Maffucci’s Syndrome with Oral Manifestations

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Maffucci’s syndrome is a rare congenital mesodermal dysplasia combined with dyschondroplasia and hemangiomatosis. About 150 cases have been reported till now. Maffucci’s syndrome is often combined with other neoplasms. Herein, we report a patient with Maffucci’s syndrome and hemangiomas on the dorsum of the tongue, which is rare in this syndrome.

Keywords: Chondroma • hemangioma • Maffucci • oral manifestations

Introduction

Maffucci’s syndrome is a rare congenital nonhereditary mesenchymal dysplasia manifested by enchondromatosis (dyschondroplasia) and soft tissue hemangiomas, which has a high incidence of malignant transformation. Till now, about 150 cases have been reported in the literature.1,2

Maffucci’s syndrome affects the skin and skeletal system with multiple enchondromas and hemangiomas occurring during the first two decades of life.3,4

Although the distribution of the osseous and soft tissue alterations is similar, the sites of enchondromas and hemangiomas need not coincide. Involvement of the viscera and mucous membranes by hemangioma is a rare finding in this syndrome.5,6

A review of the literature shows that about 5% to 10% of the reported cases have some involvement in the head and neck region.7 Oral manifestations are limited to occasional hemangiomas.7

The management of skin and oral lesions are surgical and depends on the size and location of the lesions. Severe affection of the involved site may lead to amputation.5

We report a case of Maffucci’s syndrome with hemangiomas on the dorsum of the tongue, which is rare in this syndrome.

Case Report

A 23-year-old man presented with chief complaint of two painless nodular masses on the dorsum of his tongue, which interfered with mastication and swallowing. He was suffering from multiple enchondromatosis in his left hand with deformity and shortening.

The bony lesions of the hand had appeared at the first year of his life and its shortening was caused by previous corrective surgery.

Multiple round- to oval-shaped cartilaginous nodules were noticed in his affected hand. Some small blue cutaneous painless nodules were found in the left hand too (Figure 1).

Because the tumors on the patient's tongue interfered with swallowing and mastication, he was operated on to excise the tumors in the summer of 2006. He is now free of the tumor after one year.

The tongue’s lesions were two pieces of purple-brown, oval, and elastic soft tissue with polyloid surface measuring 1.5×2.5×2cm, which enlarged recently (Figure 2).

Radiographic findings of the patient’s hand revealed phleboliths within the hemangiomas.

Two excisional biopsy samples were taken...
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from the tongue’s hemangioma and hand’s enchondroma. The evaluation of the tongue's biopsy sample indicated a benign vascular lesion composed of tortuous and dilated spaces with flattened endothelial lining. These spaces were filled by red blood cells. No evidence of malignancy was found.

Microscopic evaluation of the hand's enchondroma showed mature lobules of hyaline cartilage in addition to foci of myxoid degeneration, calcification, and enchondroma ossification, which were the characteristic features of the disease.

Discussion

Multiple enchondromas (dyschondroplasia, Ollier’s disease) with co-existing soft tissue hemangiomas is termed as Maffucci’s syndrome. Maffucci first reported this condition in 1881. Carletone et al. proposed the eponym "Maffucci's syndrome" in 1942.

No familial, pattern, sexual, or racial predilection is detected in this syndrome.

Malignancies other than musculoskeletal ones have also been reported in Maffucci's syndrome. The overall prevalence of malignancies associated with Maffucci's syndrome is 23% to 100% in different studies.

Although malignant changes are almost common in this syndrome, there was no evidence of malignant changes in our patient at the time of diagnosis and after a six-month follow-up.

Management of Maffucci’s syndrome aims at the relief of symptoms and early detection of malignancies. Operative procedures for the skeletal lesions, such as corrective osteotomy and lengthening of the arm and leg, have been reported. Sclerotherapy, irradiation, and surgery for vascular lesions have been described. In our case simple excisional surgery on the tongue brought out good results.

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