

CASE REPORT

Malignant mucosal melanoma of the eustachian tube

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Abstract Malignant mucosal melanoma is a rare condition. Although the head and neck region is the most common site for mucosal melanoma, a melanoma arising in the eustachian tube is rare. Here we present a case of mucosal melanoma arising in the right eustachian tube.

Key words Malignant melanoma · Eustachian tube · Magnetic resonance imaging

Introduction

Malignant melanoma is a neoplasm formed by melanocytes or melanocyte precursors. Although melanoma usually occurs on the skin, this tumor may also arise from the mucosa. Mucosal melanomas of the head and neck account for more than 1% of all melanomas, and about 50% of these tumors arise in the oral cavity; melanoma of the eustachian tube is a rare condition. Here we present a case of malignant melanoma arising in the eustachian tube.

Case report

In 2006, a 41-year-old man presented to a local otolaryngology clinic with progressive fullness of the right ear and hearing impairment for more than 3 months. On examination, there was a middle ear effusion on the right side. Exudative otitis media was diagnosed, and antibiotic therapy was given. Due to lack of improvement, he was referred to our hospital.

The initial endoscopic examination revealed a dark mass in the cartilaginous portion of the right eustachian tube (Fig. 1), and no changes in the nasopharyngeal mucosa. Routine laboratory tests were normal. He had no past history of cutaneous or mucosal melanoma.

Contrast-enhanced computed tomography (CT) of the nasopharynx showed thickening of the soft tissues with enhancement at the cartilaginous portion of the eustachian tube (Fig. 2a). The bony segment of the tube was intact on CT with a bony window setting.

Magnetic resonance imaging (MRI) was performed with a 1.5 tesla system with 3 mm slice thickness and 0.6 mm interslice gap. The MRI showed fluid in the right mastoid antrum and a mass (28 × 35 mm in diameter) arising from the cartilaginous portion of the eustachian tube (Fig. 2b,c). On contrast-enhanced T1-weighted images, the tumor showed moderate, homogeneous enhancement (Fig. 2d). Chest CT, abdominal ultrasonography, and fluorodeoxyglucose positron emission tomography (FDG-PET) detected no metastases.

A biopsy was performed for suspected malignant melanoma. Examination of hematoxylin-eosin (H&E)-stained sections showed that the tumor cells varied in shape and size (Fig. 3a). On immunohistochemical examination, the tumor cells were positive for S-100

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protein, melanoma antibody HMB45 (Fig. 3b), and Melan-A.

The patient received carbon ion radiation therapy (total dose 57.8 Gy) without adjuvant chemotherapy and is currently well without metastasis 8 months after treatment.

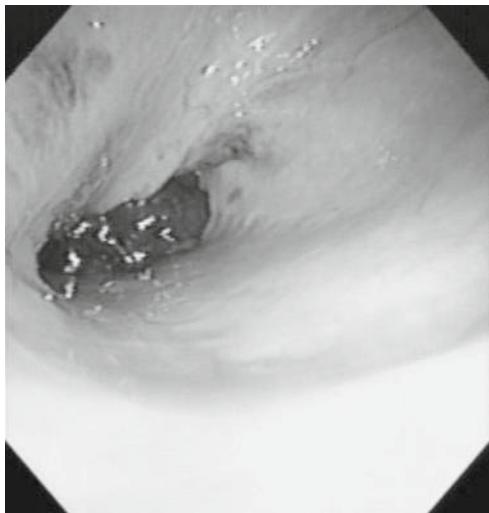


Fig. 1. Endoscopy shows a black tumor in the deep pharyngeal part of the right eustachian tube

Discussion

Malignant melanoma of the skin is a neoplasm of dermal melanocytes. It is strongly related to exposure to the sun, but the etiology of mucosal melanoma is unknown.¹ Melanoma primarily affects the skin and much less frequently arises from the mucosa, with mucosal melanomas of the head and neck accounting for about 1% of all melanomas or 6.3% of all head and neck melanomas.^{2,3}

The nasal cavity, paranasal sinuses, and oral cavity are the major sites for mucosal melanoma of the head and neck.^{1,4–7} In the nasal cavity, the sites most frequently involved are the anterior part of the nasal septum and the inferior and middle turbinates. In the oral cavity, common sites include the palate and gingivae.⁸ The eustachian tube is rarely affected. To our knowledge, only four cases have been reported in the English-language literature,^{9–11} and there was no discussion of CT or MR findings.

MR imaging is superior to CT for detecting melanoma and defining its extent. Malignant melanoma shows high attenuation on unenhanced CT because of its melanin content and hemorrhage.⁵ The MR appearance of melanotic melanoma is characteristic because of paramagnetic material in the tumor. The tumor shows high signal intensity on T1-weighted imaging (T1WI)

Fig. 2. **a** Contrast-enhanced computed tomography (CT) of the nasopharynx demonstrates a homogeneously enhanced mass in the deep part of the right eustachian tube. The mass shows high intensity on T1-weighted imaging (**b**), low intensity on T2-weighted imaging (**c**), and diffuse homogeneous enhancement on contrast-enhanced T1-weighted imaging (**d**)

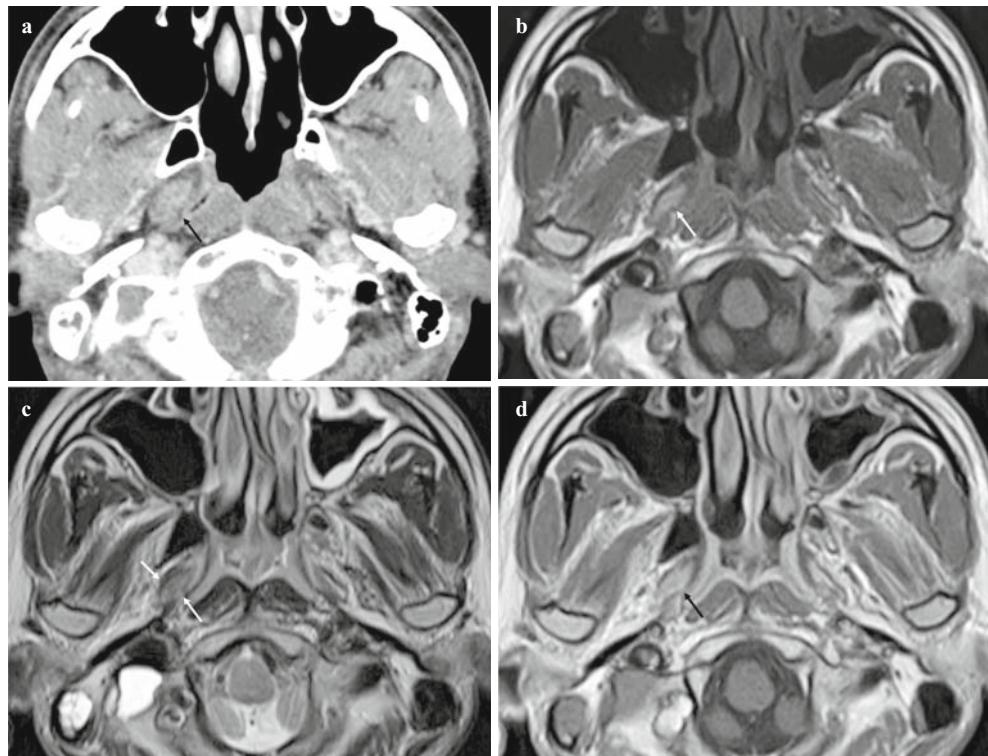
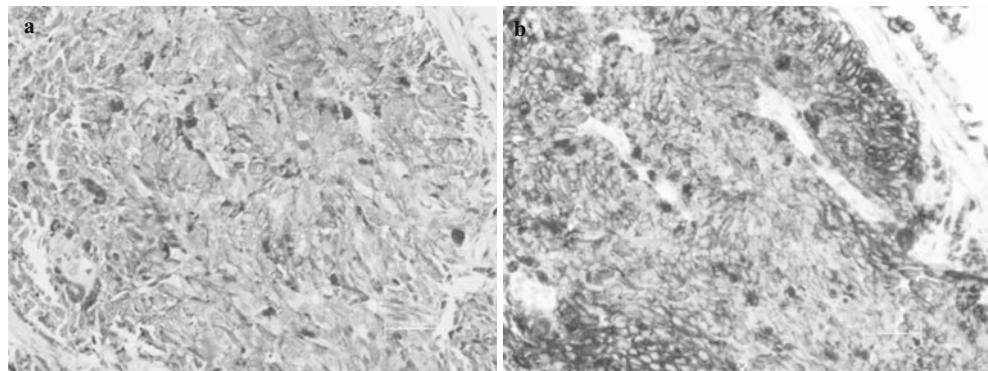


Fig. 3. Tumor cells vary in shape and size on H&E-stained sections (**a**) and are positive for HMB45 (**b**). ($\times 40$)



and low signal intensity on T2WI.^{3,5,12–14} Melanin is paramagnetic and decreases the T1 and T2 relaxation times of the surrounding protons, so the melanin content of a melanoma determines its intensity.^{13,14} Because of their rich vascularity, melanomas show strong enhancement on contrast-enhanced CT and MRI. In our case, the tumor had characteristic MR findings, but CT was less helpful. This was probably because the tumor arose in the cartilaginous portion, and there was no difference in attenuation from the surrounding normal structures.

The differential diagnosis includes lesions containing other paramagnetic agents such as manganese or iron, fungal infection, and hematoma. Fungal infection occurs in patients with underlying immunosuppression or diabetes mellitus, and the lesion is usually infiltrative. Metastasis from hemorrhagic tumors and mucinous adenocarcinoma can also show low intensity on T2WI and high intensity on T1WI.

More than 95% of melanomas are S-100 protein-positive and are negative for cytokeratins. However, S-100 protein is not specific for melanoma. More specific markers include HMB45, Melan-A, and anti-tyrosinase, but they stain in only about 75% of lesions.¹⁵

The prognosis for patients with mucosal melanoma is worse than for those with cutaneous melanoma because mucosal melanoma tends to be more advanced at the time of diagnosis. Cutaneous melanoma has a 5-year survival rate of 80.2%, whereas head and neck mucosal melanoma has a rate of only 31.7%.² However, this survival rate is still higher than that for mucosal melanoma at other sites, such as the female genital tract or the anorectum. The prognosis associated with mucosal melanoma is not influenced by the size of the primary lesion. Rich vascular and lymphatic perfusion of the mucosa promotes tumor spread, and positive lymph nodes have a significant adverse effect on survival. Chang et al. reported that lymph nodes are positive in 26.0% of head and neck melanomas.² Approximately 19% of patients present with lymph node metastasis, another 16% develop nodal metastasis after treatment, and 10%

present with distant metastasis.⁸ Lee reported that patients with local control had significantly longer survival than those with persistent local disease,¹⁶ so aggressive local treatment seems to be beneficial.

In general, surgery is the treatment of choice for malignant melanoma because of its radioresistance. In our case, wide excision would have been necessary for complete resection, so radiotherapy was selected instead. Radiotherapy has recently become more effective owing to better understanding of melanoma radiobiology, and surgery combined with radiation therapy has been increasingly employed for mucosal melanoma. In addition, carbon-ion therapy has recently been performed for malignant melanoma. There is no effective systemic chemotherapy for mucosal melanoma, so chemotherapy is used only for palliation.

Conclusion

We report a rare case of mucosal melanoma arising from the eustachian tube. Although primary eustachian tube melanoma is rare, it should be kept in mind when the tumor shows characteristic MR findings.

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