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Mucosal melanoma arising in the eustachian tube

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Abstract
Mucosal melanoma is a very rare disease. Most cases have their origins in the head and neck region. To date, only three cases of melanoma originating from the eustachian tube have been reported. We present a case of mucosal melanoma of eustachian tube origin in which a complete excision was performed. In this case, the patient underwent adjuvant radiotherapy and the mass size greatly decreased, which aided subsequent surgical excision. Systemic chemotherapy was not utilized in this case; however, it is often used for palliative purposes. A literature review is also presented.

Key words: Melanoma; Eustachian Tube; Radiotherapy

Introduction

In the head and neck region, cutaneous melanoma occurs more frequently than mucosal melanoma. In the United States, only 1.3 per cent of 84 836 melanoma patients presenting during the period 1985–1994 were diagnosed with the mucosal type.[1] The head and neck area was the primary site in 55 per cent of these cases. Only three cases of melanoma originating from the eustachian tube have been reported.[2] We present a case of mucosal melanoma arising from the eustachian tube, which was diagnosed at our institution in 2002. The patient was treated with neoadjuvant radiotherapy and complete surgical excision was performed.

Case presentation

A 62-year-old woman presented to the otolaryngology outpatient clinic with a chief complaint of 18 months of hearing impairment and nine months of bloody discharge from the right ear. She underwent tympanostomy tube insertions in 1998 and 1999.

On 9 July 2001, exploratory tympanoplasty was performed on the right ear, which revealed a granuloma and polyp obstructing the eustachian tube. The initial physical examination revealed a mass lesion protruding from a defective portion of the right tympanic membrane, and a dark coloured mass was noted in the right Rosenmuller fossa of the nasopharynx. A biopsy of the external auditory canal mass revealed melanoma (Figure 1), and magnetic resonance imaging (MRI) showed a submucosal mass of the right nasopharynx invading through the eustachian tube into the inner ear (Figure 2a and 2b). No metastatic lesion or other primary site was found on physical examination, whole body bone scan and abdominal ultrasonography.

On MRI, the right eustachian tube was thought to be the primary site, and radiotherapy was employed because the patient refused operation. The patient received a total of 6500cGy (500cGy twice a week for six and a half weeks). No acute complications of radiotherapy were evident, although the patient suffered from mild xerostomia. A follow-up MRI taken two months after the onset of radiotherapy revealed that the size of the mass had decreased. After the completion of radiotherapy, surgical excision via a type C infratemporal fossa approach was performed (Figures 3 and 4). At the time of writing, one year following surgery, the patient was being followed with regular outpatient visits and there was no sign of recurrence (Figure 2c and 2d).

Pathology

On light microscopic examination, the tumour was composed of sheets of ovoid-to-spindle shaped cells, some containing coarse dark brown pigments in the cytoplasm. The nuclei were moderately pleomorphic, with frequent nucleoli and mitotic figures. Immunohistochemical studies revealed diffuse, strong cytoplasmic staining for S-100 protein and HMB-45 antigen in the tumour cells. A diagnosis of malignant melanoma was subsequently made on the basis of these histological and immunohistochemical features (Figure 1).

Discussion

Mucosal melanoma was first reported by Weber in 1859, and since then approximately 1000 cases have been reported.[3] Mucosal melanoma originating in the head and neck area occurs more commonly in the nasal cavity than in the oral cavity. The commonest sites of nasal cavity mucosal melanoma are the anterior septum (33 per cent) and the lateral wall of the nasal cavity (28 per cent).[3] In cases of melanoma found in the middle-ear space, the disease is usually diagnosed in its advanced stages. Therefore, the primary site cannot easily be distinguished from the middle-ear mucosa, the eustachian tube or the nasopharyngeal mucosa. To date, a total of six cases have been reported in the literature. In two of these cases the primary site could not be determined, in another two cases the disease was limited to the middle-ear region, and in the remaining two cases the disease involved the eustachian tube and the nasopharynx.[2,4–6]

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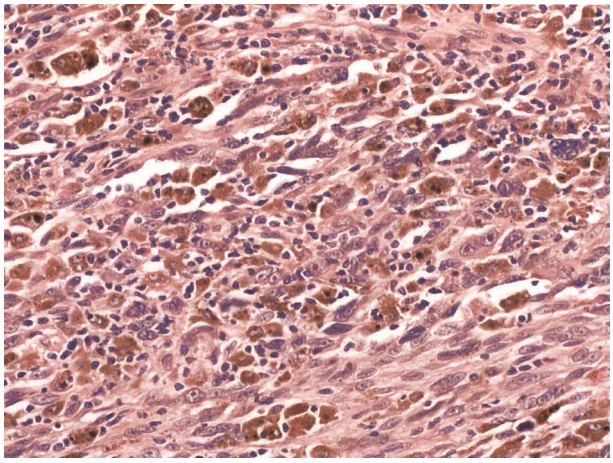

FIG. 1

The tumour is composed of ovoid-to-spindle shaped cells showing prominent nucleoli and vesicular nuclei. Many tumour cells contain dark brown melanin pigments in their cytoplasm (H & E; ×200).

In the present case of mucosal melanoma, the tumour extended from the submucosa of the nasopharynx to the middle ear and, based on radiographic imaging, we determined that the eustachian tube was the origin of the tumour.

When the individual tumour cells are found to be melanin-rich, the diagnosis is usually not difficult. However, amelanotic lesions may resemble other benign and malignant tumour types, so stains that have been employed most commonly for the diagnosis of melanoma include S-100 protein and HMB-45. S-100 is a sensitive yet non-specific marker for melanoma, while HMB-45, a monoclonal antibody derived from a melanoma extract, is more specific. Nevertheless, HMB-45 may be detected occasionally in carcinoma cells. More recently, melan-A, a melanoma-specific marker, has been added to the diagnostic armamentarium in order to more effectively distinguish between melanomas and other tumours when equivocal results have been obtained with the more traditional markers described above. Melan-A, developed from a human melanoma cell line, has been shown to be highly specific in its ability to distinguish metastatic melanoma from other lesions such as undifferentiated carcinoma, various sarcomas, high grade lymphomas and plasmacytomas.[3]

There is no established staging system for mucosal melanoma originating in the head and neck region (apart from that arising in conjunctivae). However, clinically, the disease has been categorized into three stages: stage I for localized disease, stage II in cases of lymph node metastasis, and stage III when distant metastasis is present.[3] Considering that mucosal melanoma has a low rate of lymph node metastasis and that nodal metastasis has no determined effect on the five-year survival rate, it is inappropriate to stage mucosal melanoma according to the classification guidelines of cutaneous melanoma.[7,8] Recently, mucosal melanoma has been classified according to the site of origin, into melanoma that arises from the oral cavity mucosa, and melanoma that arises from the sinonasal and nasopharyngeal mucosa. Between these two groups, there is a statistically significant difference in the rate of nodal metastasis, however there is no difference in five-year survival rates or regional and distant recurrence rates.[8,9] In the present case, elective neck dissection was not performed since the tumour was a mucosal melanoma originating from the eustachian tube.

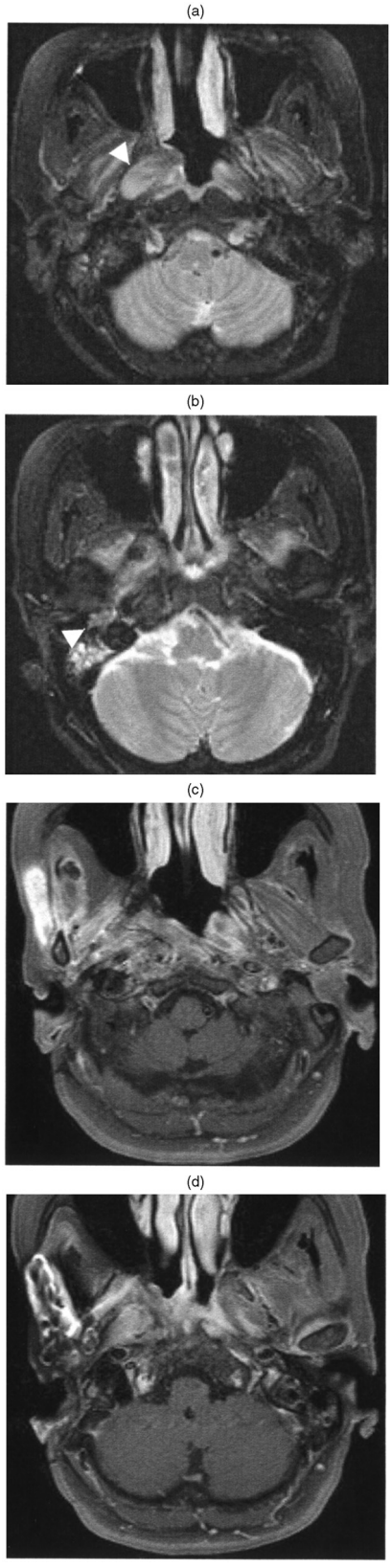

FIG. 2

(a) and (b), initial temporal magnetic resonance imaging (MRI). The mass lesion is seen in the right-sided nasopharynx submucosa (2a arrowhead) and extends into the middle-ear cavity via the eustachian tube (2b arrowhead). The eustachian tube is considered to be the origin of the mass. (c) and (d), temporal MRI taken one year post-operation. There is no evidence of tumour recurrence.

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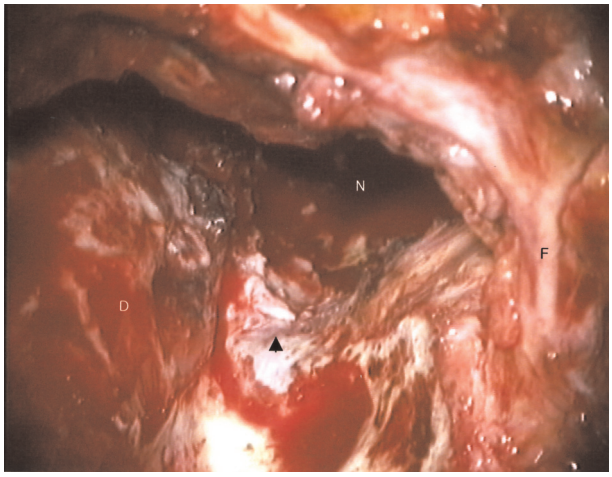

FIG. 3

Intra-operative findings following tumour removal (via type C infratemporal fossa approach).
D = middle cranial fossa dural plate; F = main trunk of facial nerve; N = nasopharynx; arrowhead = carotid artery.

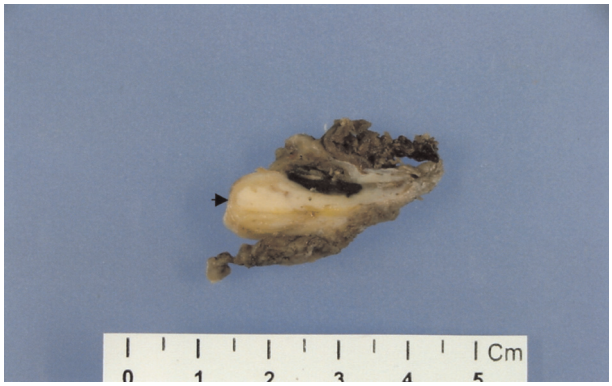

FIG. 4

The surgical specimen, covered by fibrous tissue. The surrounding tissue contains some muscular tissue. Hemisection along the canal shows a black-brown pigmentation lining the mucosal surface of the canal. Arrowhead = eustachian tube cartilage.

Surgical excision is believed to be the best treatment modality for melanoma.[8] Radiotherapy is known to be ineffective in most cases; however, adjuvant radiotherapy can be considered if surgical excision with adequate margins is not achieved due to anatomic complexity or local recurrence.[8] Currently, high-dose, hypo-fractionated radiotherapy is utilized, and it is increasingly preferred over the standard radiotherapy methods due to its low morbidity rate, despite the fact that there is no evidence of improvement in the five-year survival rate.[3,10] In this case, surgical excision was performed after the mass size was reduced by neoadjuvant high-dose, hypo-fractionated radiotherapy, which was given to a total of 6500 cGy, using 6-MV energy, three-dimensional, conformal radiotherapy technique, over six and a half weeks. Currently, there is no established regimen for chemotherapy in cases of mucosal melanoma, and it is usually used for palliative purposes only.[8]

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