The Journal of Laryngology and Otology
February 1996, Vol. **110**, pp. 163–166

Primary malignant melanoma of the palatine tonsil: a case report

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Abstract
The authors report a clinical case of a primary malignant melanoma of the right palatine tonsil in a 75-year-old woman.

Key words: Melanoma; Tonsillar neoplasms

Introduction

Malignant melanoma is a relatively common neoplasm of the skin; although less common, melanomas also arise in the eye, in the meninges and in the mucosal membranes of digestive and upper respiratory tracts (Svane-Knudsen, 1957; Conley and Pack, 1974; Shah *et al.*, 1977; Lund, 1982; Berthelsen *et al.*, 1984; Rapini *et al.*, 1985). Melanocytes, from which melanomas may potentially arise, have been demonstrated in the larynx (Goldman *et al.*, 1972), nasal cavity (Zak and Lawson, 1974) and mouth (Becker, 1927).

Mucosal melanomas are usually of the acral lentiginous type (Rubin and Farber, 1988) and represent 0.3–10 per cent of all head and neck melanomas (Guzzo *et al.*, 1993). They most frequently occur in the nasal cavity, nasal sinuses, hard palate and lips (Berthelsen *et al.*, 1984; Guzzo *et al.*, 1993). The oropharynx is one of the less common

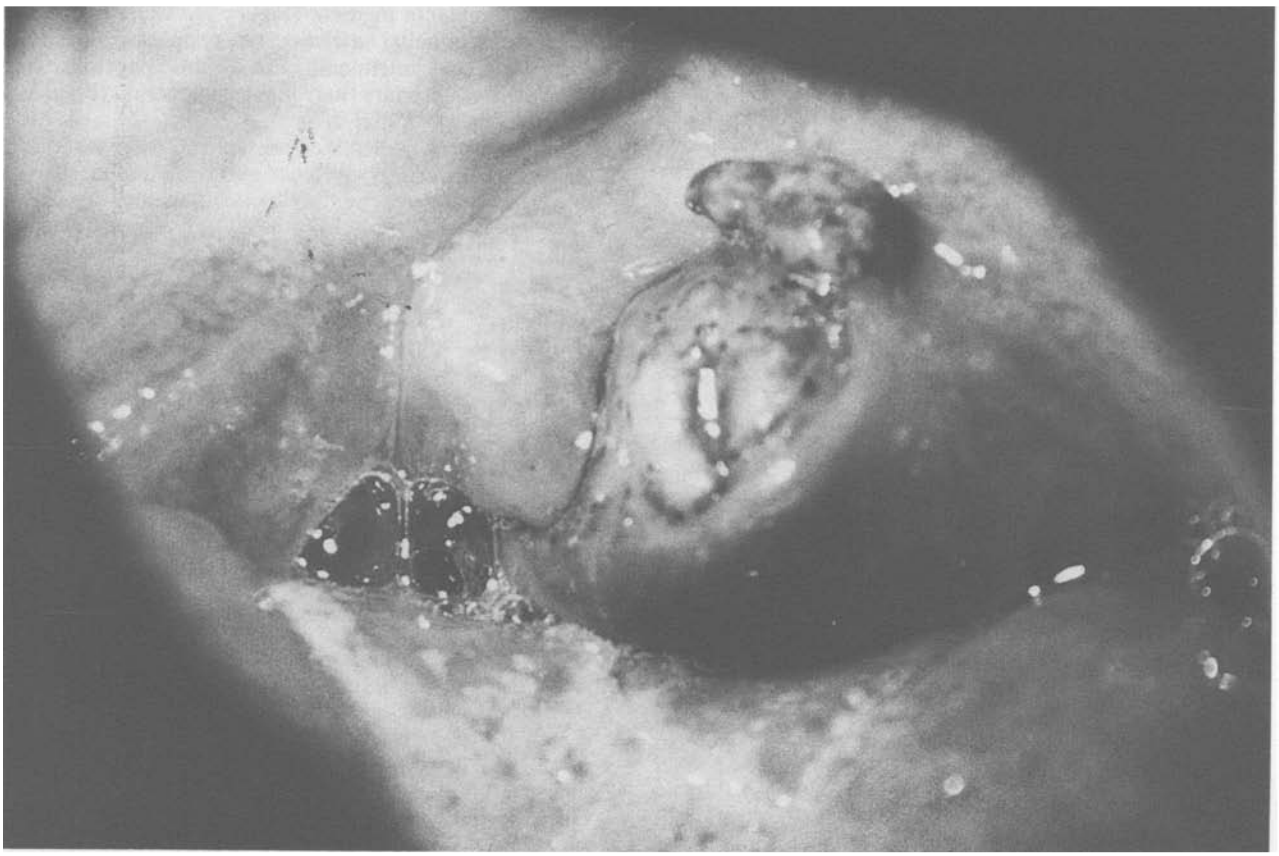

Fig. 1
Malignant melanoma of the left tonsil.

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Accepted for publication: 24 November 1995.

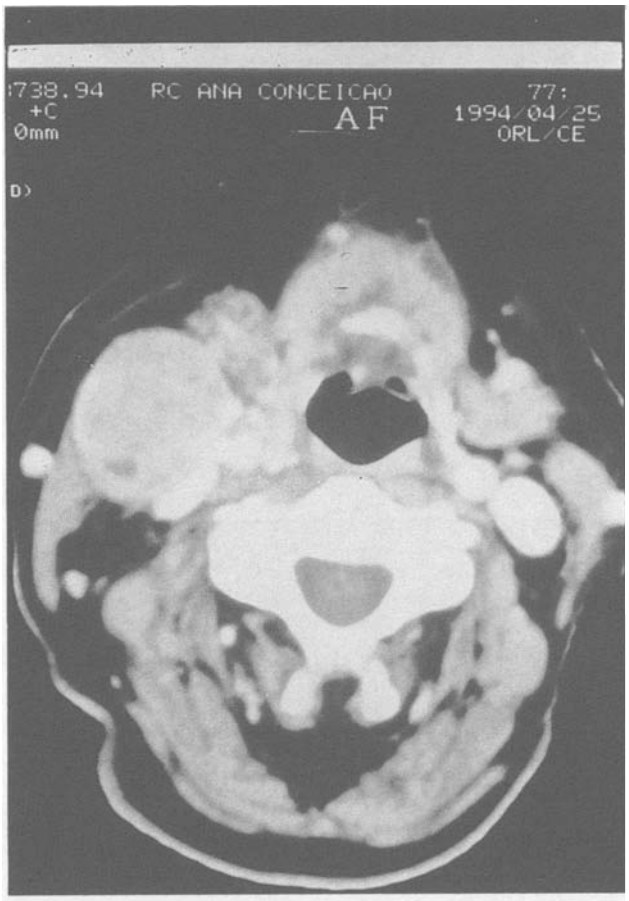

FIG. 2
CT scan of the pharynx and neck showing the jugular lymph node.

localizations and, as far as we know, there have been only three cases reported in the literature of primary malignant melanoma of the palatine tonsil (Howarth, 1943; Svane-Knudsen, 1957; Michaels, 1987).

Case report

A 75-year-old woman complaining of odynophagia and progressive high dysphagia, presented herself to our Department of Otolaryngology. Two months earlier, she had noticed a painless right cervical mass. About one month afterwards, she started to complain of odynophagia, progressive dysphagia and sialorrhoea, though denying asthenia, anorexia and weight loss. Up until then, she had always been a healthy person.

On examination, a pigmented lesion was seen on the medial surface of her right palatine tonsil, with necrosis and ulceration (Figure 1); both the anterior and posterior pillars, the lateral half of the tonsil and the soft palate were apparently free of disease. On the right side of the neck, there was a jugular lymph node, 3 × 3 cm, hard, painless to palpation and not adherent to either superficial or deep layers. The rest of the examination was unremarkable.

The CT scan of the pharynx and neck (Figure 2) showed the jugular lymph node to be partially compressing the internal jugular vein and pushing anteriorly the submandibular gland. The para-pharyngeal fat tissue was not invaded. It was decided to perform a tonsillectomy.

The previous evaluation included normal blood analysis, and a normal X-ray. The pulmonary function tests revealed a moderate mixed obstructive-restrictive component and the ECG showed a bi-fascicular heart block.

Tonsillectomy was performed and was uneventful; it was easy to separate the tonsils from their beds, which were macroscopically normal. The capsules seemed to be preserved, as histological report later confirmed. The pathological diagnosis was malignant melanoma.

An abdominal ultrasound (US) was then performed which was normal, a bone scan, and a cerebral CT, with no metastasis. The patient was observed by a dermatologist but no cutaneous melanoma was found.

After discussion with our oncological group, it was decided to perform adenectomy and to precede the treatment with local radiation and tamoxifen (20 mg/day). The patient agreed to radiation and drug therapy but refused any surgical treatment.

At the present time (six months of follow-up) the patient is doing well and with no signs of local recurrence. The jugular lymph node is about the same size as before.

Pathological findings

After surgery, the specimens received at the Department of Pathology were sampled, embedded in paraffin and stained with haematoxylin-and-eosin and Masson-Fontana's method for histological examination. Immunocytochemical study with immunoperoxidase techniques were performed using antibodies for demonstration of S100 protein, HMB 45, Cytokeratins (CAM 5.2, Becton-Dickinson) and leukocyte common antigen. Ultrastructural study was not performed.

The right tonsil contained a solid malignant neoplasm of mixed epithelioid and spindle cell pattern, containing abundant melanin pigment (Figure 3). There was prominent intraepithelial melanocytic neoplasia of surface epithelium of junctional nested and pagetoid types indicative of primary malignant melanoma (Figure 4). The tumour was 8 mm thick.

Immunocytochemical study revealed CAM 5.2 and LCA markers (respectively epithelial and lymphoid markers) to be both negative. It was not possible to identify definite positivity for S100 protein (present in melanocytes as well as other cells (Filipe and Lake, 1990), but there was a clear positivity for HMB 45, a monoclonal antibody with putative specificity for melanoma cells (Wick *et al.*, 1988).

Discussion

Carcinomas and lymphomas are the commonest malignant neoplasms of the tonsils (Friedmann, 1986). Primary malignant melanoma is extremely rare (Svane-Knudsen, 1957; Friedmann, 1986; Michaels, 1987), making it important to rule out metastatic melanoma; this may affect one or both tonsils in cases of disseminated melanoma (Filipe and Lake, 1990). The presence of an intraepithelial, neoplastic component ('junctional activity') is the most important histological evidence supporting the clinical suspicion of a primary tumour. However, metastatic malignant melanoma with epithelial tropism has been described (Kornberg *et al.*, 1978), making a careful clinical search for a potential primary site essential. Patients with metastatic melanoma in the absence of a recognized primary tumour have been reported (Weedon, 1992), either because the primary lesion underwent spontaneous regression (Chang and Knapper, 1982; Giuliano *et al.*, 1982), localized in lymph nodes or in visceral organs (Shenoy *et al.*, 1987), or was undetectable in the skin (Giuliano *et al.*, 1982; Shenoy *et al.*, 1987).

In the present case, there was no evidence of a present or past primary lesion after a careful examination, and

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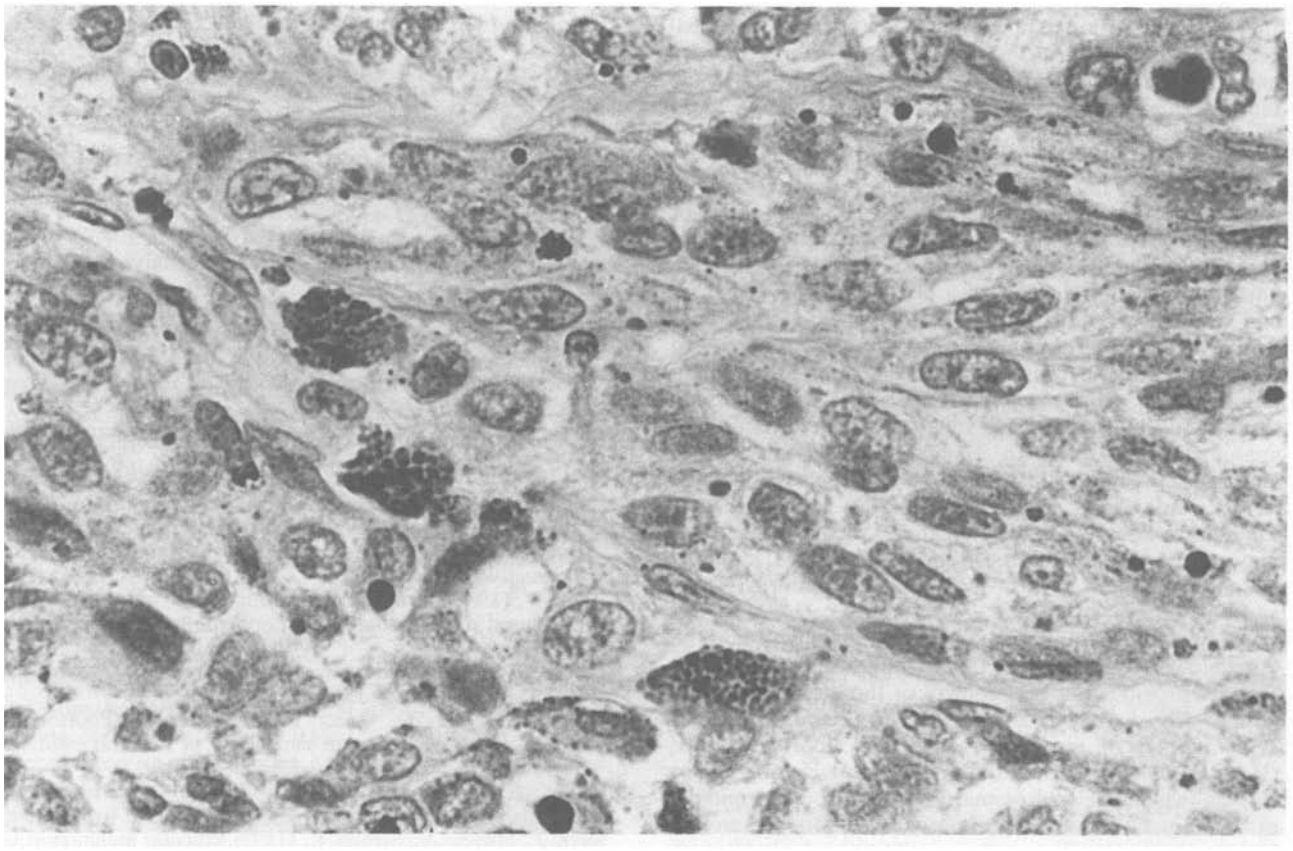

FIG. 3
Atypical pleomorphic melanoma cells containing melanin pigment, together with melanophages (H & E; × 400).

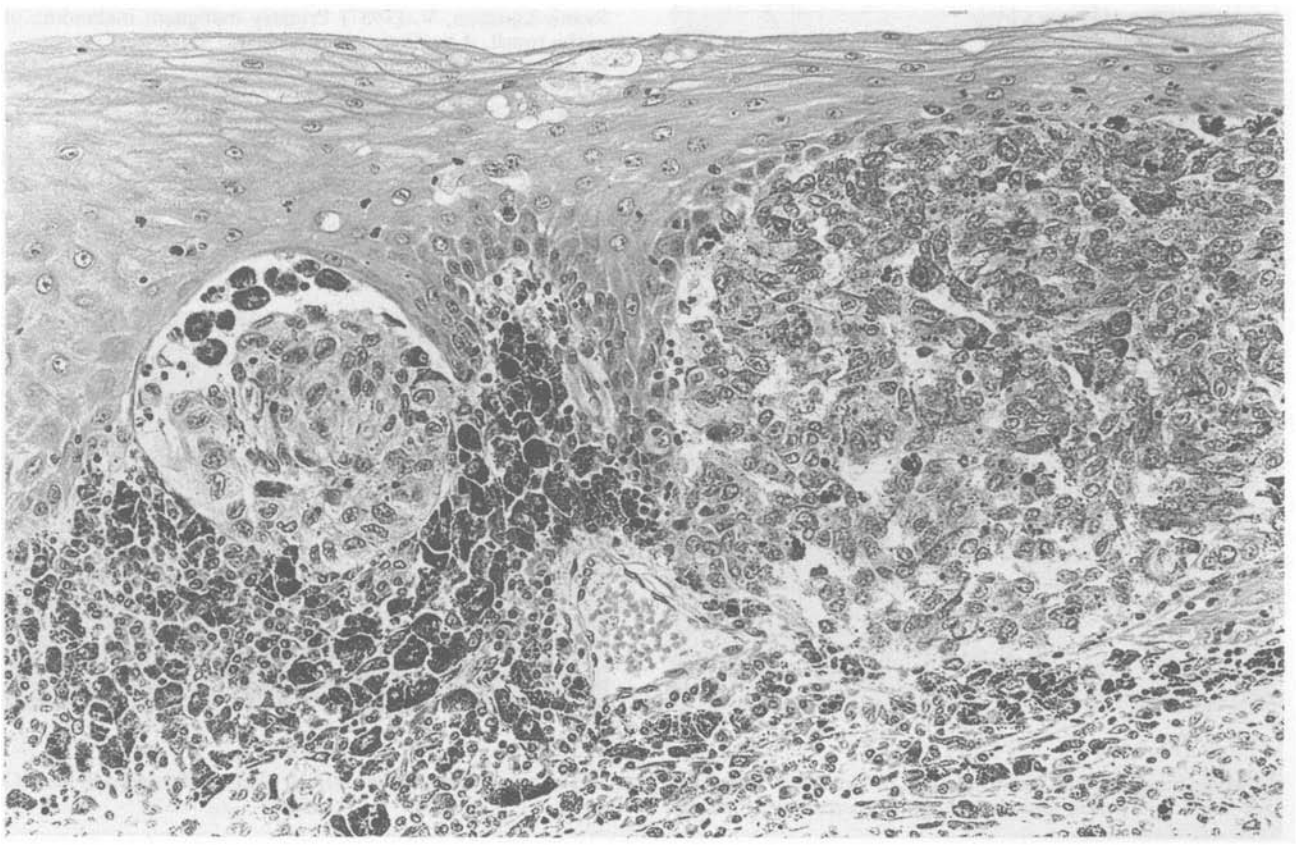

FIG. 4
Nested atypical melanocytic proliferation at the mucosal-submucosal junction. (H & E; × 200).

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there was a striking 'junctional activity' at histological preparations (Figure 4). So, the diagnosis of primary malignant melanoma seems reasonable. Although most mucosal melanomas are of the acral lentiginous type (Rubin and Farber, 1988), the present case demonstrates a vertical growth phase pattern.

In spite of the improvement in prognosis of the cutaneous melanomas in recent years, prognosis of mucosal melanomas has remained poor and worse than for cutaneous forms (Rapini *et al.*, 1985). Many reasons have been proposed for such a poor prognosis (Eneroth and Lundberg, 1975; Eisen and Voorhees, 1991): these neoplasms produce nonspecific symptoms, causing delay in the diagnosis; they affect old people, with inefficient immune systems; mucosae have rich vascular and lymphatic efferents, promoting early metastasis; there are unusual anatomical constraints to the extent of surgery; these neoplasms seem to be histologically aggressive, with a high mitotic index and marked anaplasia. The maximum tumour thickness (Breslow thickness) is the single most important factor of prognosis in clinical stage I and II melanomas of the vertical growth phase pattern (Rubin and Farber, 1988).

All forms of therapy seem to have little influence on the outcome of the disease (Eisen and Voorhees, 1991). Surgery provides the best control of the disease but only temporarily. Local recurrences are frequent even with wide margins of resection (Eisen and Voorhees, 1991; Guzzo *et al.*, 1993). Radiotherapy has a place either after surgery, when disease free margins cannot be guaranteed, or, in old patients, as an alternative to surgery (Eisen and Voorhees, 1991). The presence of local invaded lymph nodes does not seem to influence the prognosis, at least in oral cavity mucosal melanomas (Guzzo *et al*, 1993).

Prospective studies of melanoma in this location are needed as their biological behaviour seems to be different from that of cutaneous forms.

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