XII./1.: Tumours of the ear

XII./1.1.: Tumours of the external ear

XII./1.1.1.: Precancerous tumours

Cutaneous horn

Usually develops on the concha of aged persons.

Figure 1.: Cutaneous horn

Senile keratosis

Generally it is a flat, yellowish-brown smooth mass with indistinct borders that stands out of the surface of the skin. Often carcinoma spinocellulare develops from it. Both lesions or the fast growth of the benign melanoma indicate malignisation.

XII./1.1.2.: Malignant tumours

Basalioma (Basal cell carcinoma)

Its most frequent spot of appearance is the edge of the helix, often the top of the concha. Probably long exposure to intensive sunshine contributes to its appearance; it is typically an illness of old agricultural labourers and often it is bilateral. A small, slowly growing mass appears on the rim of the concha. As it is itching the patient will scrape off the crusted epithelium, the epithelium will grow again, and finally ulcerates. The wound will not heal and slowly, unstoppably, it will grow. With indolent patients it may reach a fair size by the time the patient shows it to a doctor. The surgical excision of the concha rim basalioma has very good results.
Adenocarcinoma

It can originate from the glands of the skin of the acoustic meatus or it may come from degenerated originally benign ceruminoma.

Planocellular cancer

It may appear anywhere on the concha in the shape of one uneven raised edge infiltrated ulcer. It is most frequent in the cavum concha and in the external acoustic meatus.

Malignant lymphoma

Generally it spreads from the parotis to the external meatus.

Sarcoma

They appear rarely, and the appearance on the external ear is rare. Osteo- and angiosarcoma may appear.
Rhabdomyosarcoma

It can occur at a very early age, even in newborn infants, most frequently under the age of six, but up to the age of 14. In the external meatus a polyp-like mass appears which bleeds easily and grows fast, in some cases it even protudes from the external meatus.

Malignant melanoma

It appears as a small, fast growing, painless dark tumour or it may come from a pigmented naevus already present at birth. The quick growth of the lesion may indicate malignity.

Therapy.

Surgical removal, irradiation and citostatic treatment systemically or through regional supplying arteries by perfusion. The therapy to be used depends on the character of the tumour, its place and size, and the age, general status and cooperation of the patient.

Metastatic development

With the exception of basalioma, malignant tumours, depending on their position, develop metastases early to the retroauricular and neck lymphatic glands; tumours in the frontal regions and at the entrance of the external meatus disseminate to the praeauricular, the lymphatic glands attached to the parotis and the base of the skull. This is the reason why the ablasic removal of relatively small tumours also demand a rather large surgical intervention (extensive resection, neck dissection), and in the case of forward progression the parotis needs to be resected, together with the n. facial in it. A great number of the patients do not accept this solution or at least postpone the operation, and this way they miss the most favourable time for the operation.
Carcinoma in the external meatus will soon involve the tympanic cavity and the processus mastoideus and progresses toward the base of the skull, therefore surgical ablation requires petrous bone resection. Modern radiation therapy procedures and cytostatics have improved results, nevertheless advanced state malignant tumours have, with the exception of basalioma, a rather poor prognosis.

XII./1.2.: Tumours of the middle ear

XII./1.2.1.: Cylindroma (adenocystic carcinoma)

In the past this type of tumour used to be considered semimalignant. It can start from the salivary glands of the tympanic cavity. Clinically it can be mistaken for discharge from the ear and torpid middle ear inflammation. Characteristically the tumour spreads along the nerves, so one can primarily count for the facial nerve being coated and the spreading will follow the facial nerve. In case of early detection irradiation therapy following surgical removal may bring good results.

XII./1.2.2.: Middle ear carcinoma

In the majority of cases it is extremely difficult to decide whether the carcinoma appeared in the middle ear and spread from there into the external auditory meatus or the carcinoma or the flattened epithelium is spreading into the cavity system of the middle ear. Early detection is further complicated by the fact that this carcinoma generally appears together with chronic middle ear suppuration and the symptoms (otorrhoea, hearing loss) have been present before too. Alarming symptoms are the onset of pain and perhaps facial paralysis. Histological examination confirms the diagnosis. The prognosis is generally very bad. Ablastic surgical solution even through petrosa resection is not always feasible because the tumour often progresses to the tip of the petrosa, or to the middle scala through the tegmen tympanin. In addition to the operation irradiation and citostatic therapy can be considered.

XII./1.3.: Tumours of the internal ear

Acoustic neuroma

Schwaann-cell based tumours constitute 7-10% of brain tumours, 40% of back scala tumours, and more than 70% of cerebellopontine angle tumours. It is very rare in childhood, most tumours are found between 30-50 years of age, but the period of hearing deterioration can be very long, it may even be 30 years.

The VIII brain nerves as they leave the brain stem are covered for about 10 mm by neuroglia and having bored the way through the pia mater they run on in the Schwann cell made sheath. Tumours form most often at the neuroglial-neurolemmal junction, at the neurolemmal stretch of the upper vestibular nerve, inside the internal meatus. Consequently the correct name would be: vestibular nerve schwannoma. The tumour can appear on the acoustic nerve and the facial nerve but that is much less frequent. The tumour is benign and stems from one direction. The tumour has clear borders, does not infiltrate the tissues nearby, it has a sheath. The tissue structure is characterised by the clustered or swirling formation of elongated cells, at places loose reticular structure and
rounder cells appear. Fatty degeneration, haemosiderin increase and cyst formation can occur. Within the tumour the n. vestibulocochlear fibres degenerate.

Schwannoma must be told apart from the clinical picture of neurofibromatosis 2 (NF 2), which is an autosomal dominantly hereditary illness, affecting both sides.

The tumour grows slowly and compresses neighbouring vein and nerve systems, fills the internal meatus, steps into the pontocerebellar cysytem, while it is widening the wall of the internal acoustic meatus, and reaching the brain stem and the cerebellum it dislocates the pons, the myelencaphalon, and the cerebellum. The liquor circulation disorder created by the growing tumour leads to increased intracranial pressure. The size of the tumour does not always match the gravity of the symptoms.

The stages of the illness:

**Stage I:** Small intrameatal tumours, symptoms indicating n. cochleovestibular and n. facial impairment.

**Stage II:** Less than 2.5 cm tumours stepping out of the internal meatus, the n. trigeminus is affected.

**Stage III:** Large tumours, 2.5-5 cm, affecting the a n. glossopharyngeus, a n. vagus and n. accessorius, with cerebellar or brain stem symptoms.

**Stage IV:** Tumours over 5 cm, brain stem compression, prostrate state, eye fundus stasis, neurological symptoms

This slow growing tumour does not have any symptoms for a long time. The first, and even for years the only symptom is tinnitus, that is why patients with one sided tinnitus need to be appropriately examined. An indication of the impairment of the one sided, high pitch localised neural hearing loss, with no recruitment, bad speech comprehenson, the stapedius reflex shows increased fatigue.

Brain stem elicited response test (BERA) allows early diagnosis. The latency of V wave lengthens compared to the average and the opposite side, the I-V interpeak latency is 0.5 ms longer than the normal average and compared to the opposite side it is 0.3 ms longer.

Larger tumours cause greater discrepancies, greater latency figures and the other waves are either deformed or missing. During electrocochleography cochlear microphonia are normal, hair cells are working, only the transmission of the stimulus is hampered. Otoacoustic emission is normal. Due to the place of the tumour clearly retrolabyrinth-like symptoms can be expected but the intrameatal tumour through compressing the veins causes endolabyrinth-like hearing loss or Menière-like symptoms too. Rarely the circulation disorder can cause as introductory symptoms sudden deafness or facial paresis.

The slow deterioration of vestibular functioning causes mild indistinct disturbance of balance. The failure of one-side of the function is continuously compensated, this way causing hardly any complaint.

With open eyes there is no spontaneous nystagmus or any loosening procedure, e.g. after head shaking the nystagmus can be detected as it lashes towards the healthy side. With closed eyes latent spontaneous nystagmus can be registered using electronystagmography.
With standing, walking tests some slight insecurity can be seen. The kaloric reaction is severely limited or is entirely missing.

With the progression of the tumour the nystagmus-picture becomes more varied. We can see an irregular glance direction nystagmus: made up of a fine wave of second degree falling out nystagmus and one primary nystagmus directed at the side of the tumour, which originates from the brain stem. (Bruns-nystagmus)

Signs of the facial nerve being involved: less frequent blinks, smooth naso-labial folds, fatigability of the mimic muscles, primarily at the innervation of the labial commissure, Hitselberger symptom: tactile hypaesthesia when touching the back wall of the auditory meatus. Reduced tear and saliva secretion. The involvement of the nervus trigeminus can be seen in reduced cornea reflex tactile hypaesthesia on the face.

In the III.rd stage of the tumour the compression of the lower brain the nerves, the cerebellum, and the brain stem appear. This indicates a significant size of space occupation. The aim of the otorinolaryngological examination is early diagnosis and treatment as early as possible.

**Radiological signs of acoustic neuroma:**

Stenvers-radiography: it can show the dilation of the internal meatus only in half of the cases, very often bringing false-negative results. (This way these days the diagnostic value of this technique is questionable, if it is worth having it done at all.)

CT test: diagnoses the destruction of the internal meatus and gives an exact diagnosis of large tumours.

MR test: with gadolinium contrast material show small intrameatal tumours, it serves the mapping of the relationship between the brain stem and the tumour.

X-ray tests with positive contrast material have become superfluous since we have CT and MR.

Liquor tests are no longer part of the diagnostic tools, in spite of the fact that in the liquor in the case of larger tumours the total protein level is higher.

**Differential diagnosis:**

Non-tumour related pathologies. Menière-disease and syndrome, sclerosis multiplex, sudden hearing loss, clinical pictures involving tinnitus. Sometimes the neuroma, creating circulation disorder, misleadingly appears in the shape of acute hearing reduction with a spell of dizziness.

Other space occupying processes: arachnioideal cyst, congenital cholesteatoma, pontin glioma, meningeoma, distant metastases, recessus lateralis plexus papilloma, haemangioma.

Neurofibromatosis 1 (Recklinghausen-disease, NF1): it can be sporadic or familial with autosomal dominant line of heritage, the gene being on chromosome 17. Incidence of clinically multiplex hyperpigmented macula and neurofibromas, visual nerve glioma, but the occurrence of acustic tumours is not proven.
Neurofibromatosis 2 (NF 2): autosomal dominant heritage, on chromosome 22 the hereditary form of the mutation of the NF 2 (merlin) gene can be demonstrated. Its frequency is 1:50 000.

The appearance of bilateral acustic neuroma and other neurological manifestation, paravertebral neurofibroma. It may start at a young age, under 20, and also after 50. The prognosis for young patients is bad.

**Therapy:**

Operation, radical removal of the tumour, using microchirurgical methods, saving cranial nerve functions. The smaller the tumour the better functional results are expected after the operation. From a cosmetic point of view leaving the facial nerve intact is the most important but to preserve the level of hearing is significant from the patient’s quality of life, especially in the case of NF 2, where bilateral developments are expected. The facial nerve, strongly elongated, dislocated, broken into its fibres runs along the tumour’s sheath. Primarily, preceding direct compression, the circulatory disorder may damage the n. facial funcion. Further surgical complication is damage to a. cerebelli anterior inferior.

Stereotaxic radiation surgery: an aimed single large dosage radiation with linear accelerator or Co 60 gamma rays. (gamma knife) It can be used with tumours less than 2.5-3 cm, and in the case of old, high risk patients, if the patient’s other ear is deaf. Success rate: in 80 % of the cases the size of the tumour decreases or it stops growing. A further advantage is that it is an out- patients procedure, involves less stress and no surgical risk.

If due to the slow growth of the tumour it gives rise to very few complaints we can just ‘wait and scan’, provided we inform the patient about possible consequences. Very close follow-up, annual MR required.

**Follow up**

Whatever therapy has been done the yearly MR control is justified.

Acustic neuroma is a good example of the necessity of collaboration between related branches of knowledge. Because of the primary otological symptoms early diagnostics are in the hands of the ear specialist. He must think in time of having imaging tests made. The operation itself is often made by neurosurgeons, but hearing rehabilitation is again the task of the ear specialist. In the case of bilateral tumour NF 2 familial examination, and genetic advice is needed. Before deafness sets in the patient must be taught how to read lips. An instrument of hearing rehabilitation is grafting a brain stem hearing implant.