympanic part. The bone of the internal auditory meatus is often widened funnelwise by the slow growth of the neoplasm. The surface of the neoplasm is smooth and lobulated. The cut surface is yellowish, often with areas of haemorrhage. The nerve of origin, usually the vestibular division of the eighth nerve, may be identified on the surface of the tumour and is often stretched by the latter. A fluid exudate may be observed in the cochlea and vestibule.

Microscopic appearances. Acoustic neuroma has the features of a neoplasm of Schwann cells with arrangement of the cells into a specific, almost organoid pattern. It is customary to define two areas of different appearance in the tumour as Antoni A and Antoni B types (Figure 1). The tumor cells in both areas are Schwann cells or their derivatives. Antoni A areas show the spindle cells of the neoplasm closely packed together. There is a tendency to palisading of nuclei, i.e. formation of nuclei of cells into rows which are aligned at right angles to the cells Verocay bodies may be present in the Antoni A areas. These are whorled formations of palisaded tumour cells resembling tactile corpuscles. Palisading and Verocay bodies may, however, be absent. The degree of cellularity of the neoplasm can be high or low. In the latter case there may be areas of fibrous tissue. The spindle cells frequently show a moderate degree of pleomorphism, hut mitotic figures can rarely be seen in the acoustic tumours. The presence of pleomorphism does not denote a malignant tendency on the part of the neoplasm. Antoni B areas show a loose reticular pattern, sometimes with histiocytic proliferation. These areas are rarely prominent in acoustic neuromas. Thrombosis and necrosis may be present in some parts of the neoplasm. Granular or homogeneous fluid exudate is usually present in the perilymphatic spaces of the cochlea and vestibule. This may arise as a result of pressure by neoplasm on veins in the internal auditory meatus (Figure 2).

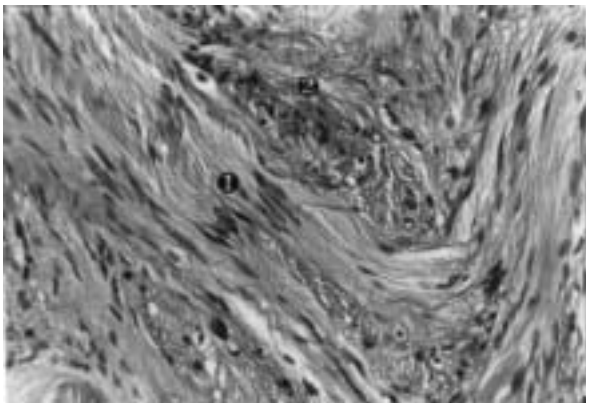

Figure 1. Acoustic tumor containing dense and Antoni type I and vacuolar and Antoni type II tissue. Palisading of the nuclei (1) and Verocay bodies (2) are visible. ×430

Electron microscopy. The ultrastructure of acoustic neuroma is characterized by the

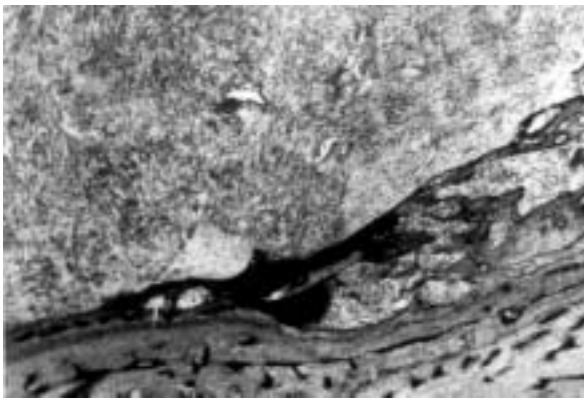

Figure 2. Vestibular schwannoma arising from the utricular nerve. ×46

presence of Schwann cells in both Antoni A and B areas. These cells have a network of thin, interdigitating processes covered by a fine basal lamina in all areas. The collagen of acoustic neuromas frequently shows fibres of a giant size. These have a periodicity of cross bands of up to 150 nm as compared with the periodicity of normal collagen of 64-71 nm[7].

Natural history. Acoustic neuromas are always benign neoplasms. They may cause serious symptoms and even death due to damage to temporal bone and cerebral structures, but develop no malignancy.

Meningioma

Meningiomas are usually intracranial masses. They arise from arachnoid *villi*, which are small protrusions of the arachnoid membranes into the venous sinuses. Arachnoid *villi* may be found in parts of the temporal bone, including the inner ear, and on occasion meningiomas may arise from these structures as primary neoplasms of the inner ear region. The most likely position for a primary inner ear meningioma is in the wall of the internal auditory meatus, where arachnoid *villi* are normally frequent[8].

The histological appearances of a meningioma are those of a tumour with a whirled arrangement of cells: meningotheliomatous if the tumour cells appear epithelioid, psammomatous if calcification of the whirled masses is prominent and fibroblastic if the tumour cells resemble fibroblasts. Meningiomas as well as acoustic neuromas may appear in the inner ear in the bilateral acoustic neuroma syndrome[9]. The meningioma is a slowly growing tumour of the temporal bone which has had a reputation for complete benignity. Our experience with this neoplasm, has, however, indicated its propensity for local recurrence and invasion (Figure 3).

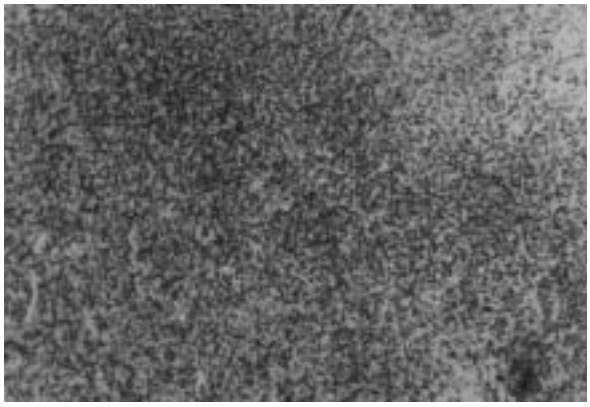

Figure 3. Meningioma. ×115

Conclusions

Neurinoma of the cerebello pontine angle is certainly a benign tumor. Our procedure of choice in case of serviceable hearing-puretone average threshold better then 50dB and speech discrimination score greater then 50%, referred as 50/50 rule, is middle fossa approach in case of intracanalicular tumor, retrolabyrinthine approach in case of extrameatal tumor. In case of no serviceable hearing or tumor greater then 2.5 cm extrameatal, we adopt translabyrinthine approach.

It is very important to differentiate meningiomas from neuromas. MRI in thin sections using T1 and T2- weighted techniques with and without gadolinium contrast agent shows vascular encasement or displacement. In the T2 weighted images soft tumors appear white in contrast to firm tumors. Cerebral angiography is performed on a routine basis and has three important functions: first, to define the relationship of the tumor to vascular structures, second to evaluate the vascular supply of the tumor and subsequent embolization if this is deemed possible and third to evaluate tolerance to temporary vascular occlusion clinically and quantitatively using Xenon/CT cerebral blood flow techniques. Petroclival meningiomas derive their blood supply predominantly from the meningohypophyseal branch of the internal carotid artery. When the tumor is supplied by the external carotid artery (ascending pharyngeal, middle meningeal branches, occipital artery), preoperative embolization of these external carotid artery feeders can be extremely helpful with tumor excision by reducing intraoperative bleeding. Embolization of the meningohypophyseal arteries is occasionally feasible.

The great difference between neuromas and meningiomas is that the first grow leaving vascular and neural structures outside their capsule, while the second grow encasing

neurovascular structures. This is why we agree with Al Mefty: Meningiomas of vault are angels, meningiomas of petrous bone are devils.

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