



Case Report

Malignant Melanoma of the External Auditory Canal

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Abstract

Primary malignant melanoma rarely occurs in the external auditory canal. In the few reported cases, treatment with lateral temporal bone resection, superficial parotidectomy, and neck dissection were suggested. We describe a 52-year-old man with this disease, managed by sentinel node biopsy with lymphoscintigraphy guidance, lateral temporal bone resection, and postoperative radiotherapy. The patient refused superficial parotidectomy and neck dissection. Although there was no locoregional recurrence on follow-up, pulmonary metastasis was diagnosed in the third year after treatment. The incidence, symptoms, treatment, and prognosis of external auditory canal malignant melanoma are also discussed in this article. (*Tzu Chi Med J* 2009;21(3):244–247)

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1. Introduction

The cause of malignant melanoma of the skin is mainly attributed to overexposure to the ultraviolet component of sunlight. Genetic factors, skin freckles, fair skin, light hair, age, and immunosuppression are other predisposing factors (1,2), with Caucasians being more susceptible. Malignant melanoma arising from the external auditory canal (EAC) is a rare disease, with few reports in the medical literature (1–4). In this article, we report a case in an Asian patient.

2. Case report

A previously healthy 52-year-old man presented with a 4-month history of a blockage sensation in his right ear. Inspection showed a protruding dark fleshy mass filling the entire right EAC, arising from the posterior aspect (Fig. 1). Neurologic and head and neck examination showed no other specific findings. Axial and coronal views of a preoperative computed tomography scan of the temporal bone showed a soft-tissue mass (about 1.3×0.8×0.6 cm) filling the right EAC (Fig. 2). Biopsy specimens obtained from this mass

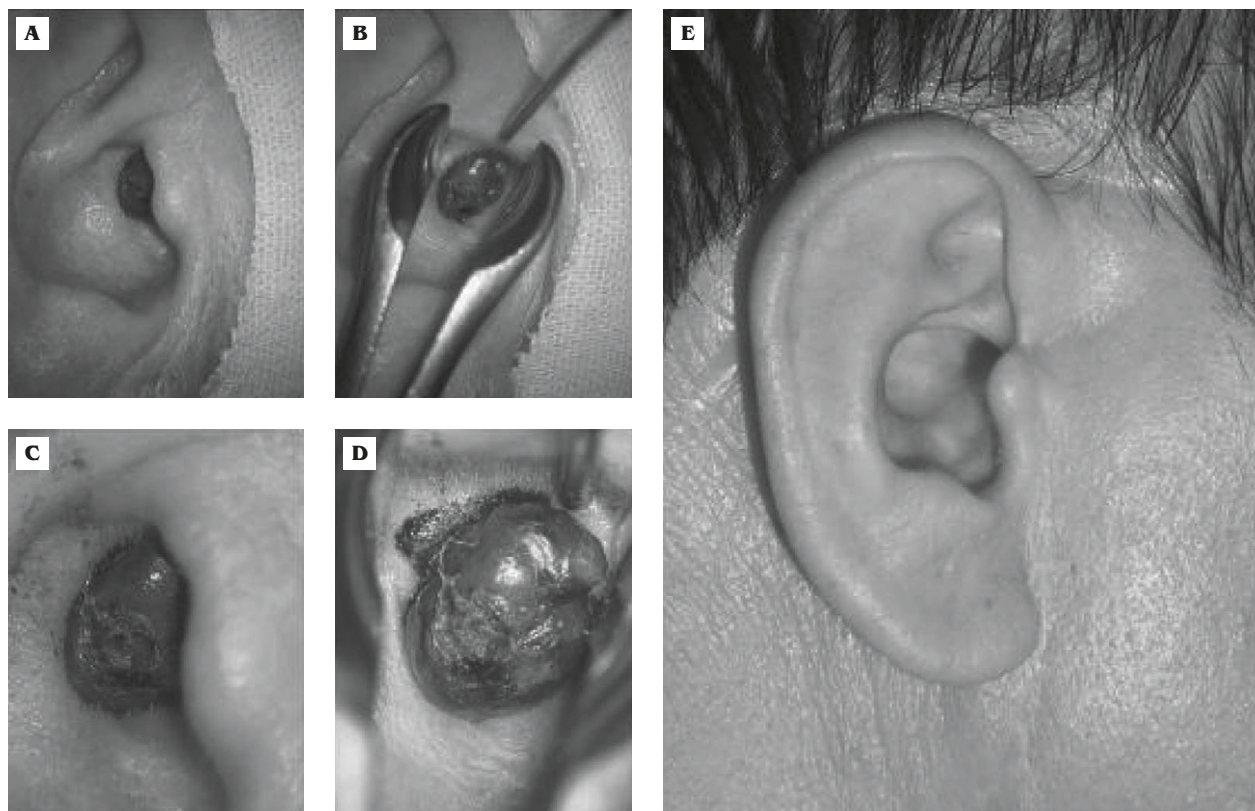


Fig. 1 — (A–D) Macroscopic views of a protruding dark fleshy mass filling the entire right external auditory canal arising from dark skin at the posterior aspect of the external auditory canal wall. (E) Macroscopic view of the ear 3 years post-operatively. Physical examination showed a dry cavity without local recurrence.

revealed sheets of oval- to spindle-shaped cells with melanin pigments of great variation in size and shape (Fig. 3). These cells stained positively with the HMB-45 and S-100 (Fig. 3) stains, but negatively with the cytokeratin AE1/AE3 stain. Melanin pigments—as confirmed by a Fontana-Masson stain—were present in both the oval- to spindle-shaped cells and the macrophages in the stroma. Malignant melanoma of the EAC was diagnosed.

Whole body positron emission tomography showed no evidence of distant metastasis. The patient was informed of the necessity of a lateral temporal bone resection, superficial parotidectomy, and ipsilateral neck dissection. However, the patient refused the extended superficial parotidectomy and neck dissection. Lymphoscintigraphy (Fig. 3) with sentinel node biopsy showed no nodal metastasis. The patient subsequently underwent a lateral temporal bone resection and postoperative radiotherapy. The tumor was resected with a 2 cm safety margin and the pathology report revealed that the margins were all disease-free. The patient was regularly followed in our outpatient department for 3 years. Unfortunately, metastasis developed in the lungs in the third year, although there was no local recurrence (Fig. 1).

3. Discussion

Malignant melanoma of the ear is estimated to occur in 1–4% of all skin melanomas and about 7–20% of melanomas of the head and neck region (1,3,4). The mean age at diagnosis is reported to be around 50 years; all age groups are affected except for younger populations. The most frequently involved areas of auricular melanoma are the helix and ear lobule. Malignant melanoma confined to the EAC is rarely reported in the literature (1,2,5,6).

The presenting symptoms of malignant melanoma of the EAC are hearing loss, sensation of a blocked ear or foreign body, and discharge from the ears (1,2,5,6). These nonspecific symptoms are often ignored by patients until near total blockage of the EAC occurs. There should be a high index of suspicion of a possible malignant melanoma when a pigmented lesion changes color, enlarges rapidly, or ulcerates with poor healing. Excisional biopsy of these lesions is advised.

The optimal management of malignant melanomas of the ear is total surgical excision with at least a 1–2 cm safety margin in consideration of possible satellite lesions (3). Sometimes, it is more difficult to excise malignant melanomas inside the EAC because

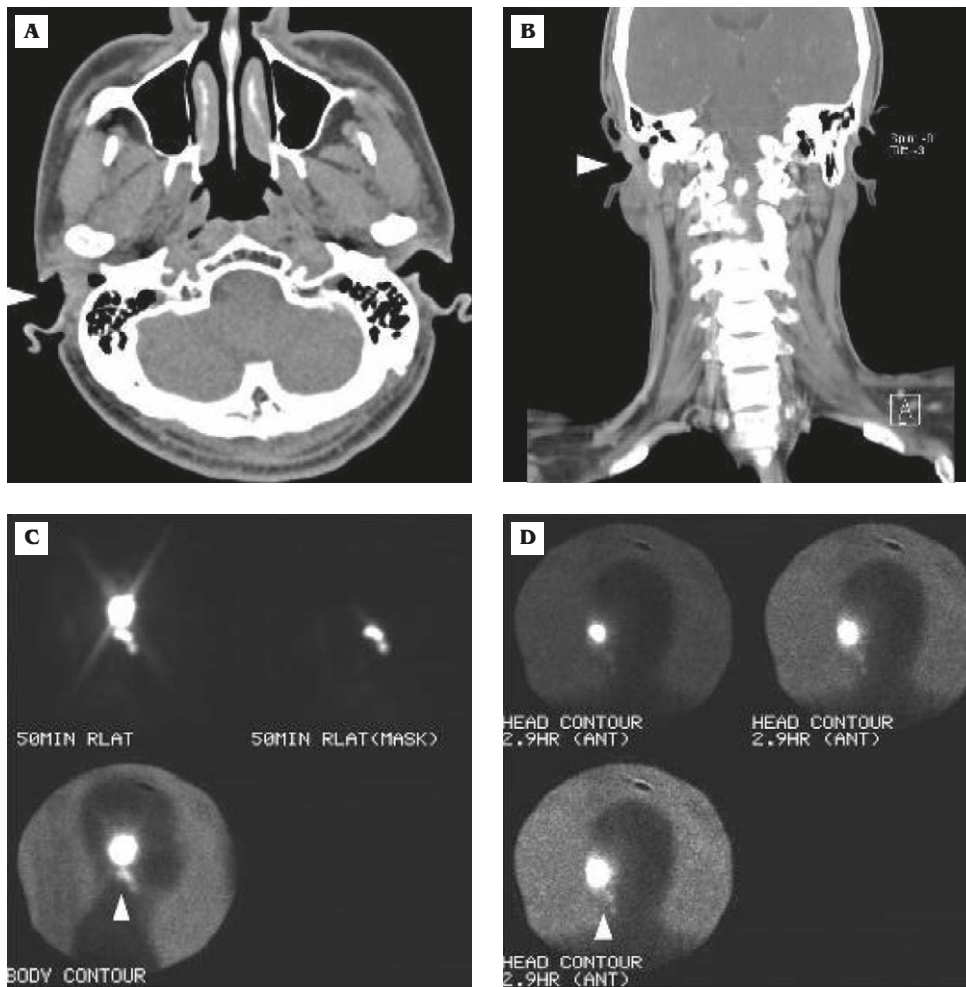


Fig. 2 — (A, B) Axial and coronal views of preoperative computed tomography. White arrowheads show a soft-tissue mass measuring about 1.3×0.8×0.6 cm filling the right external auditory canal. (C, D) Right lateral view and anterior view on lymphoscintigraphy (white arrowheads) show the sentinel lymph node below the main tumor. Sentinel lymph node mapping and excisional biopsy were completed before the operation on the main tumor.

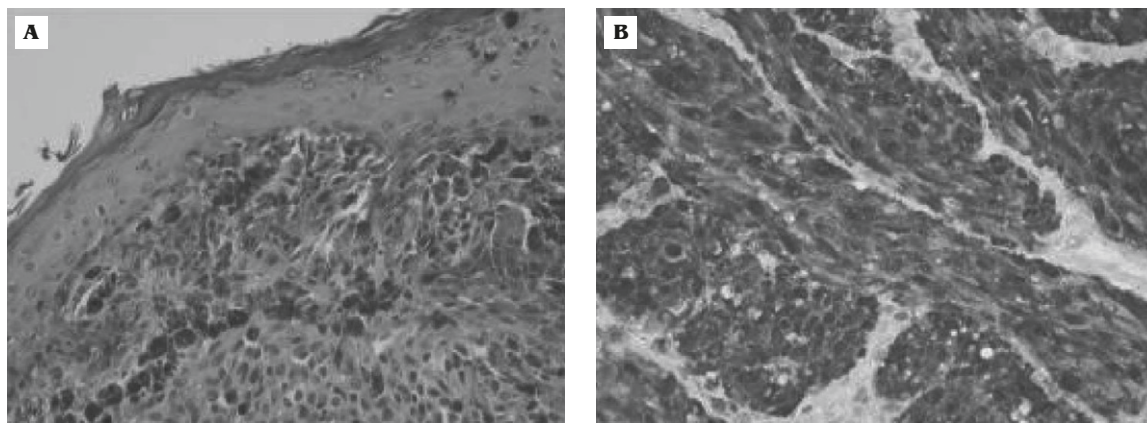


Fig. 3 — (A) Nests of polyhedral and spindle-shaped tumor cells with melanin pigments in the cytoplasm and polymorphism of the nuclei (hematoxylin & eosin, 100×). (B) The tumor cells stained positively with the S-100 protein immunohistochemical stain (200×).

of the limited space between the lesion, and the neighboring delicate and vital structures of the middle/inner ears. Wide local excision of a malignant melanoma in the EAC often leads to recurrence of disease and mortality within 2 years. Removal of the tumor with a margin of surrounding bone is the minimal requirement in treating the primary focus (1).

For the clinically negative neck, the treatment is rather controversial. Routinely, a superficial parotidectomy with a neck dissection has been suggested (1–3,5,6). However, several prospective trials of patients with cutaneous malignant melanomas could not find a definitive benefit of elective lymph node dissection on patient survival (7,8). Therefore, the value of elective neck dissection for the patient with melanoma of the ear is still undetermined.

Morton et al first reported a sentinel lymph node biopsy to drain a lymph node in a malignant melanoma of the head and neck (9). Later studies showed that lymphoscintigraphy with sentinel node biopsy helped in decision making regarding neck dissection and yielded a high success rate and low morbidity in patients with a clinically negative neck (7). However, many large retrospective studies failed to show a positive relationship between positive results from a sentinel node biopsy and survival. Nevertheless, it remains the most useful staging and prognostic indicator at the present time and the results of sentinel node biopsy do help high risk patients in the determination of possible adjuvant therapies (7,8).

When there is neck metastasis, functional neck dissection and high doses of interferon alpha 2b are suggested. Other possible adjuvant therapies such as immunomodulation therapy, vaccination, radiation therapy and chemotherapy have been discussed, although no definitive survival benefits have been reported (10).

Tumor thickness is the most important prognostic factor of malignant melanoma of the skin, but a nodular pattern, ulceration, and regional or distant metastases are associated with a poor prognosis. Other poor prognostic factors for malignant melanoma in the EAC include delayed diagnosis because of its deep location, a greater tendency for metastasis due to the thin epithelium, a short distance between the tumor and the dermal lymphatics, and a previously failed surgical excision (1).

In summary, malignant melanomas of the EAC are extremely rare, especially in Asians, and the utility of sentinel node biopsy has not been mentioned in previous case reports (1,2,5,6). We reported a case that was staged and treated using lymphoscintigraphy with sentinel node biopsy, lateral temporal bone resection, and postoperative radiotherapy. Both whole body positron emission tomography scanning and sentinel node biopsy before treatment showed no regional or distant metastasis, and the patient was under complete locoregional control after treatment. However, pulmonary metastasis eventually developed after 3 years. This highlights EAC melanoma as a grave disease with a systemic predisposition and poor prognosis; long-term follow-up is necessary despite good locoregional control.

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