



This work is licensed under a Creative Commons Attribution-NonCommercial 4.0 International License.

Hemangioma Turned Into a Gigantic Tongue: A Rare Case of Maffucci Syndrome

Kevser Peker

ABSTRACT

Background: This is a rare case of Maffucci syndrome who underwent orthopedic surgery owing to enchondroma and who had hemangiomas in the tongue tip.

Case Report: After anesthesia induction, a 60-year-old male patient was intubated with direct laryngoscopy without any trauma to the hemangioma. However, before extubation, the edema in the patient's tongue was observed to slightly increase. After the endotracheal tube was removed, the patient had stridor, the airway was obstructed, the patient could not tolerate this condition, and he was subsequently reintubated. It was observed that the hemangioma in the tongue tip was dilated. The patient was followed up in the intensive care unit for 1 day and was extubated the following day with the reduction of hemangioma and tissue edema.

Conclusion: The examination of hemangiomas, especially in the upper airway, requires careful attention from an anesthetic point of view.

Keywords: Anesthesia, airway, gigantic hemangioma, Maffucci syndrome

INTRODUCTION

Maffucci syndrome was first described in 1881 (1). Its clinical manifestations are cutaneous, soft tissue, and visceral hemangiomas and multiple cartilaginous masses of the bones of the extremities (2). The etiology is not clear. Hemangiomas associated with Maffucci syndrome vary between 5% and 10% in the head and neck region, and oral manifestations are limited to occasional hemangiomas (3). The examination of enchondroma and hemangiomas, especially in the upper airway, requires careful preparation from an anesthetic point of view (4).

After a written consent was obtained from the patient, we report a case of hemangiomas at the tongue tip that developed into severe respiratory distress in the early postoperative period and was subsequently diagnosed as having Maffucci syndrome.

CASE REPORT

A 60-year-old male patient was admitted to the department of orthopedics and traumatology because of increased pain in a lesion on a finger. This lesion was considered as enchondroma, and the patient was taken to the operating room. Because the surgical team planned to take an iliac wing graft, general anesthesia was considered for the patient. The patient had hemangioma at the tongue tip with Mallampati classification III. An otorhinolaryngology consultation was requested. They did not consider intervention for hemangioma. The other difficult airway predictors were not seen. Necessary preparation (stylet, bougie, videolaryngoscope, fiberoptic bronchoscope device) was done owing to the anticipated difficulty with intubation. In the operating room, after routine monitoring, the patient was induced with fentanyl 1 mcg/kg, 2 mg/kg propofol, and 0.6 mg/kg rocuronium. The patient was intubated (spiraled endotracheal tube of size 7.5 + stylet) using direct laryngoscopy (Cormack-Lehane grade 2b) without trauma to the hemangioma at the tongue tip. Anesthesia was maintained using 2% sevoflurane in a mixture of 60% nitrous oxide and 40% oxygen. The intraoperative course of the surgery was uneventful, and the patient was hemodynamically stable. At the end of the surgery, it was observed that the hemangioma at the tongue tip and tissue edema had slightly increased in the patient's tongue before extubation. Necessary antiedema treatment was performed. Sugammadex was used for decurarization. After adequate ventilation was observed, the endotracheal tube was removed with the suggestions of the otorhinolaryngology (with the preparation for emergency tracheostomy). Then the patient had stridor, the airway was obstructed, and he was subsequently reintubated because of the gigantic hemangioma (Fig. 1a). At the end of the surgery, the decision for elective mechanical ventilation for the patient was made and the patient was transferred to the intensive care unit (ICU). Methylprednisolone

Cite this article as:
Peker K. Hemangioma Turned Into a Gigantic Tongue: A Rare Case of Maffucci Syndrome. Erciyes Med J 2021; 43(1): 90-2.

Department of Anesthesiology and Critical Care, Kirikkale University Faculty of Medicine Hospital, Kirikkale, Turkey

Submitted
14.02.2020

Accepted
01.06.2020

Available Online Date
11.12.2020

Correspondence
Kevser Peker,
Kirikkale University Faculty of
Medicine Hospital,
Department of Anesthesiology
and Critical Care,
Kirikkale, Turkey
Phone: +90 318 357 52 82
e-mail:
drbacan@hotmail.com

©Copyright 2021 by Erciyes
University Faculty of Medicine -
Available online at
www.erciyesmedj.com



Figure 1. (a) Patient with gigantic hemangioma. (b) Reduction of hemangioma in the intensive care unit

(250 mg) was administered to the patient at 6-hour intervals. The patient was followed up in the ICU for 1 day and was extubated the following day with the reduction of hemangioma and tissue edema (Fig. 1b). He was sent to the ward at postoperative day 2.

Tissue samples were compatible with enchondroma, and abdominal ultrasonography revealed a nodular appearance on the liver consistent with subcapsular hemangioma (17.5×9 mm hemangioma). The radiologic findings in contrast-enhanced tomography were reported as lobulated mass cartilage characterized by osteolytic lesions associated with a large number of punctate calcifications. These radiologic findings and the clinical examination contributed to the diagnosis of Maffucci syndrome. The patient's follow-up was planned accordingly.

DISCUSSION

The diagnosis of Maffucci syndrome is made with radiologic imaging and clinical evaluation. The genetic and distinctive biochemical properties of the syndrome are not yet clear. Thus, in the present case, the patient was diagnosed as having Maffucci syndrome as a result of radiologic and histopathologic findings and the clinical presentation of vascular malformations as hemangioma in the tongue, hemangioma in the liver, enchondroma in the extremity, and deformations in the bone structure.

Ollier disease is also characterized by enchondromas, as in Maffucci syndrome. In Ollier disease, it was reported that enchondromas start to be observed in early childhood (5). However, the presence of multiple hemangiomas in early childhood differentiate Maffucci syndrome from Ollier disease (6). In addition, the hemangiomas on the tongue tip of the present patient had existed since childhood.

In terms of anesthesia in Maffucci syndrome, the location of hemangiomas and enchondromas is important. Fiberoptic bronchoscopy and intubation should be preferred for patients with oral cavity hemangiomas so as not to tear the hemangioma (6). In this case, the patient was intubated using direct laryngoscopy without any damage. However, at the end of the surgery, the hemangioma extended to the root of the tongue resulting in tissue edema, and the tongue became gigantic.

The hemangiomas in Maffucci syndrome can sometimes be visceral (4). In the present patient, the radiologic results were consistent with liver hemangioma. In addition, it should never be forgotten that the hemangiomas and enchondromas in Maffucci syndrome may show malignant transformation and necessary follow-up should be made (6).

Enchondromas are intramedullary tumors caused by hyaline cartilage modification and may cause pathologic fractures as a result of osteophytic changes (7). Complications include spontaneous fractures, skeletal deformations, and malignant transformation (7). In most cases, enchondromas are seen in the bones of the extremities, especially in the hands and feet. However, enchondromas may also occur from the sinonasal canal, the petrous apex, the cerebellopontine angle, and the mandible, skulls, ribs, and vertebrae (4). The importance of these tumors for anesthesia in these patients is that careful positioning is necessary to avoid pathologic fractures because of their presence and location (8). In addition, localization of tumors in the airway tract may require difficult airway equipment such as tracheostomy.

Maffucci syndrome is a sporadic disorder occurring only in isolated patients without familial transmission. Whether there is a single gene defect or a combination of somatic mutations is unclear (9). The presence of such mutations may be guiding through future studies to understand the malignant transformations in these patients. In terms of anesthesia, patients with Maffucci syndrome with malignant transformations will require more attention during the perioperative and postoperative management.

Various histologic variants have been described in terms of vascular malformations observed in Maffucci syndrome. Cavernous vascular cavities in some cases may cause thrombus and phlebitis (10). This also requires careful consideration in these patients in the perioperative period.

For the present patient, the treatment for this type of benign lesion was conservative. Therefore, to evaluate the development of new enchondromas and hemangiomas and other malignancies such as ovarian or liver cancer, it was decided to perform periodic follow-up with radiographic imaging every 3 months.

In conclusion, because these lesions may cause a difficult airway, the observation of enchondroma and hemangiomas especially in the upper airway in a patient with Maffucci syndrome requires careful preparation (fiberoptic bronchoscopy for intubation to avoid tearing the lesions, tracheostomy preparation for difficulty because of bleeding) from an anesthetic point of view.

Informed Consent: Written informed consent was obtained from patients who participated in this study.

Peer-review: Externally peer-reviewed.

Conflict of Interest: The author have no conflict of interest to declare.

Financial Disclosure: The author declared that this study has received no financial support.

REFERENCES

1. Maffucci A. Di un caso encondroma ed angioma multiplo. [Article in Italian]. *Mov Med Chir* 1881; 3: 399–412.

2. Fletcher CDM, Bridge JA, Hogendoorn PCW, Mertens F, editors. WHO Classification of Tumours of Soft Tissue and Bone. Pathology and Genetics of Tumours of Soft Tissue and Bone. 4th edition. Lyon: IARC Press, 2013.
3. Moorthy AP. Oral manifestations in Maffucci syndrome. *British Dental J* 1983; 155: 160–1. [\[CrossRef\]](#)
4. Singh GP, Bindu B, Pandia MP, Bithal PK. Maffucci syndrome: Anaesthetic management and review of literature. *J Neuroanaesthesiology and Crit Care* 2015; 2(02): 136–8. [\[CrossRef\]](#)
5. MacGillis K, King D. Nonoperative Management of Multiple Hand Enchondromas in Ollier Disease With Progressive Ossification. *Am J Orthop (Belle Mead NJ)* 2015; 44(9): E343–6.
6. Prokopchuk O, Andres S, Becker K, Holzapfel K, Hartmann D, Friess H. Maffucci syndrome and neoplasms: a case report and review of the literature. *BMC Res Notes* 2016; 9: 126. [\[CrossRef\]](#)
7. Bisaccia M, Colleluori G, Meccariello G, Piscitelli L, Falzarano G, Medici A, et al. A rare case of Maffucci syndrome. From the diagnosis to the management. *Canadian Open Orthopaedics and Traumatology J* 2016; 3(1):1–8.
8. Zhou X, Zhao B, Keshav P, Chen X, Gao W, Yan H. The management and surgical intervention timing of enchondromas: A 10-year experience. *Medicine (Baltimore)* 2017; 96(16): e6678. [\[CrossRef\]](#)
9. Hopyan S, Gokgoz N, Poon R, Gensure RC, Yu C, Cole WG, et al. A mutant PTH/PTHrP type I receptor in enchondromatosis. *Nat Genet* 2002; 30(3): 306–10. [\[CrossRef\]](#)
10. Pansuriya TC, Kroon HM, Bovée JV. Enchondromatosis: insights on the different subtypes. *Int J Clin Exp Pathol* 2010; 3(6): 557–69.