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Unknown primary nasopharyngeal melanoma presenting as severe recurrent epistaxis and hearing loss following treatment and remission of metastatic disease: A case report and literature review☆

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ABSTRACT

INTRODUCTION: Primary nasopharyngeal melanoma is an exceedingly rare pathology with unclear etiology and oftentimes obscure clinical presentation. Despite improved diagnostic capabilities, these lesions are often diagnosed at an advanced stage and associated prognosis is poor, partly due to high rates of recurrences and metastasis.

PRESENTATION OF CASE: A 74-year-old woman was diagnosed with metastatic melanoma to the liver, of unknown primary. Just prior to the time of diagnosis, she experienced several episodes of severe epistaxis which she managed conservatively. Her symptoms eventually subsided without further medical evaluation. The patient was initially treated with interleukin-2 (IL-2) for her advanced disease, but her cancer progressed. She was then enrolled in a protocol for percutaneous hepatic perfusion (PHP) with melphalan and had complete radiographic resolution of disease, yet her nosebleeds recurred and persisted despite conservative measures. Six years after her initial diagnosis, a nasopharyngoscopy demonstrated a pigmented lesion in the posterior nasopharynx. Surgical resection was performed (pathology consistent with mucosal melanoma) followed by radiation therapy. She has since had complete resolution of bleeding and shows no evidence of cancer.

DISCUSSION: To our knowledge, this is the first report of a diagnosis of primary nasopharyngeal melanoma 6-years following complete remission of metastatic disease. Surgery remains the primary treatment for disease and symptom control in this setting.

CONCLUSION: Timely diagnosis of nasopharyngeal melanomas remains challenging. Thorough clinical evaluations should be performed in such patients, and attention should be paid to recurrent and persistent symptoms, such as epistaxis and hearing loss. This may allow for earlier detection of primary disease.

1. Introduction

Whereas cutaneous melanoma is a familiar entity to many clinicians and patients alike, primary head and neck mucosal melanoma (HNMM) is a rare pathology with an unclear etiology and oftentimes obscure clinical presentation [1,2]. Similar to cutaneous melanoma, there has been an increase in the incidence of HNMM in the United States since the late 1980s, most notably in white females ages 55–84 [3,4]. Despite advances in diagnostic capabilities and treatment approaches, prognosis remains poor and the 5-year overall survival rate is 10–30%, partly due to high rates of recurrences and metastasis [4].

Nasopharyngeal melanoma is an exceedingly rare type of HNMM and its incidence is usually reported with nasal cavity and sinonasal tract melanomas. Mucosal melanomas in these locations account for 4% of HNMMs and 4% of nasal tract/nasopharyngeal neoplasms [5]. Hearing loss, epistaxis, and nasal congestion are presenting symptoms, however the diagnosis is often delayed several months following symptom onset [6,7]. We present a report of a woman who had a complete response to systemic treatment of

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metastatic melanoma of unknown origin. Six years later, a diagnosis of primary nasopharyngeal melanoma was made after an extensive work-up for recurrent symptoms of severe right-sided epistaxis and hearing loss. Herein, we describe the clinical decision-making and multidisciplinary management involved in the care of the patient.

2. Presentation of case

In February 2007, a 74-year-old woman presented with abdominal pain and generalized malaise. A CT showed multiple liver masses and subsequent biopsy was consistent with metastatic melanoma. Extensive work-up did not reveal a primary source of melanoma. Just prior to the time of diagnosis, she experienced severe epistaxis that she treated conservatively; these symptoms resolved and she did not pursue further medical work-up for her nosebleeds. She was initially treated with interleukin-2 (IL-2) for stage IV disease, but her cancer progressed. In November 2007, she enrolled in a National Institutes of Health (NIH) study investigating percutaneous hepatic perfusion (PHP) with melphalan for ocular or cutaneous melanoma metastatic to the liver. She tolerated two doses, but stopped therapy because of systemic toxicity. Despite no further treatment, her burden of disease decreased at subsequent visits and she was declared to have complete radiographic eradication of all tumor sites by April 2009.

In January 2013, the patient presented to an otolaryngology clinic for follow-up of chronic right-sided cerumen impaction and recurrent severe right-sided epistaxis. Following an unremarkable clinical exam, she was continued on a moisturization regimen with saline nasal spray, gel, and a humidifier. Symptoms persisted and the aforementioned regimen was modified to include other over-the-counter agents. She was later evaluated in April 2013 with similar complaints as well as diminished hearing. On exam, her physician noted right nasal turbinate swelling and septal ooze, as well as right ear cerumen impaction. Nasal packing was performed and the patient returned home, but bleeding persisted. The patient sought medical evaluation at a local hospital, and a limited nasal endoscopy was performed, revealing no bleeding source and a widely patent nasal cavity with no suspicious lesions. She subsequently returned to her original otolaryngology clinic and a more complete nasopharyngoscopy was performed, revealing a dark-nodular pigmented lesion in the right posterior nasopharynx with no evidence of satellite melanosis (Fig. 1). Imaging showed a 1.3 × 1 cm lesion in the right fossa of Rosenmuller obstructing the eustachian orifice and a right tympanomastoid effusion (Fig. 2). Staging PET scan demonstrated metabolic hyperactivity in this area extending into the ipsilateral torus tubarius and posteriorly into the eustachian tube orifice but without further evidence of local or metastatic disease. She underwent partial nasopharyngectomy, posterior septectomy, and resection of right torus in June 2013 via an endoscopic endonasal approach. Final pathology report demonstrated malignant mucosal melanoma (positive for S-100, HMB45, and Melan-A; BRAF and c-kit mutation-negative) and adjuvant radiation therapy (total of 70 Gy) was administered in September. She has since been doing remarkably well. In October 2013, several months following her surgery, examination of her right ear demonstrated a clear external auditory canal, with no evidence of effusion, retraction, or perforation. Surveillance imaging, clinical exams, and laboratory studies through November 2014 have been unremarkable for recurrent or metastatic disease, and she has had no further episodes of epistaxis.

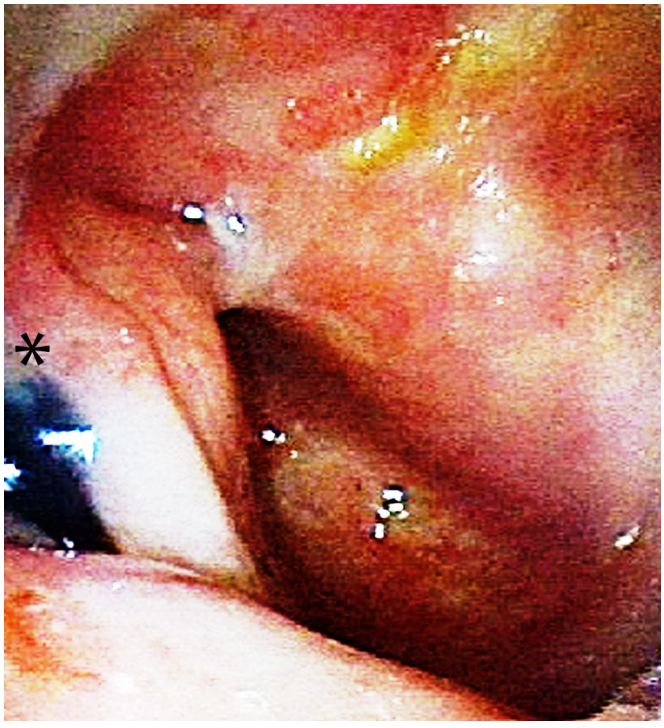

Fig. 1. Endoscopic evaluation revealed a dark melanotic lesion located in the right nasopharynx (bottom left).

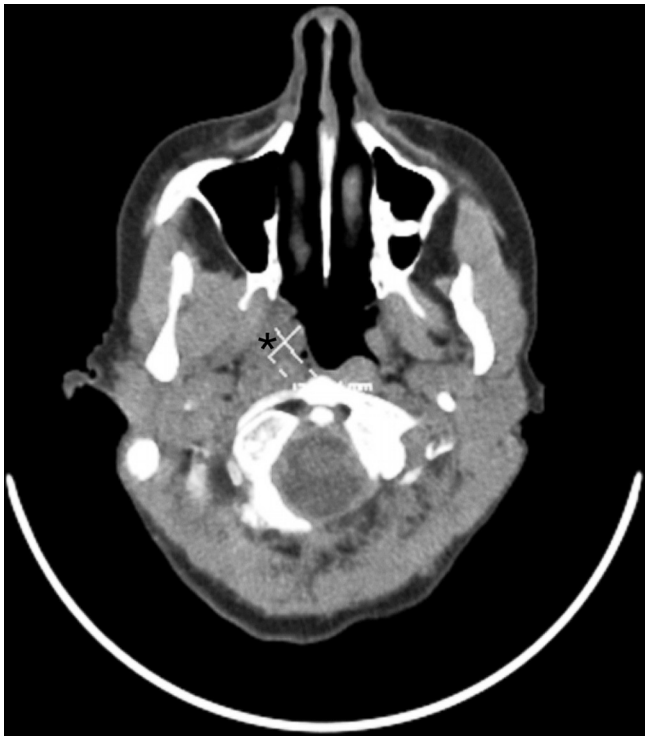

Fig. 2. Computed-tomography demonstrating a 1.3 × 1 cm lesion in the right fossa of Rosenmuller involving the eustachian orifice and tympanomastoid effusion.

3. Discussion

Primary mucosal melanoma accounts for approximately 0.03% of all cancer diagnoses, and head and neck cases account for 0.7–3.8% of melanomas [2,8]. Given its rarity, much of the management approach to nasopharyngeal melanoma is derived from