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Malignant melanoma of the ear - a literature review of a rare otological tumor

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Purpose

The aim of this study is to present the current state of knowledge on melanoma located in the ear.

Review methods

The PubMed and Google Scholar databases were used for the literature review. The following phrases were searched for in English: “ear melanoma”, “inner ear melanoma”, “external ear melanoma”, “middle ear melanoma”.

Abstract

The purpose of this article is to raise awareness of a malignant melanoma which is usually associated with a location on the exposed skin and occurs rarely in the area of the ear. Ear as an organ is divided into three parts - external, middle and inner ear. In each of these parts the process of carcinogenesis can occur. The tumor develops from mutated melanocytes. External ear melanoma contributes for only about 1% of all cutaneous melanoma and occurs mostly to people over the age of 60. Lesions in this area should raise concern and a detailed differential diagnosis should be implemented. The main treatment method is surgery, however due to rarity of the tumour no standard approach has been presented. A rarer location is malignant melanoma in the middle and inner ear, which can extend to the Eaustachian tube. There are barely a few cases described in the English literature so far. Symptoms include ear discharge, hearing impairment, and a sensation of fullness in the affected ear. As well as in the tumor of the external ear, also in this case, there are no standard procedures. However it is suggested to choose the surgical approach that can be combined with both radiotherapy and chemotherapy. The prognosis remains poor despite the significant development of medicine.

Key words: melanoma malignum, ear melanoma, inner ear melanoma, external ear melanoma, middle ear melanoma

Introduction

The ear is divided into three main parts - external, middle and inner ear. External ear, which consists of the auricle and external auditory canal, both covered with skin, is the only one exposed to the external environment. It is furtherly separated by the tympanic membrane from the middle ear. The middle ear is composed of tympanic cavity and mastoid air cells. Through the Eustachian tube it communicates with the nasopharynx. The tympanic cavity incorporates hearing ossicles (malleus, incus and stapes), chorda tympani and two muscles: tensor tympani muscle and stapedius muscle. The middle ear is covered by mucosa. The inner ear is located in the petrous part of the temporal bone. The bony labyrinth is filled with the perilymph, in which the membranous labyrinth is located. The labyrinth is divided into vestibular and cochlear parts, connected via a narrow duct. Those structures are responsible for hearing and balance.[1]

The World Health Organisation (WHO) in its 5th Edition of Classification of Head and Neck Tumours specifies several types of ear tumours, but from the anatomical point of view divides them only into two categories: the first one is external ear tumours and the second one is middle and internal ear tumours. Such a combination is due to anatomical closeness as well as radiological and clinical coincidence occurring among patients with tumours of this region. It mentions that neoplasms of this region are particularly rare and distinguishes 13 types of ear and temporal bone tumour types.[2] The classification features Squamous Cell Carcinoma (SCC), which is considered to be the most common tumour of the external and middle ear. It points out that in general the most common tumour of the temporal bone is Schwannoma.[2, 3, 4, 5]

Melanocytes are skin cells that contain melanosomes - melanin collecting granules. Melanin is one of the main contributors for pigmentation of both skin and hair. Melanocytes can absorb ultraviolet radiation (UVR), therefore protect against UVR-induced DNA damage.[6] However when exposed to certain factors, after gaining multiple somatic mutations i.e. in genes of key signaling pathways, melanocytes can uncontrollably lead to melanocytic neoplasm and give origin to both benign and malignant lesions, the latter called melanomas.[7] The most common type of malignant melanoma is cutaneous malignant melanoma.[8]

Mucosal melanoma covers less than 1% of all malignant melanomas and its rarity remains stable. It is most frequently situated in the head and neck region, followed by anus and rectum, female reproductive organs and urinary tract. Due to its rarity no proven risk factors

have been yet presented and it is suggested that surgery with adjuvant radiotherapy is the most frequently chosen option. The 5-year survival rate stays low at only 25%.[9] As mentioned before the ear structures can be covered with both skin and mucosa, which are two types of tissues from which melanoma can develop.

External ear

External ear melanoma (EEM) is a rather rare and infrequently encountered tumor, accounting for about 1% of primary cutaneous melanomas. This tumor is mostly diagnosed in patients over 60 years old. It is more often found in men (86.5%) than women and individuals with fair skin color.[10 - 18]

Studies conducted between 1973 and 2012 showed an increase in the incidence of EEM (111.9%) among adolescents and young adults. This aligns with the growing trend of total melanoma malignum incidence in individuals aged 15-39 years.[16]

EEM mainly occurs on the auricular helix and ear lobes. The most common subtype is superficial spreading melanoma (40.1%), which typically affects individuals between 30 and 40 years of age and presents as a slightly raised, multicolored lesion. Rarer forms of EEM are lentigo maligna (33.7%) and nodular melanoma (16.4%).[12, 14, 15]

The prognosis of external ear melanoma (EEM) is inconsistent as well as the treatment approach. Some researchers propose a more conservative choice of treatment, considering EEM to be a tumor with a good prognosis, while others advocate for more invasive treatments, viewing EEM as an aggressive cancer. Further research is needed to standardize management strategies for EEM.[13, 18]

For patients who undergo early surgical intervention, survival rates are very favorable.[16, 18] The 5-year survival rate for patients with stage I and II EEM is satisfactory, reaching 93%. However, for patients with stage III and IV EEM, the prognosis is poor, with 5-year survival rates of 56.6% and 20.5%, respectively.[18]

Prognostic factors that worsen survival rate include advanced age, tumor thickness, ulceration, lymph node involvement, and distant metastases.[12, 18]

In terms of symptoms, the lesion often does not raise any suspicion and may resemble a birthmark.[16] In superficial spreading melanoma, a pigmented, unevenly colored lesion with asymmetric features and mildly raised areas may be observed.[12] In lentigo maligna melanoma, a brownish, asymmetric spot with uneven color and irregular borders may be noticed.[12, 17] For nodular melanoma, a red-purple exophytic tumor with confluent satellite papules is

characteristic.[12, 14] If the tumor spreads from the external auditory canal to the middle ear, symptoms such as hearing loss, facial paralysis, otalgia, vertigo, and otorrhea may occur.[16]

Clinically and dermoscopically, no significant differences have been observed between melanoma of the external ear and melanoma of the face.[15, 19] Melanoma may not present with alarming symptoms, as it is painless and can easily be mistaken for a birthmark or other pigmented skin abnormality. As a result, some patients seek medical attention late when the tumor is already in an advanced stage, although most cases of EEM are diagnosed at an early stage.[12, 15, 16]

The diagnosis is confirmed through an excisional biopsy of the lesion.[12, 14, 15] Dermoscopy is helpful in diagnosing the condition, and in some cases, it can predict the type and stage of the tumor.[10, 12, 14, 19] Dermoscopy features that suggest superficial spreading melanoma include a blue-white veil, irregular dots and globules, rhomboidal structures, obliteration and asymmetric pigmentation of follicular openings, white depigmentation areas, and a circle within a circle pattern.[10, 12, 15] Dermoscopy features for lentigo maligna melanoma include a homogeneous, bluish area with rhomboidal structures, pigmentation of follicular openings, and the presence of a circle within a circle pattern.[10, 12, 15, 17] For nodular melanoma, dermoscopic features include a bluish veil, atypical vascular pattern, and irregular dots and globules.[12, 14]

To determine the precise location of EEM and the presence of distant metastases, which are rare, a CT scan is required.[12, 16]

There are no clear guidelines regarding the surgical treatment of external ear melanoma. As a result, the specific management approach for EEM is not standardized.[13, 18] The treatment of this tumor often differs from the standard treatment of melanomas located elsewhere due to the unique anatomy of the external ear. The cartilage of the auricle and the thin skin covering it are important factors that must be taken into account during surgical management. The unpredictable lymphatic drainage of the external ear, which may involve pre- or post- auricular sites, the parotid gland, and anterior or posterior cervical chains, must also be considered when choosing the form of treatment. Due to this peculiar anatomy, sentinel lymph node biopsy is performed to determine which nodes are at risk of involvement.[13, 16, 18]

Cartilage-preserving surgery is increasingly favored over composite excision, as it provides better conditions for later reconstruction. Currently, there is no evidence suggesting an increased risk of melanoma recurrence after cartilage-preserving surgery.[11]

No guidelines are presented specifying the margin width for excising melanoma of the external ear. It is important to remove enough tissue to ensure effective cancer management

while minimizing unnecessary disruption to the external ear's structure and function. Generally, a margin of at least 1 cm is recommended, but further research is needed in this area.[11, 14, 16, 18] For patients in whom surgery is not indicated or is not feasible, adjuvant radiotherapy can be considered.[12]

It is undeniable that early diagnosis and prompt treatment lead to longer survival for patients, so routine dermoscopic examinations of the ears by dermatologists are essential.[12]

Middle ear

Primary mucosal melanoma accounts for only 0.8%-3.7% of all melanomas.[20 - 22] It is 100 times rarer than cutaneous melanoma.[22] In most cases (over 50%), it is located in the head and neck region, mainly in the sinonasal region (66%) and the oral cavity (25%).[22]

Mucosal melanoma originating from the middle ear or Eustachian tube is particularly rare. By August 2018 there are merely 9 cases reported of this tumor with its increasing number - 21 reported up until February 2020.[20, 22, 23] Primary mucosal melanoma of the middle ear presents similarly to primary mucosal melanoma of the Eustachian tube, often involving as well the Eustachian tube.[22]

The 5-year survival rate is estimated between 20 and 35%.[23] Based on the cases described so far, middle ear melanoma has a poor prognosis—only half of the patients survived longer than 8 months.[20 - 22] Until 2021, one case of complete spontaneous regression of mucosal melanoma of the middle ear has been described.[24]

The main symptoms in patients with mucosal melanoma of the middle ear include ear discharge, hearing impairment, and a sensation of fullness in the affected ear.[20 - 23, 25] Other symptoms include constant ear pain, tinnitus, and facial nerve weakness.[20, 22, 24] In patients with mucosal melanoma of the Eustachian tube, the most common symptoms observed were hearing impairment and aural fullness. Less common symptoms included epistaxis and otorrhea.[22]

Otoscopy may reveal redness, swelling, a dark-appearing tympanic membrane, and discharge from the middle ear.[20] Nasopharyngoscopy can show a dark polyp-like lesion at the entrance of the Eustachian tube.[23, 25] Some patients may present with serous otitis media.[21, 22] The tumor may extend to the posterior end of the nasal septum or the external auditory canal.[23, 25] The diagnosis is confirmed through biopsy.[20, 23, 25, 26] In cases where the melanoma invades the bone, CT and MRI are the primary imaging modalities.[20] Positron emission tomography/computed tomography with 2-[fluorine-18]-fluoro-2-deoxy-D-glucose (FDG-PET-CT) will show increased FDG uptake value.[23, 24]

Middle ear melanoma should be differentiated from ceruminous carcinoma, squamous cell carcinoma, glomus tumor, otitis media, and mastoiditis.[20, 23]

This tumor can cause distant metastases to the lungs, liver, abdomen, intraparotid region, as well as cutaneous metastases.[20, 21]

Similar to mucosal melanoma in locations other than the middle ear and Eustachian tube, the primary treatment is surgical resection with clear margins. Radiotherapy may be used as an adjuvant treatment for advanced lesions or in cases with positive margins. It is also utilized as a palliative method for unresectable lesions or in patients not qualifying for surgery.[22, 26] Chemotherapy may be the sole treatment or used as an adjuvant therapy.[22] Molecular targeted immunotherapy appears promising.[23] Mutations in the BRAF proto-oncogene, common in cutaneous melanomas, are less frequent in mucosal melanomas. Therefore, further research is needed on immune response modulators, such as anti-PD-1 and PDL-1. [22, 23, 26]

Inner ear

As mentioned before the WHO classifies tumours of middle and inner ear as one, emphasizing the fact that tumours in this area are very rare.[2] Cerebellopontine angle (CPA) is triangular space in the posterior cranial fossa in close connection to the middle ear, containing structures like cranial nerve V, VI, VII, and VIII along with the anterior inferior cerebellar artery.[27] WHO points out that the most common neoplasm occurring in the temporal bone, as well as in the CPA region, is vestibular schwannoma, whereas CPA melanoma is sporadic.[2, 27]

Only one case of primary melanoma with origin in the cochlea has been found in the English literature.[28] Several cases of melanoma metastases to the inner ear have been described. Most of them showed that the tumour has reached the CPA.[29 - 32]

Symptoms can include sudden hearing loss, dizziness, nausea, vomiting and tinnitus as well as the headache, facial weakness and nystagmus. Some of the manifestations can occur unilateral or bilateral. To confirm the hearing impairment most of the patients underwent tone audiometry, but also the use of Auditory Evoked Potential Testing was described. All of the authors performed Magnetic Resonance Imaging (MRI).[28 - 32]

The chosen treatment method is surgery accompanied with adjuvant radiotherapy, less frequently chemotherapy or steroid administration.[28 - 30, 32] Due to the possibility of misdiagnosing inner ear melanoma as schwannoma, both of the tumours should be included in differential diagnosis in patients who present previously mentioned symptoms.[28, 29] None of the authors give any detail about long term outcomes and life expectancy.

Conclusion

Melanoma is a malignant tumor originating from melanocytes, primarily found in the skin. It accounts for about 65% of skin cancer-related deaths.[20] A concerning trend is the increasing incidence of melanoma in adolescents and young adults (ages 15-39), making it the third most common cancer in this age group.[16]

Melanoma of the external ear is a relatively rare tumor in a difficult-to-examine location. It can easily be mistaken for a birthmark or other benign skin lesion, which may lead to patients not seeking medical help in the early stages of melanoma development. This contributes to delayed diagnosis and treatment, resulting in poorer prognosis.[14, 16] Therefore, routine dermatological check-ups to exclude EEM, especially in young individuals, are encouraged, as the incidence of melanoma of the external ear and other body parts is increasing in this age group.[12, 15, 16]

Rarely (about 1.3%), melanoma can also affect mucous membranes, mainly in the nose and paranasal sinuses. Primary mucosal melanoma in the middle and inner ear is extremely rare, with only a small group of cases reported so far.[2, 20] Sensorineural hearing loss and vestibular dysfunction are a frequent reason why patients search for urgent otolaryngological help. Even though primal or metastatic malignant melanoma located in the inner ear or CPA is a rare cause of those symptoms, in case of lack of other diagnosis or further exacerbation of ailment it should be included in differential diagnosis.[28 - 32]

The prognosis for melanoma in uncommon sites, such as the inner or middle ear, is unfortunately poor or unknown due to diagnostic difficulties, late recognition and the lack of characteristic symptoms.[20, 21]

In order to improve the detection of ear melanoma, we would like to increase the awareness of this ever-growing problem. Due to the non-specificity of the symptoms reported by the patient, they can be easily misdiagnosed. A thorough interview and both basic and extended tests are necessary. The described 5-year survival time is the highest in patients who underwent surgery, but it also varies depending on the initial stage of the disease but it is considered as relatively poor.[33, 34] For this reason, rapid diagnosis and treatment are essential.

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