Massive oral hemorrhage secondary to atypical expression of Maffucci's syndrome: A case report

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A healthy adult male who had massive oral hemorrhage was diagnosed to have atypical presentation of Maffucci's syndrome. Extensive search of the literature confirmed the reports of this syndrome involving maxillofacial region only in two cases till date. **(Int Chin J Dent 2002; 2: 60-62.)**

Key words: Maffucci's syndrome, massive oral hemorrhage.

INTRODUCTION

Maffucci's syndrome is a congenital disorder that has been described among patients of all races with no sexual predilection.[1] It is characterized by the presence of multiple enchondromas and soft tissue angiomas. The presenting symptom of the patient may be skeletal or vascular. Skeletal lesions appear as dyschondroplasias where as soft tissue lesions consist of typical cavernous hemagiomas.[2] The visceral involvement by hemagiomas has been reported in 17% of the cases with Maffucci's syndrome and only 2 cases involving maxillofacial region have been so far found in literature.[2,3]

CLINICAL REPORT

A 50-year old man was referred from the medical unit to diagnose and manage recurrent massive oral hemorrhage. History revealed that the patient used to develop massive oral hemorrhage which used to last for short duration. As the frequency of hemorrhagic attacks increased, the amount of blood loss warranted admission to the hospital. At the time of referral, the patient had received supportive treatment for the hemorrhage which had occurred 24 hours earlier. Patient was conscious and in a state of hyperdynamic circulation. The complete blood picture and coagulation profile were found to be normal.

Intra oral examination of the patient revealed diffuse angiomatous lesions extending on both sides of the tongue involving the ventral surface and lateral border present since childhood (Fig. 1). The lesion was soft in consistency and bluish red in color. On examination of the body there was a subcutaneous angioma on the left side of the chest wall (Fig. 2). Since the patient had experienced recurrent massive oral hemorrhage, and in order to prevent further episodes, elective bilateral lingual artery ligation was done. Biopsy of the intra

oral lesions were taken which confirmed the clinical diagnosis of cavernous hemangiomas.

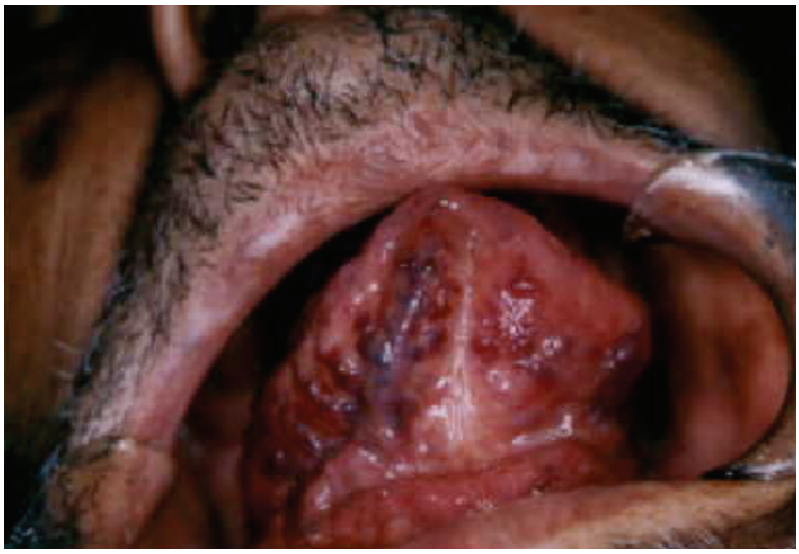

Fig. 1. Bluish-red colored diffuse angiomas on the ventral surface lateral aspects of the tongue.

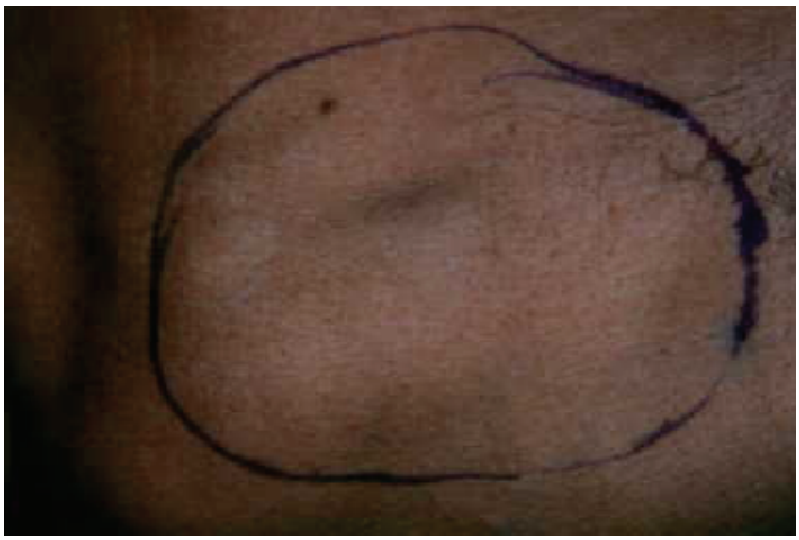

Fig. 2. Subcutaneous angioma on the left side of the chest wall.

DISCUSSION

Maffucci described this syndrome characterized by enchondromatosis, bone deformities, hemangiomas and phlebolithiasis. Kast and Von Recklinghausen described it in 1859. This syndrome has been reviewed by Canleton, Bean, Anderson, Lewis, and Ketcham.[4] This syndrome should not be confused with Ollier's disease, Klippel-Trenaunay-Weber syndrome and Proteus syndrome.

In the above reported case, as the angiomas involved large areas of the tongue bilaterally and to prevent further oral hemorrhage, elective bilateral lingual artery ligation was done. The patient is kept under constant observation to rule out the possibility of malignant transformation of the lesion.

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