Intracanalicular meningioma

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Introduction

A combined lesion of the VIIth and VIIIth cranial nerves must carry a sinister connotation, especially in the absence of skull trauma or inflammatory middle ear disease. The most common cause of this unusual association of signs is a space-occupying lesion of the cerebello-pontine angle, most frequently an acoustic neurinoma.

This report concerns a patient in whom facial paralysis and deafness were due to intracanalicular meningioma, a condition which is both extremely rare and difficult to manage.

Case report

History

J.M., a male, aged 14 years, was seen on 6 November 1973 with the complaints of left-sided facial weakness and deafness for the past four and two years respectively. There was no history of vertigo, tinnitus or headache.

Past history

J.M. was treated elsewhere previously by three courses of ACTH for suspected uncomplicated left-sided Bell's palsy over a period (20 November 1969 to 25 July 1972) without much improvement. He was then referred to the Otology Department of the same hospital, when the left-sided deafness was detected. Straight X-rays of the petrous bones failed to reveal any structural abnormality of the internal auditory meatus.

Decompression of the mastoid segment of the facial nerve was carried out on 8 November 1972. This part of the nerve appeared to be normal. Tissue taken from a granular area over the horizontal segment of the facial nerve was examined histologically and reported on as follows: 'Shows rather spindle-shaped cells which by their arrangement are suggestive of elements of the dura. Search through several levels has revealed a tiny psammoma body.'

Lack of desired clinical improvement resulted in referral of this patient to the Eye and Ear Clinic of the Royal Victoria Hospital, Belfast.

Examination

Clinical examination showed an almost complete lower motor neurone type of VIIth nerve palsy and complete deafness in the left ear. There was no nystagmus, and tests of clinical balance were performed normally. The remaining ear, nose and throat findings were normal.

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Investigations

(*a*) Pure-tone audiogram showed total deafness in left ear.

(*b*) Vestibular function analysis showed complete loss of labyrinthine function of the affected side. There was no spontaneous nystagmus. Position tests were negative.

(*c*) Electromyographic study revealed severe denervation of facial muscles and the nerve excitability test was also negative on the left side. The secretomotor function of this nerve was also absent.

(*d*) X-ray polytomography showed dilatation of the lateral end of the left internal auditory canal representing a 50 per cent increase in size as compared with the right side.

(*e*) CSF protein content was normal.

(*f*) Myodil cisternography revealed absence of filling of the left internal auditory meatus.

Diagnosis

A diagnosis of a space-occupying lesion of the left internal auditory meatus affecting the VIIth and VIIIth cranial nerves was made.

Surgical treatment

(19 December 1973.) Exploration left internal auditory meatus via middle fossa approach (House).

The greater superficial petrosal nerve was followed posteriorly to the geniculate ganglion whose surface had a distinctly abnormal granular appearance.

Further removal of bone uncovered the internal auditory meatus completely filled with tumour tissue. Attempts to separate the peripheral end of the facial nerve from the tumour mass were unavailing and eventually the tumour was removed piecemeal leaving the proximal stumps of the branches of the VIIth and VIIIth nerves. The geniculate ganglion was also removed with the labyrinthine portion of the VIIth nerve. The walls of the internal meatus were taken back in all directions for about 2 mm. in order to lessen the risk of leaving tumour extensions and the inner end of the meatus plugged with a free temporalis muscle graft.

On receipt of a histological report indicating meningioma, a search of the relevant literature was undertaken to obtain guidance regarding the future management of this case. In a most erudite and comprehensive survey, Nager and Masica (1970) emphasized the potential of this tumour to spread throughout the temporal bone. It was clear that there was a high risk of residual disease in this patient and therefore a second operation through a postaural transmastoid approach was carried out on 2 January 1974.

In this, most of the remaining petrous temporal bone was systematically removed by drilling. Islands of what appeared to be tumour tissue were located in the bone anterior and medial to the geniculate ganglion and in the bone adjoining the anterior and inferior walls of the internal auditory meatus. A small piece of granular tissue located in the ampullary end of the lateral semi-

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circular canal and all the remaining facial nerve between the stylomastoid foramen and the internal auditory meatus were sent for histological examination. Recovery was uneventful and the patient was discharged on 10 January 1974.

Pathology report

Biopsy specimens were received on 19 December 1973 and 2 January 1974. On each occasion tissue was embedded in paraffin and sections were stained with haemotoxylin and eosin. The initial specimens (19 December 1973) were taken from the internal auditory meatus and the region of the geniculate ganglion. Each consisted almost entirely of tumour. The tumour was a well-differentiated meningioma of the syncytial type in which there was a predominant whorled pattern with extensive psammoma body formation. There was slight focal calcification. Mitotic figures were not seen.

At operation on 2 January 1974, four specimens were sent for histology: (*a*) geniculate ganglion region; (*b*) lateral semicircular canal; (*c*) facial nerve and stapes; (*d*) wall of internal auditory meatus. The specimen from the facial nerve consisted almost entirely of meningiomatous tissue and in the geniculate ganglion region a few fragments of tumour could be identified although most of the tissue showed reactive changes, presumably resulting from previous surgery. The other sites examined did not contain tumour.

Discussion

Meningioma, a tumour generally held to arise from the arachnoid villi and often situated in close relationship to the major intracranial venous sinuses, is comparatively rare in the posterior fossa. It accounts for about 6 per cent of all tumours of the cerebello-pontine angle of which about half arise in the internal auditory meatus.

Tumours arising from the internal auditory meatus cause pressure effects at first on the contents of the meatus and later on the brain stem, cerebellum and nearby cranial nerves. In this patient the tumour did not extend medial to the porus of the internal auditory meatus and thus the symptoms and signs were limited to those arising from direct involvement of the VIIth and VIIIth nerves within the temporal bone. Headache, Vth nerve signs and cerebellar involvement were therefore not present. Several unusual aspects of this case deserve emphasis.

1. The age of the patient. This was much less than that reported by Nager (1970) who found the average age of reported cases was 45 years.

2. Nager drew attention to the propensity of intracanalicular meningiomas to develop small but potentially threatening extensions into the labyrinth and the middle ear. Thanks to Nager's report we were alerted of this danger and carried out a second, and as it proved, vitally important operation which revealed such extensions.

3. Reports of meningioma in the middle ear are extremely rare (Chouard *et al.*, 1972). Whether they can *ever* have origin from the structures of the middle ear is debatable: the presence of ectopic arachnoid tissue in the tubo-tympanic cleft has not been reported. In reality, it seems more likely that meningioma in the middle ear is a lateral extension from a site of origin in the internal auditory meatus.

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4. The CSF protein findings in deafness due to retrocochlear lesions must be evaluated in the knowledge that normal CSF protein levels do occur when a meningioma is confined to the internal auditory meatus.

5. Our experience in this case underlines the contribution towards an early diagnosis of investigative aids such as polytomography and contrast studies of the posterior fossa. It also emphasizes the inestimable value of reports such as that of Nager and Masica (1970) which drew our attention to the urgent need for a second more radical operation in this patient. A satisfactory outcome in such cases is largely due to these factors in combination with the pioneering endeavours of surgeons such as William F. House (1961) who developed the microsurgical middle fossa approach to the internal auditory meatus. The desirability of a combined otologic, neurosurgical and pathological team to deal with such a complex situation as occurs in intracanalicular meningioma is well demonstrated in this case.

For a complete reading on this subject, readers are referred to the report of Nager and Masica (1970).

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