Malignant Auditory tumors

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objectives

1-Squamous cell carcinoma of external ear

2-Embryonal rhabdomyosarcoma

3-Vestibular schwannoma
Squamous cell carcinoma of external ear

Definition
malignant tumour of stratified squamous epithelium It represents 55% of malignant tumors of the pinna

Epidemiology
The average age at diagnosis is 65-70 years for the pinna lesions and there is a male predominance

Etiology
Actinic over exposure and frostbite have been suggested as causes of the pinna lesion. The canal tumours have been linked with the same tumour type in the middle ear as possibly resulting from prolonged chronic inflammation

localization
the majority of squamous cell carcinoma of external ear arise on the pinna a lesser number arise in the external canal

External ear sites of involvement in the pinna in a study of 52 patients show

<table>
<thead>
<tr>
<th>Number of Patients</th>
<th>Site</th>
</tr>
</thead>
<tbody>
<tr>
<td>27</td>
<td>Helix</td>
</tr>
<tr>
<td>11</td>
<td>Posterior auricle</td>
</tr>
<tr>
<td>6</td>
<td>Antihelix</td>
</tr>
<tr>
<td>3</td>
<td>Triangular fossa</td>
</tr>
<tr>
<td>3</td>
<td>Concha</td>
</tr>
<tr>
<td>2</td>
<td>Lobule</td>
</tr>
</tbody>
</table>
Prognosis and predictive factors

Squamous cell carcinoma of the pinna is an aggressive disease with a high propensity for local recurrence.

The outcome of the disease following surgical excision is related to the clinical stage at presentation, the higher the stage the worse the outcome.

Metastatic spread of squamous carcinoma of the pinna and external auditory meatus to lymph nodes is unusual.

Lesions arising in the canal have a worse prognosis because of the late diagnosis and invasion of adjacent structures.

Squamous cell carcinoma of middle ear

Etiology

An origin from long-term chronic inflammation of the middle ear has been suggested. However, malignant neoplasia in its earlier stages has clinical features similar to those of chronic otitis media. More over biopsy is not usually carried out during surgery when a diagnosis only of otitis media has been made. Therefore, longstanding squamous carcinoma of the middle ear may go undiagnosed.

Localization

The neoplasm soon expands to involve much of the middle ear.

There is extension by tumour through the bone on the medial wall of the Eustachian tube to infiltrate the perineurium of nerves in the carotid canal.

The tumour also penetrates the thin layer of bone between the posterior mastoid air cells and the Dura with subsequent invasion along the Dura and into the internal auditory meatus.

Clinical features

This tumour is usually advanced at presentation. The patient usually complains of pain in the ear, bleeding and a serosanguinous discharge from the ear canal. Seventh nerve palsy is an important sign indicating infiltration beyond the middle ear.
Embryonal rhabdomyosarcoma

Definition
A primitive malignant tumour with phenotypic and biological features of embryonic skeletal muscle.

Epidemiology
Rhabdomyosarcoma is rare in any part of the body. There is a distinct group arising in the head and neck of children, often very young, with a predilection for the palate, middle ear and orbit.

CASE REPORT
A boy, aged 4 years, presenting otalgia and irritability for the last 30 days where was diagnosed an otitis media. After a short course of antibiotics and analgesics the child developed deviation of the eye to the right, facial paralysis, and hearing impairment on the right.

The neurologic examination findings included facial and abducens nerve palsy. Otoscopic examination showed a fragile and reddish tissue in the external tympanic membrane was intact presenting hyperemia and bulging. Pure-tone audiometry detected 45-dB hearing loss in this ear. There was no detectable lymphadenopathy.

A computed tomography (CT) of the head showed destruction of the petrous part of temporal bone and right carotid canal associated another lesion with soft-tissue component in the right frontal bone.
MRI T2 weighted images shows hyperintense lesions with soft tissue component located in the petrous part of the right temporal bone extending to the sphenoid and clivus (A,B,C) and associated with lesion of the right frontal bone (D)

MRI T1 weighted images showed hypointense lesion located in the petrous part of the right temporal bone (A) with contrast enhancement (B,C) and associated with lesion of right frontal bone (D)

The impairment of the right abducens nerve has occurred by involvement of Dorello canal and the facial and vestibulocochlear nerve by involvement of the internal acoustic meatus and the ipsilateral tympanic cavity.

Biopsy of the right external acoustic meatus showed chronic otitis externa with granulation tissue and rare atypical cells. The immunohistochemical methods confirmed, through the antibody expressions for desmin and myogenin, the diagnosis of embryonal rhabdomyosarcoma.

The tumor was resected by radical mastoidectomy via postauricular with meatoplasty and the child was started on multi agent chemotherapy and radiotherapy.
Vestibular schwannoma

Definition
A benign nerve sheath tumour arising in the internal auditory canal.

Synonyms
Acoustic neuroma, acoustic neurinoma, neurilemmoma

Epidemiology
Vestibular schwannoma is the most common neoplasm of the temporal bone. Unilateral vestibular schwannoma accounts for 5-10% of all intracranial tumours and for most of the cerebellopontine angle tumours. It is found in about 0.8% of consecutive adult necropsies. The age at presentation is the fifth or sixth decade. It also is seen in younger people in association with neurofibromatosis type 2.
**Etiology**

Solitary vestibular schwannoma occurs sporadically, and does not seem to be associated with a gene mutation. The etiology is unknown.

**Localization**

Vestibular schwannoma was formerly considered to arise most commonly at the glial-neurilemmal junction of the eighth cranial nerve. Such a site of origin has now become doubtful. In one study of five temporal bones with small vestibular schwannomas, the tumour arose more peripherally. The vestibular division of the nerve is usually affected. Rarely, the cochlear division is the source of the neoplasm. Growth takes place from the site of origin of the tumour, both centrally onto the cerebellopontine angle and peripherally along the canal.

Vestibular schwannoma is usually unilateral, but may be bilateral, in which case the condition is neurofibromatosis 2.

**Clinical features**

Progressive unilateral hearing loss (90% of patients) and tinnitus (70% of patients) are the clinical manifestations, due to cochlear involvement. Less common symptoms are: headache, vertigo, facial pain and facial weakness. The neoplasm may grow slowly for years without causing symptoms and may be first diagnosed only at post-mortem. Diagnosis is usually made by MRI scanning. In small, slowly growing tumours, an option for management is non-surgical, using MRI scanning at intervals to observe growth.

Surgical removal may be carried out by drilling from the external canal through the temporal bone or by craniotomy and middle fossa approach to the internal auditory meatus, or by stereotactically guided gamma knife surgery.
Small vestibular schwannoma (S) in autopsy temporal bone specimen. It is arising from the vestibular division of the eighth nerve and causing a small indentation only of the bony wall of the internal canal.

MRI of bilateral vestibular schwannomas in a 32 year old man with NF2.

Thank you