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Primary Melanoma of the Palatine Tonsil in an Adult Filipino Patient: A Case Report

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ABSTRACT

Objectives: To discuss a case of primary melanoma of the palatine tonsil in a 57-year-old man presented with a dark, pigmented tonsillar mass initially managed as a case of arterio-venous malformation, and review the literature on its presentation, diagnosis, management and outcomes.

Methods:

Design: Case Report
Setting: Tertiary Government Training Hospital
Patient: One

Results: A 57-year-old man presented with a pigmented, bluish-black mass (7.2 cm) on the right tonsillar area with dysphagia and odynophagia. A CT scan interpretation considered large tonsillar malignancy, right with infiltrations of the soft palate, lingual tonsils and pre-epiglottic space. The initial impression was an arteriovenous malformation and preoperative arterial embolization was followed by a tonsillectomy. The final biopsy result was mucosal melanoma. Refusing further treatment, he expired nine months later in the emergency room, after presenting with decreasing sensorium and desaturations, jaundice and abdominal distension.

Conclusion: To the best of our knowledge, this is the first reported case of tonsillar melanoma in the Philippines. Primary tonsillar melanoma is rare but its diagnosis is still possible (although it is usually diagnosed in advanced stages). Despite improvement in surgical techniques and adjuvant therapies, its prognosis remains poor. Regular oral cavity screening may help in early detection.

Keywords: *palatine tonsil; melanoma; primary; mucosal*

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Melanoma is a tumor produced by the malignant transformation of melanocytes derived from the neural crest. The incidence of melanoma has rapidly increased worldwide. Its incidence is increasing faster than that of any other cancer except lung cancer in women.[1] Melanoma usually occurs on the skin. According to the GLOBOCAN 2020 report, cutaneous melanoma is the 18th most common malignancy worldwide.[2] However, neural crest cells migrate and can develop into melanoma in other locations such as the brain, head and neck mucosa, and genitourinary and gastrointestinal tract.[3] While the head and neck comprised the majority of the extracutaneous melanoma, a primary melanoma of the tonsil is exceedingly rare with only 27 reported cases as of 2018.[4] Herein, we report another case of primary melanoma of the palatine tonsil and review the literature on its presentation, diagnosis, management and outcomes.

CASE REPORT

A 57-year-old Filipino man presented to our institution due to a mass on the right palatine tonsil with bluish-black discoloration. Five months prior, the patient noted mass in the right tonsillar area associated with intermittent odynophagia. A discoloration with gradual enlargement of the tonsil was noted after a month. Odynophagia was temporarily controlled with mefenamic acid. The mass eventually progressed to have a black discoloration which prompted consult with a private ENT specialist. However, the patient refused to undergo a tonsillar biopsy, and was referred to a government hospital for further work-up and management.

Our patient did not have any medical comorbidities or take any maintenance medications. There was no history of weight loss, early satiety or heredofamilial disease such as malignancy. He was a 10-pack year smoker and an alcoholic beverage drinker.

Oropharyngeal examination revealed a Brodsky 3+ bluish black, exophytic mass with ulceration at the right tonsil. *(Figure 1)* Both nasal endoscopy and otoscopy were unremarkable. There was no suspicious/malignant-looking lesion on his skin and no palpable cervical lymphadenopathies. The cranial nerve examination was within normal limits. The initial impression was an arteriovenous malformation of the tonsil thus an angioembolization was advised. Due to the size of the mass and pending schedule for embolization, an elective tracheostomy was performed since airway compromise was imminent, with the patient complaining of dysphagia to solids and 2-pillow orthopnea.

A computed tomography (CT) angiography of the head and neck showed a fairly defined, heterogeneously enhancing soft tissue mass within the region of the right tonsillar pillar measuring 7.2 x 5.6 x 4.9 cm with narrowing of the oropharyngeal space and nasopharynx. *(Figure 2)* The right parapharyngeal space and vallecula were obliterated. Bilateral level IIA cervical lymph nodes were enlarged, the largest measuring 0.7cm. The tonsillar branch of the right facial artery was supplying the mass. A tonsillar malignancy was also considered during this time.

The patient eventually underwent a trans-arterial embolization of the feeding artery using Onyx™ Liquid Embolic System (LES) glue (Medtronic, Minneapolis, MN, USA). *(Figure 3)* Two days after embolization, a unilateral tonsillectomy without margins was performed by a senior resident. The final biopsy of the specimen was reported as consistent with mucosal melanoma. *(Figure 4)* Two weeks after surgery, the patient followed up at the clinic without significant change in his status. He was advised a metastatic work-up and neck dissection. However, he refused to undergo further diagnostics and management.

Nine months post-tonsillectomy, the patient was brought back to the emergency room with decreasing sensorium and desaturations, and physical examination revealed a distended abdomen and jaundice. He expired after 3 hours on mechanical ventilation. The case was signed out as acute respiratory failure type 2 secondary to hepatic encephalopathy, probably secondary to hepatic metastasis from malignant melanoma Stage IV, right tonsillar area (T4bN1M1) status post tracheostomy tube insertion and right tonsillectomy (July 2021).

DISCUSSION

Up to 90% of head and neck melanomas are cutaneous lesions, commonly occurring on the face.[5] In contrast, mucosal melanomas (MM) in the head and neck only comprised less than 1% of all melanomas with the most common locations being the oral cavity, paranasal sinuses and nasal cavity.[6] Melanomas occurring in the oral cavity should always be among the possible differential diagnoses if oral pigmentations are discovered during examination, since they are considered to be worrisome and life-threatening conditions.[7]

The pathogenesis and biologic behavior of MM of the head and neck are poorly understood. It is unlikely that ultraviolet radiation from prolonged sun exposure, which is a major risk factor for cutaneous melanoma, is implicated in MM. However, cigarette smoking has been suggested as a risk factor since pigmented oral lesions are more prevalent (up to 37%) among smokers.[6] Oral MM has a higher incidence among Asians, Asian-Indians, Hispanics, and Africans and their peak incidence is in the 60s without gender preference.[7]

Among the mucosal melanomas, primary palatine tonsil melanomas are considered extremely rare. In a 2018 literature review, Osorio *et al*.[4] compiled for the first time all reported cases, 26 in total, of primary tonsillar melanoma. We searched MEDLINE (PubMed), Google scholar, HERDIN plus, the ASEAN Citation Index (ACI), the Western Pacific Region Index Medicus (WPRIM), and the grey literature using the keywords

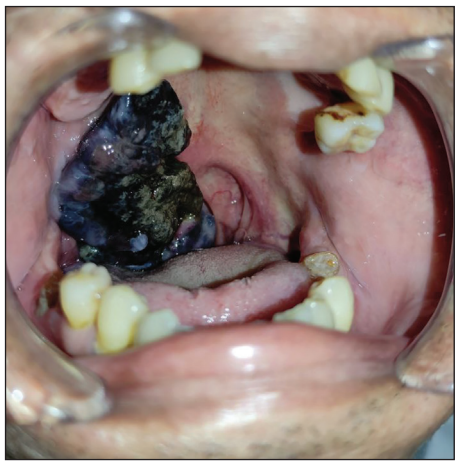

Figure 1. Oral cavity examination showing a raised, lobulated mass with bluish-black discoloration over the right tonsillar area extending to the soft palate and oropharynx

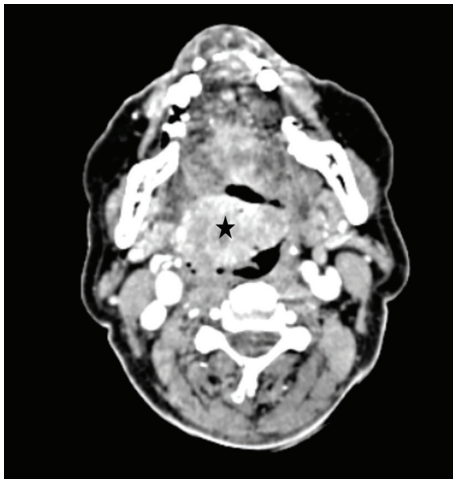

Figure 2. Contrast-enhanced CT-scan, axial view at the level of the oropharynx showed a 7.2 x 5.6 x 4.9 cm fairly defined, heterogeneously enhancing soft mass within the region of the right tonsillar pillar (star)

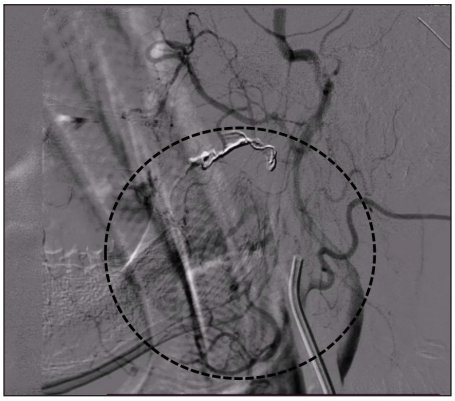

Figure 3. Computed Tomography-guided trans-arterial embolization; facial branch of the right external carotid artery embolized using Onyx™ glue. A large tumoral blush is seen in the right oropharyngeal area (dotted lines).

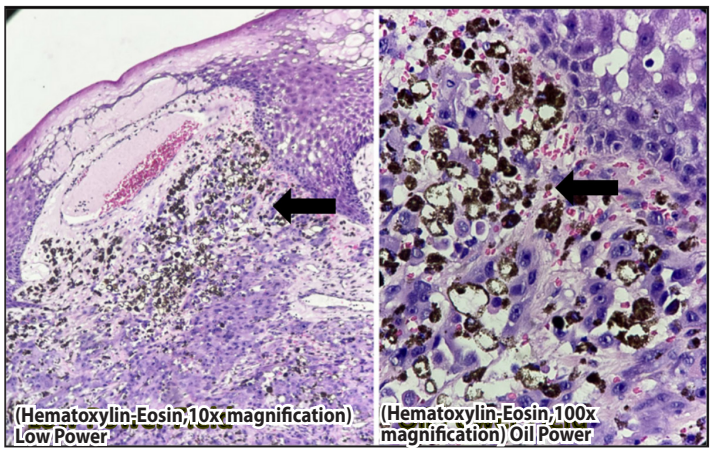

Figure 4. Microscopic examination, Hematoxylin-Eosin stains, left, low power (10x magnification); and right, Oil immersion (100x magnification), showed atypical melanocytes exhibiting marked cytologic atypia with invasion to the surrounding tissue, and with prominent melanin pigmentation (arrows)

"tonsil" AND "melanoma" and found four additional cases bringing the total number of reported cases to 30.[8, 9, 10,11] believe there are now 31 cases in total, including our report. We can summarize these cases, excluding 10 of the cases from the review paper of Osorio *et al.*[4] as they were not reported with sufficient information. The mean age of presentation was 54.5 $\pm$ 13.7 years and more than 80% of the patients were male. The average size of the mass was 3.7 cm (range 0.85 – 7.6) with no predilection on the laterality. They typically appear as a bluish-black mass with the pigmentation of the tonsillar area and patients commonly complain of dysphagia and odynophagia. More than 30% of the patients also presented with cervical lymphadenopathy. [4, 8-11]

Going back to our present case, based on gross examination of the mass, a differential diagnosis of oropharyngeal/tonsillar arteriovenous malformations (AVM) was initially considered, although the subsequent CT scan interpretation suggested malignancy. With the initial consideration of a vascular tumor, we still referred the patient to Interventional Radiology for trans arterial embolization prior to tonsillectomy for excision of mass and final biopsy. This remains to be the most conventional modern approach for AVMs.[12]

Metastasis of a skin melanoma to the tonsil is also rare with fewer than 30 cases reported in the literature. The average occurrence of this metastasis is 6 years from its presentation but with a 5-year survival rate of less than 5%.[13] Since there is difficulty in differentiating a primary or metastatic tonsillar melanoma, it is prudent to thoroughly examine the patient for a primary cutaneous source.

Primary work-up of mucosal melanoma includes contrast-enhanced CT scan and/or magnetic resonance imaging (MRI) to determine the anatomic extent of disease. Chest and abdominopelvic CT scan, brain MRI, and/or fluorodeoxyglucose (FDG)-positron emission tomography

(PET)/CT scans are necessary for metastatic workup when clinically indicated.[14]

Most of the reported cases of primary palatine tonsillar melanoma were confirmed after tonsillectomy.[4] Because of its rarity, tonsillar melanoma is treated in the same manner as other mucosal melanomas of the head and neck. The National Comprehensive Cancer Network (NCCN) guidelines for mucosal melanoma recommend surgery for localized mucosal disease (T1 - T3) or advanced local disease (T4a) involving the deep structures/soft tissue with or without regional lymph node metastasis. Surgical procedures specific to reported cases of tonsillar melanoma range from simple tonsillectomy to a more radical surgery including pharyngectomy with flap reconstruction, and resection of the soft palate and constrictor muscles.[4] In patients with a confirmed nodal disease or T4a lesion, neck dissection and postoperative radiation are recommended to improve locoregional control. However, as of this writing, the NCCN guidelines prefer a multidisciplinary approach and enrollment of patients to clinical trials when lesions are T4a or have a nodal disease.[14] Additionally, the surgical specimen should be verified using appropriate staining such as HMB-45, S-100, and/or Melan-A.

Systemic therapy for mucosal melanoma is similar to what is used for cutaneous melanoma including Anti PD-1 and combined targeted therapy such as Dabrafenib/trametinib. However, they are commonly used in metastatic or unresectable diseases. Currently, there is no data to support its use in the adjuvant setting.[14]

Based on the review of Osorio *et al.* two patients already had metastatic disease to the lung and gastrointestinal tract, respectively, and patients succumbed to their disease within 4 to 12 months.[4] Oral mucosal melanomas are preceded by oral pigmentations that last several months to years without any symptoms. This may explain patients' delay in seeking medical attention.[7] In addition to its dismal prognosis, prompt recognition and treatment are paramount.

The NCCN guidelines recommend a follow-up interval of every 1 to 3 months in the first year, every 2 to 6 months in the second year, every 4 to 8 months on years 3 to 5, and annually thereafter.[14] However, the frequency of medical examination/follow-up often is not standardized. Relapse rates of oral melanomas, in general, have been reported to be 10% to 20% after complete surgical excision and appropriate adjuvant therapy; even relapses beyond the 10th year have been documented.[6, 15]

To the best of our knowledge, this is the first reported case of tonsillar melanoma in the Philippines. Primary melanoma of the palatine tonsil is rare, and prognosis remains poor despite improvement in surgical techniques and adjuvant therapies. Although it is usually diagnosed in advanced stages, its early detection may still be possible with regular oral cavity screening.

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