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CASE REPORT

Primary malignant melanoma of the middle ear mucosa: a case report

Orta kulak mukozasının primer malign melanomu: Olgu sunumu

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Primary involvement of the middle ear and mastoid mucosa by malignant melanoma (MM) has been rarely reported. We report an 81-year-old Caucasian woman presenting with otalgia, left-sided aural fullness, and hearing loss. Otoscopic examination revealed a purple coloured polyp that appeared to be vascular and occupy left external auditory canal (EAC). The computed tomography scanning showed a mass in soft tissue density, filling the tympanic and the mastoid cavities, eroding the mastoid cortex, and invading medial and central parts of EAC. A biopsy was performed with the diagnosis of MM. The patient underwent subtotal temporal bone resection with radical neck dissection. Dacarbazine was given postoperatively. The case presented herein emphasizes the need to be aware that MMs can arise in the middle ear and mastoid cavity. An index of suspicion in the differential diagnosis of temporal bone tumours is necessary for early recognition.

Key Words: Neoplasm; malignant melanoma; mucosa; temporal bone; dacarbazine.

Orta kulak ve mastoid kavite mukozasının malign melanoma bağlı primer tutulumu nadirdir. Sol kulakta ağrı, dolgunluk ve işitme kaybı yakınmalarıyla başvuran 81 yaşındaki kadın hastanın otoskopik muayenesinde mor renkli, vasküler görünümde kitlenin sol dış kulak yolunu tıkadığı görüldü. Bilgisayarlı tomografide yumuşak doku dansitesindeki kitlenin orta kulak ve mastoid kaviteyi doldurduğu, mastoid korteksi erode ederek dış kulak yolunun medial ve orta kısımlarını invaze ettiği görüldü. Kitleden alınan biyopsi sonucu malign melanom olarak bildirildi. Hastaya subtotal temporal kemik rezeksiyonuna ek olarak radikal boyun diseksiyonu uygulandı ve ameliyat sonrası dönemde dakarbazin verildi. Temporal kemikten kaynaklanan tümörlerin ayırıcı tanısında malign melanomdan şüphelenmek erken tanıya olanak sağlamaktadır.

Anahtar Sözcükler: Neoplazi; malign melanom; mukoza; temporal kemik; dakarbazin.

Malignant melanoma of the temporal bone may emanate from the squamous epithelium of the auricle and the external auditory canal (EAC), or rarely from the mucosa of the middle ear and the mastoid cavity.[1-4] The tumor can arise as a primary lesion or as a result of metastasis. Primary involvement of the middle ear and mastoid mucosa by malignant melanoma has been rarely reported.[2-4] The formulation of the adequate management options for mucosal melanoma is controversial due to its rarity in this location. The surgical therapy has been combined with radiation therapy and chemotherapy.[5] We report the description of a rare case of a malignant melanoma of the middle ear mucosa with no other primary site.

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CASE REPORT

An 81-year-old Caucasian woman had one month history of a bloody discharge from her left ear. The patient described otalgia, left-sided aural fullness, progressive hearing loss, and tinnitus. She denied any vertigo and had no evidence of facial palsy. Her past medical history was significant for type-2 diabetes mellitus, hypertension, and cardiac arrhythmias. Her family history was unremarkable.

On physical examination, left sided otoscopy revealed a dark purple colored polyp that apperead to be vascular (Fig. 1). The tympanic membrane could not be examined, because the polyp occluded the medial two thirds of the EAC. Right sided otoscopy was normal. Tuning fork tests were consistent with a left sided conductive type hearing loss. There was no palpable cervical lymphadenopathy, and the remainder of the head and neck examination was normal. Pigmented skin lesions or suspicious nevi were not detected by whole body dermatological examination.

The results of audiological evaluation revealed a mixed type hearing loss with an air-bone gap of 47dB on the left ear. Word discrimination score was 72% on the left ear. The preoperative high-resolution computed tomography (CT) scanning of the temporal bone showed an irregular mass in soft tissue density, filling the tympanic and the mastoid cavities, eroding the mastoid cortex along the posterior wall of the EAC, and invading medial and central parts of the EAC (Fig. 2a-c). The mass appeared confluent with

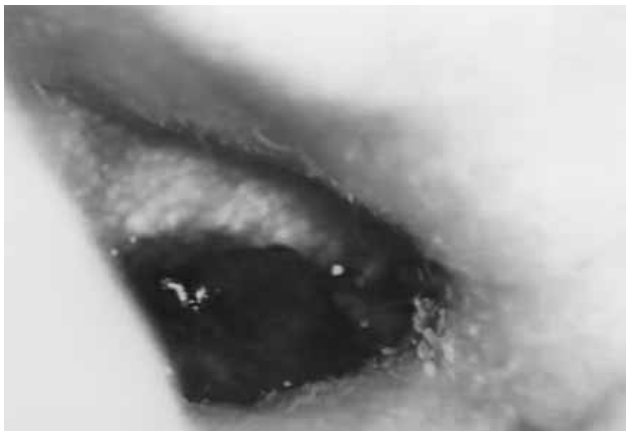

Fig. 1 - *Left otoscopy revealing a dark purple colored polyp occluding the medial two thirds of the EAC.*

the tympanic membrane. The ossicles were detected to be intact. Digital substraction angiography showed the mass to be a relatively avascular lesion.

A biopsy of the hemorrhagic mass in the left EAC was performed with the pathologic diagnosis of malignant melanoma. Complete body survey to consider metastasis and to confirm the diagnosis of primary malignant melanoma of the middle ear was performed with bone scintigraphy and thoraco-abdominal CT studies, which were completely negative. Routine laboratory tests and liver function tests were all within normal limits.

Preoperative examination with the patient under anesthesia revealed a mildly bleeding mass in the deep EAC after violation. The mass was attached to

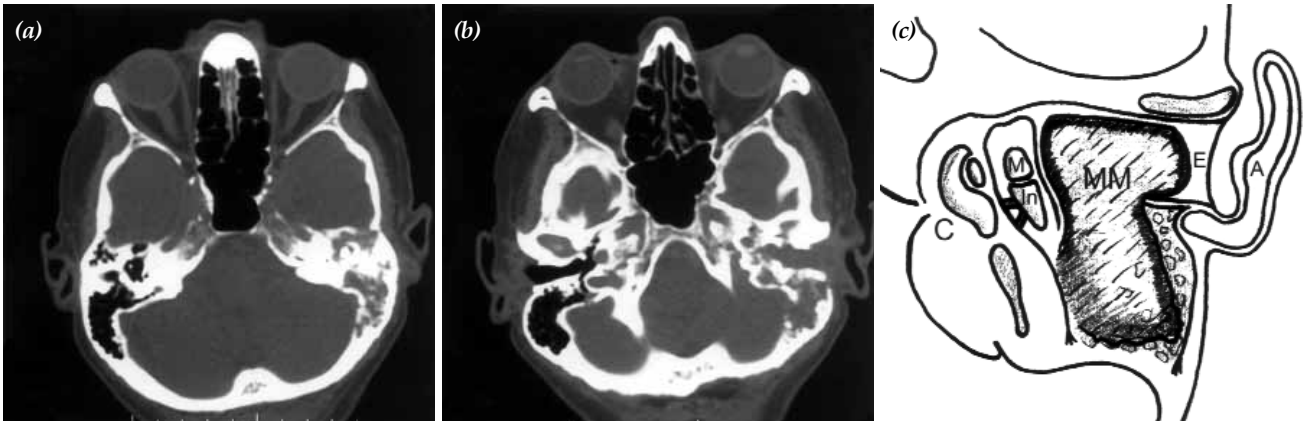

Fig. 2 - *Preoperative axial computed tomographic scan at the level of cochlea* ***(a)****, and external auditory canal (EAC)* ***(b)****, showing an irregular mass in soft tissue density, filling the medial and central parts of the EAC, destructing the mastoid cortex along the posterior wall of the EAC and invading the mastoid cavity. The mass appears confluent with the tympanic membrane and the ossicles are detected to be intact. The invasion pattern of the malignant melanoma (axial section) has been illustrated* ***(c)****: A, auricle; E, external auditory meatus; MM, malignant melanoma mass; M, malleus; In, incus; C, cochlea.*

the postero-inferior wall of the medial two thirds of the EAC without revealing a cleft to examine the tympanic membrane. The patient underwent left subtotal temporal bone resection with radical neck dissection in combination with periparotid lymph node dissection. Facial nerve function was intact postoperatively.

The pathologic examination of the specimen was positive for malignant melanoma and immunoreactive for the melanin specific antigen, HMB45 (Fig. 3, 4). The surgical specimen showed laterally compressed tympanic membrane with a marginal perforation of the posteror and inferior quadrants. The hypotympanum, tympanic sinus, facial recess, posterior epitympanum, mastoid aditus and mastoid cavity were involved with intact middle ear ossicles. The tumor infiltration to the posterior belly of digastric muscle and the sternocleidomastoid muscle at the mastoid apex region was detected. Microscopic metastasis of melanoma was present in the highest 3 lymph nodes adjacent to the parotid gland and EAC, out of 37 isolated lymph nodes.

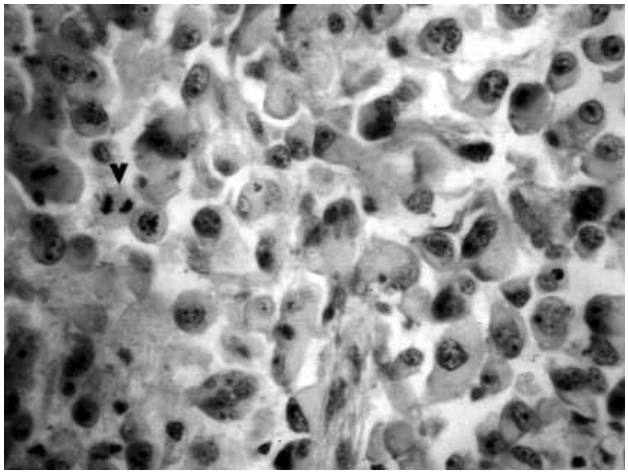

Fig. 3 - *Representative histologic section of malignant melanoma. Original magnification (H-E x 400).*

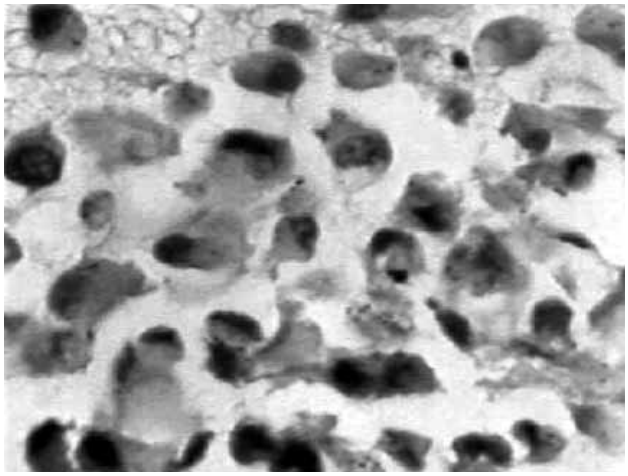

Fig. 4 - *Photomicrograph showing the tumor immunoreactive for the melanin specific antigen, HMB45 stain. Original magnification (H-E x 400).*

After discussing the options with the patient, medical oncology consultation was obtained postoperatively. Dacarbazine chemotherapy regimen (250 mg/m^2) was planned and continued for 5 days. No complications occurred during chemotherapy.

Although the therapy was considered to be effective, our patient died three months after the operation due to a possible cardiovascular accident. The patient's family refused autopsy, so the cause of death could not be investigated and the possibility of metastases could not be confirmed.

DISCUSSION

In terms of proportion of all melanomas, primary mucosal melanomas of the head and neck represent 0.4% to 1.8% of the total.[6] The malignant melanoma of the middle ear mucosa is rarely reported (Table I).[2-4,7-11]

Cutaneous malignant melanoma of the auricle is thought to arise as a result of intense sun exposure, but the specific pathophysiological factors underlying the development of the primary malignant melanoma of the middle ear mucosa and the deep EAC are not well known.[1,5] Genetic predisposition and the patient's phenotype are parts of the etiology.[1,5]

Patients often present with long-standing otalgia,

TABLE I

THE REPORTED CASES OF PRIMARY MALIGNANT MELANOMA OF THE MIDDLE EAR

Author	Year	Number of patients
Oueslati Z, et al.[7]	2001	1
Uchida M and Matsunami T.[3]	2001	1
Urpegui Garcia A, et al.[8]	1999	1
Banerjee A, Meikle D.[9]	1992	1
Sherman IW, et al.[4]	1991	1
McKenna EL Jr, et al.[2]	1984	1
Kagan AR, et al.[10]	1982	1
Muir CS, Nectoux J.[11]	1980	1

otorrhea, bloody discharge and loss of hearing.[12] These presenting signs and symptoms are very nonspecific, and are also associated with chronic infections of the EAC and middle ear. Because of the location and the frequent lack of pathognomonic symptoms, there is often a delay in diagnosis.[12] Diagnosis of malignant melanoma is established by biopsy of a suspicious lesion.[3] Pathologic diagnosis of melanoma hinges on the identification of intracellular melanin.[3]

Successful management of a melanoma depends on the early recognition. Complete radiological evaluation requires CT of the temporal bone and the neck for the evaluation of the extent of invasion of neighbouring structures. Routine metastatic workup should include a thoraco-abdominal radiograph, radionuclide bone scan and liver function tests.[5]

Surgery currently offers the best probability for cure of malignant melanoma of the head and neck.[5] The surgical management of the head and neck melanomas is often limited by the surrounding anatomical structures. Extension of the tumor into the mastoid cavity and EAC may necessitate concomitant subtotal temporal bone resection, mastoidectomy, and a parotidectomy.[12] Careful surgical planning minimizes the cosmetic and functional consequences of extirpation and decrease the tumor burden.

The nodal status is one of the most important predictors of survival. The removal of occult metastatic disease by various dissections of the neck will reduce the incidence of recurrence.[13] The extension of neck dissection to include periparotid lymph nodes is essential. The technique of lymphoscintigraphy and sentinel lymph node biopsy to identify patients with nonpalpable lymph node metastases could greatly increase the yield of elective lymph node dissection.[13]

Radiation therapy may play an increasing role in the treatment of patients with unresectable local disease or those who refuse surgery. Radiation therapy is not effective in completely eradicating the tumor and improving the overall survival, but improves regional disease control.[5] Chemotherapy has generally be reserved for patients with systemic disease, and has not enhanced survival or regional control rates. Dacarbazine (DTIC) and nitrosureas have been the mainstays of chemotherapy.[5] Immunotherapeutic agents (interferon and interleukin-2), and vaccines against melanoma associated antigens are promising.

Malignant melanoma of the middle ear is unusual and difficult to diagnose clinically, so a high index of suspicion is necessary for early recognition. Before deciding on a treatment plan, it is important to discuss the poor patient survival outcome and high morbidity that exists with extensive resections and adjunctive therapy with radiotherapy or chemotherapy. The case presented herein emphasizes the need to be aware that melanomas can arise in the middle ear and mastoid cavity.

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