



Transoral Robotic Excision of Hemangioma Tongue in Rare Case of Maffucci Syndrome

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Abstract

Maffucci syndrome is an extremely rare disorder characterized by benign overgrowths of cartilage (enchondromas), skeletal deformities and cutaneous lesions composed of abnormal blood vessels (hemangioma). Enchondromas arise in bones, most frequently in the hands and feet, and less often in the legs and long bones of the arm. We present a case report of hemangioma right side base of tongue and operated enchondroma left foot labelled as Maffucci syndrome. Patient underwent MRI contrast neck and after confirming the diagnosis, patient underwent robotic tongue base excision of hemangioma. No blood transfusion was required as robotic surgery is minimally invasive with minimal bleeding. Patient stay in hospital was only 1 day and recovered completely after the surgery and is doing well currently with no recurrence.

Keywords: Maffucci syndrome; Hemangioma tongue; Transoral robotic surgery

Introduction

Maffucci syndrome is a rare disease first reported in 1881. Maffucci syndrome is characterized by the presence of multiple enchondromas combined with multiple soft tissue hemangiomas or lymphangioma [1]. There is a tendency for malignant transformation of enchondromas into chondrosarcomas or of hemangiomas into vascular sarcomas. Patients with Maffucci syndrome also are susceptible to the development of other malignant lesions such as glioma. It is caused by somatic mutations in the Isocitrate Dehydrogenase 1 (*IDH1*) or *IDH2* genes [2-4]. Enchondromas in the metaphyseal regions of long bones may also result in deformity and limb asymmetry, as well as pathological fractures.

Case Presentation

A 33 years old female came with chief complaints of foreign body sensation in throat for 20 days. It was insidious in onset, gradually progressive. Not relieved on medications. Patient also gives history of difficulty in swallowing though not very significant. Patient also gives history of bleeding from mouth after touching the back of the tongue with finger, one episode which resolved on its own. Patient gives past surgical history of enchondroma of left foot which was excised surgically. On oropharyngeal examination there was swelling on right side base of tongue, soft in consistency, reddish appearance, and non tender (Figure 1). Rest of the ear, nose and larynx examination was normal. There was no palpable neck node (Figure 2). Patient was clinically diagnosed as hemangioma right side base of tongue. Patient underwent MRI neck with contrast which showed a well defined lesion arising from the base of tongue on right side measuring 9 mm × 7 mm and directed posteroinferiorly. It is displaying hyperintense signal on T2 and intense contrast enhancement in contrast study which is seen to persist in delayed phase. No evidence of diffusion restriction in it. Muscle of tongue shows normal signal intensity. Patient underwent Robotic (Da Vinci robot) excision of hemangioma tongue with wide margins. Tissue was sent for histopathology which was suggestive of hemangioma. Patient recovered well and is in follow up with no recurrence.

Discussion

Maffucci syndrome is one of several subtypes of enchondromatosis. Others include Ollier disease, which occurs with more frequency than Maffucci as well as metachondromatosis, genochondromatosis, spondyloenchondrodysplasia, dyspondyloenchondromatosis and cheirospondyloenchondromatosis which are even rarer. While both Ollier disease and Maffucci

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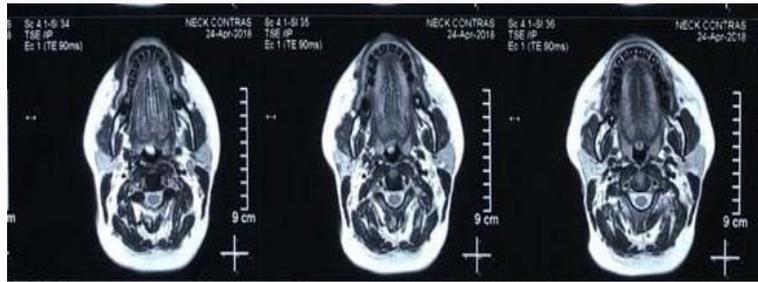


Figure 1: Preoperative scan showing contrast enhanced haemangioma on right side base of tongue.

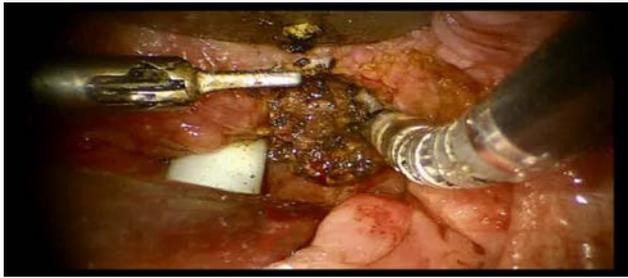


Figure 2: Intraoperative picture showing haemangioma with normal cuff of tissue.

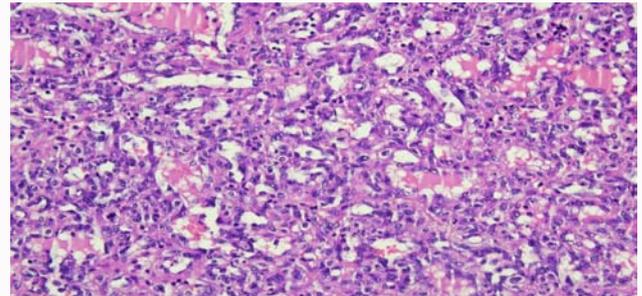


Figure 4: Histopathology slide 2 (Magnification 4X)-Vessels lined by bland plump endothelial cells. Mild infiltrate of lymphocytic plasma cells in between the vessels.

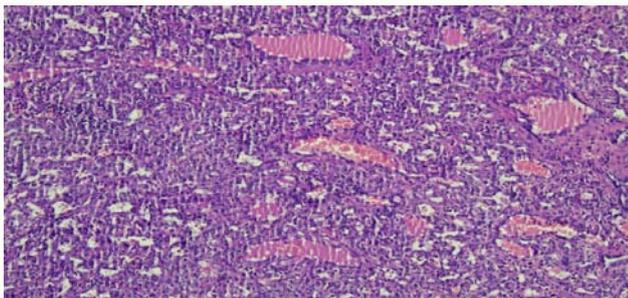


Figure 3: Histopathology slide 1 (Magnification 10X) - Ulcerated mucosa of tongue with submucosa showing a benign vascular neoplasm composed of proliferative closely packed capillary sized blood vessels.

syndrome are characterized by multiple, often unilateral enchondromas, Maffucci syndrome is distinguished from Ollier disease by the presence of hemangiomas and/or lymphangiomas [1]. Due to the rarity of the disease the incidence of malignant transformation of the lesions is not known but some reports have estimated it to be between 23% and 100% [5].

There are two primary challenges in the management of patients with Maffucci syndrome. The first is the management of the bony deformities which characterize the disease (Figure 3).

The second challenge is surveillance for malignancies, not only the malignant transformation of the primary enchondromas but also the associated soft tissue malignancies. At present, there is no standard protocol for monitoring patients with Maffucci syndrome [6] (Figure 4).

In this case we detected hemangioma base of tongue which was removed by transoral robotic surgery. Detail history revealed surgery for enchondromatosis. We are of opinion that enchondromatosis patient should be followed regularly to detect other tumors in early

stage. Literature mentions that Maffucci syndrome patient to be monitored by contrast enhanced MRI annually [7]. Base of tongue hemangioma is not very rare. Suspicion for Maffucci syndrome & detail history is advised before proceeding in these cases. Da Vinci Robotic system is minimally invasive & of great tool to operate tongue base tumors. The advantage of TORS is stay in hospital is less than 1 day & no need to arrange for blood transfusion as even in hemangioma there is very minimal bleeding.

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