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Case Report

A rare case of rynopharyngeal melanoma

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ABSTRACT

Primary mucosal melanomas (MM) of the head and neck region constitute 0.5-2% of all malignant melanomas. The rynopharynx is a region that is less often involved by malignant melanomas. Because most of mucosal melanotic lesions are painless in their early stages, the diagnosis is unfortunately often delayed until symptoms resulting from ulceration, growth, and/or bleeding are noted. Here, we document the rare case of a malignant rynopharynx melanoma of a 43 year old woman. Its treatment and the pertinent literature are discussed. No complication was recorded in the post-operative period and no further surgery was performed. The follow up showed no recurrence in the same position and with the same characteristics, even after six years. Mucosal melanomas are aggressive tumours and the prognosis in these patients is poor. Clinicians must use treatment strategies that provide functional benefit, so as to maintain quality of life without excessive toxicity.

Key Words: Mucosal malignant melanoma, rynopharynx, surgical management

INTRODUCTION

Primary mucosal melanomas (MM) of the head and neck region constitute 0.5-2% of all malignant melanomas.[1] The rynopharynx is a region that is less often involved by malignant melanomas.[2] Moore, *et al*.[3] reported that only 6.3% of all melanomas in the head and neck region are of mucosal origin. However, they are mostly located in the paranasal sinuses, hard palate and lips.[4]

The mean age at diagnosis of patients with MM is 55 years (range, 40-70 years)[5,6] and, unfortunately, these patients are candidates for limited surgical excision because of the high morbidity associated with extended surgical excision. Some authors believe that the rich lymphatic and capillary networks of the mucosa cause early dissemination of the disease so that mucosal melanomas are biologically more aggressive than their cutaneous counterparts, showing greater anaplasia, pleomorphism and vascular invasion.[7]

Because most of mucosal melanotic lesions are painless in their early stages, the diagnosis is unfortunately often delayed until symptoms resulting from ulceration, growth, and/or bleeding are noted.[8] Only 40% of the patients complained of pain or discomfort. In clinical appearance, most of these lesions were described as non pigmented, and they were diagnosed after discovery of a mass. Epistaxis is usually the most common presenting symptom among tumours of the nasal cavities, followed by obstructive symptoms, such as congestion and breathing difficulties.[9]

Wide resection of MM carries a high risk of morbidity. In addition, there is no definitive evidence that wide resection of mucosal malignant melanomas improves long-term survival. The presence of amelanotic melanoma makes diagnosis more difficult, inasmuch as most clinicians may consider melanoma only when confronted with a pigmented lesion. Therefore, any

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growing lesion, pigmented or non-pigmented, requires that a diagnosis be made, and it should be biopsied without delay. It may be appropriate to excise small masses, but large lesions should be sampled with incisional techniques.[10]

Because of the rarity of MM as a whole, and because of the unique biology and clinical challenges of MM arising from each anatomic location, understanding of this disease and its optimal management remains limited. The impact of various treatment strategies on disease control and survival has been difficult to assess because of the small size of most reported series of MM arising from any one particular site, the retrospective nature of most series, and the lack of a uniform comprehensive staging system for this disease.[11]

Here we document a rare case of malignant melanoma (MM) of the rynopharynx. Its treatment and the pertinent literature are discussed.

CASE REPORT

A-43-year old woman presented to Maxillofacial Surgery, Galeazzi Hospital, Milan, Italy in June 2006 for evaluation. On clinical examination, she presented dysarthria of speech, with signs of cerebellar ataxia. Her general medical history was contributory; with a history of malignant melanoma involving epipharynx treated in Germany twenty years earlier (1987), with a wide local excision, followed, one year later, by a right lateral neck dissection (the histological examination results were unknown). Additional details were not available but she also claimed to be suffering from Claude Bernard Horner Syndrome.

Since three years, the patient had experienced a progressive paralysis of the arms and legs. A few months ago, it had required hospitalization and proper neurological investigations. She had been diagnosed with a paraneoplastic cerebellar syndrome. Imaging studies used to evaluate this lesion, including positron emission tomography (PET), computed tomography (CT) and magnetic resonance imaging (MRI), revealed a large recurrence of malignant melanoma of rynopharynx [Figure 1]. Immediately, she underwent three cycles of adjuvant chemotherapy, waiting to be subjected to a surgical approach.

The latest imaging study to evaluate the clinical situation of patient, including MRI of brain and face showed no alteration of brain parenchyma, parapharynx, salivary glands and eyeballs, compared to previous studies.

Therefore, it was decided to submit the patient to an open surgical approach in order to remove the rynopharyngeal lesion. As the nose-palatal-genieno-flap was planned, a left paralateronasal incision was performed, extending from the lip up to the nasal root [Figure 2]. Then, the skeletonization and the osteotomy of the zygomatic bone, with release of the pterygoid process from jaw bones were done. Following this, the osteotomy of the palatine bone from 2.2 to 2.8 dental elements and the section of the soft palate from the origin to the hard palate were carried out in order to attain the exposition of the rynopharinx. The melanoma occupied the rear wall of the nasopharynx and the lateral wall bilaterally. It was decided to remove the mucosa and submucosa. The surgical specimen was sent to the Pathology Laboratory for definitive diagnosis. The gross specimen measured 4 × 3.5 cm [Figure 3] and contained part of the pharynx consisting of mucosa and submucosa and several biopsies of the surrounding areas. The microscopic examination revealed a specimen composed of fragments of mucous tissue with foci of chronic inflammation. Only the rynopharyngeal supero-posterior and superior-medial left borders were compromised by disease; other areas were free. Based on the microscopic findings and clinical history, a diagnosis of melanoma was done.

During the surgery, a tracheotomy was performed to prevent airway collapse and a nasogastric tube was positioned. No complication was recorded in the post-operative period and no further surgery was performed. The follow up showed no recurrence in the same position and with the same characteristics after six years.

DISCUSSION

The MM is a rare disease, and usually it carries a poor prognosis. Although in the past, there was some debate regarding the cellular origin of these tumours, the presence of melanocytes within the epithelial lining of the respiratory tracts is now well established.[12] It remains important, however, to differentiate primary mucosal tumours from metastases, because melanomas from other sites can metastasize (although this happens infrequently) to

organs lined with mucosal epithelia. For this reason, a prior history of cutaneous or ocular melanoma should be ruled out. Mucosal and cutaneous melanomas are known to be genetically distinct entities.

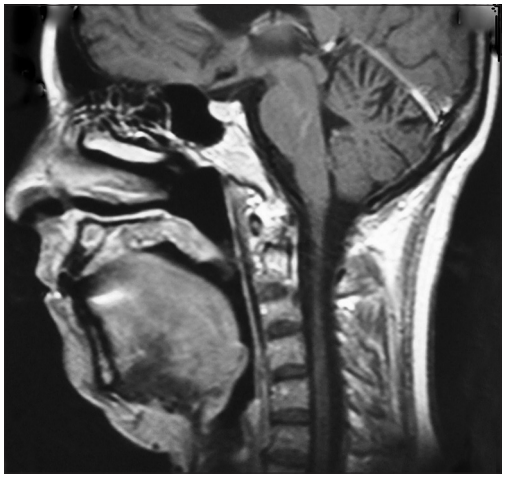

Figure 1: Pre-surgical Resonance Magentic image

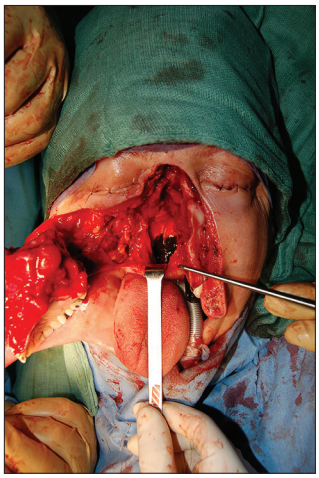

Figure 2: The surgical field showing the nose-palatal-genieno-flap

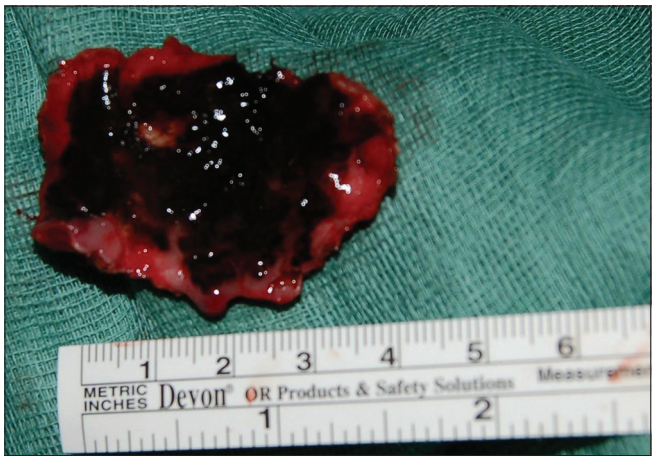

Figure 3: The rynopharyngeal melanoma excised

Bastian, *et al*. have reported that mucosal melanomas carry substantially more chromosomal aberrations and copy-number alterations than their cutaneous counterparts.[13] Importantly, the tyrosine kinase c-kit is over expressed and frequently mutated in mucosal melanomas, as opposed to rare c-kit alterations in cutaneous melanomas. No specific causative agents have yet been identified for mucosal melanomas. It is clear, however, that direct sun exposure, which has been strongly linked to cutaneous melanoma, probably is not involved in the pathogenesis of these tumours. It remains to be determined whether genetic predisposition, local micro environmental factors, infectious agents, or hormonal influences are involved.[14]

MM is a rare and aggressive neoplasm, with high rates of local, regional, and distant failure. Owing to the small size of most reported series and their retrospective nature, and the lack of uniform comprehensive staging system, the effect of various treatment strategies on disease control and survival has been difficult to assess. The optimal management of head and neck MM is not well defined. Surgical treatment has being advocated as the primary treatment modality, with growing consideration for postoperative radiotherapy, as wide surgical resection in the head and neck region is often difficult. Radiotherapy has recently been reported as a beneficial management modality, regardless of the fact that MM has been considered to be radioresistant. As significant morbidity is expected in high doses of radiotherapy to the head and neck region, new radiographic modalities with better precision are required. Furthermore, high-energy radiotherapy was suggested as a better therapy to mucosal MM due to the suggested biology of the tumour. The high rates of locoregional recurrence and distant metastasis also suggest that systemic treatment is needed. Currently, there is no role for adjuvant systemic therapy for patients who have been successfully resected, but recent developments in the understanding of the biology of melanoma and, in particular, specific growth pathways holds promise for the future.[15]

CONCLUSION

Here, a case of malignant melanoma of rynopharynx is reported. Mucosal melanomas are aggressive tumours and the prognosis in these patients is poor.

Clinicians must use treatment strategies that provide functional benefit, so as to maintain quality of life without excessive toxicity.

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