Journal of Oral and Maxillofacial Surgery, Medicine, and Pathology xxx (2017) xxx–xxx

Contents lists available at ScienceDirect

Journal of Oral and Maxillofacial Surgery, Medicine, and Pathology

journal homepage: www.elsevier.com/locate/jomsmp

Case report

Tongue neurofibroma: An oral manifestation of neurofibromatosis type 1 – case report

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ARTICLE INFO

Article history:
Received 17 January 2017
Received in revised form 27 March 2017
Accepted 25 May 2017
Available online xxx

Keywords:
Neurofibroma
Neurofibromatosis 1
Diagnosis

ABSTRACT

The neurofibroma is a benign tumor of neural origin that is relatively uncommon and even rare in the oral cavity. It is mostly associated with neurofibromatosis type 1, a dominant and rare autosomal disease characterized by the reduced tumor suppressor protein neurofibromin. The aim of this study is to describe a case of oral neurofibroma associated with neurofibromatosis type 1 and to review the relevant literature. A 41-year-old male diagnosed with neurofibromatosis type 1 reported a slow-growing nodule in his tongue, which was asymptomatic. Physical examination showed a sessile nodular lesion covered by intact mucosa located at the back of the tongue. Numerous soft-consistency nodules were observed on various areas of the skin, as well as café-au-lait skin macules. Considering the diagnoses of focal fibrous hyperplasia or benign tumor of mesenchymal origin, an excisional biopsy was performed. Histopathological examination established the diagnosis of neurofibroma. There were no signs of recurrence after 2 years of follow-up.

1. Introduction

Neurofibromas are benign tumors of neural origin that commonly affect the skin; they are rarely found as intraoral lesions. Usually they originate from non-encapsulated peripheral nerves and may be derived from Schwann sheath cells or perineural fibroblasts [1]. The oral neurofibroma can manifest in solitary form, even though, in most of the cases, it is related to neurofibromatosis type 1. Oral involvement can occur in about 72% of the cases [2,3].

Neurofibromatosis (NF) is a complex and rare neurocutaneous disease; it is genetically inherited and can manifest as one of nine types, of which NF type 1 and type 2 are the most common [4,5]. NF type 1 is the classic form of the disease corresponding to about 90% of the cases [4] and requires a multidisciplinary approach for the manifestations not only in the nervous system but also in the skin, eyes, bones and occasionally in the oral cavity [5].

The choice of treatment is surgical excision, and the prognosis is somewhat doubtful because when it is associated with the NF type 1, the oral neurofibroma has a greater risk of malignancy. Such a transformation can be associated with the malignant peripheral nerve sheath tumor (MPNST), also known as neurofibrosarcoma or malignant schwannoma [6,7].

The present study aims to report a case of tongue neurofibroma as a manifestation of NF type 1, describing its clinical characteristics, diagnostic hypothesis, diagnosis process, histopathological characteristics and treatment.

2. Case report

A 41 years old male patient sought dental assistance complaining of a nodule on the tongue without painful symptoms that he first noticed three years ago; he stated that the size of the nodule had not changed since then. The patient also reported history of neurofibromatosis type 1.

At the extra-oral physical exam, many nodules of soft consistency were observed in several skin areas of the upper body and limbs. Café-au-lait skin macules were also observed (Fig. 1). The intra-oral exam showed a sessile nodule covered by intact mucosa with normal color, measuring about 8 mm in diameter and located

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http://dx.doi.org/10.1016/j.ajoms.2017.05.009
2212-5558/

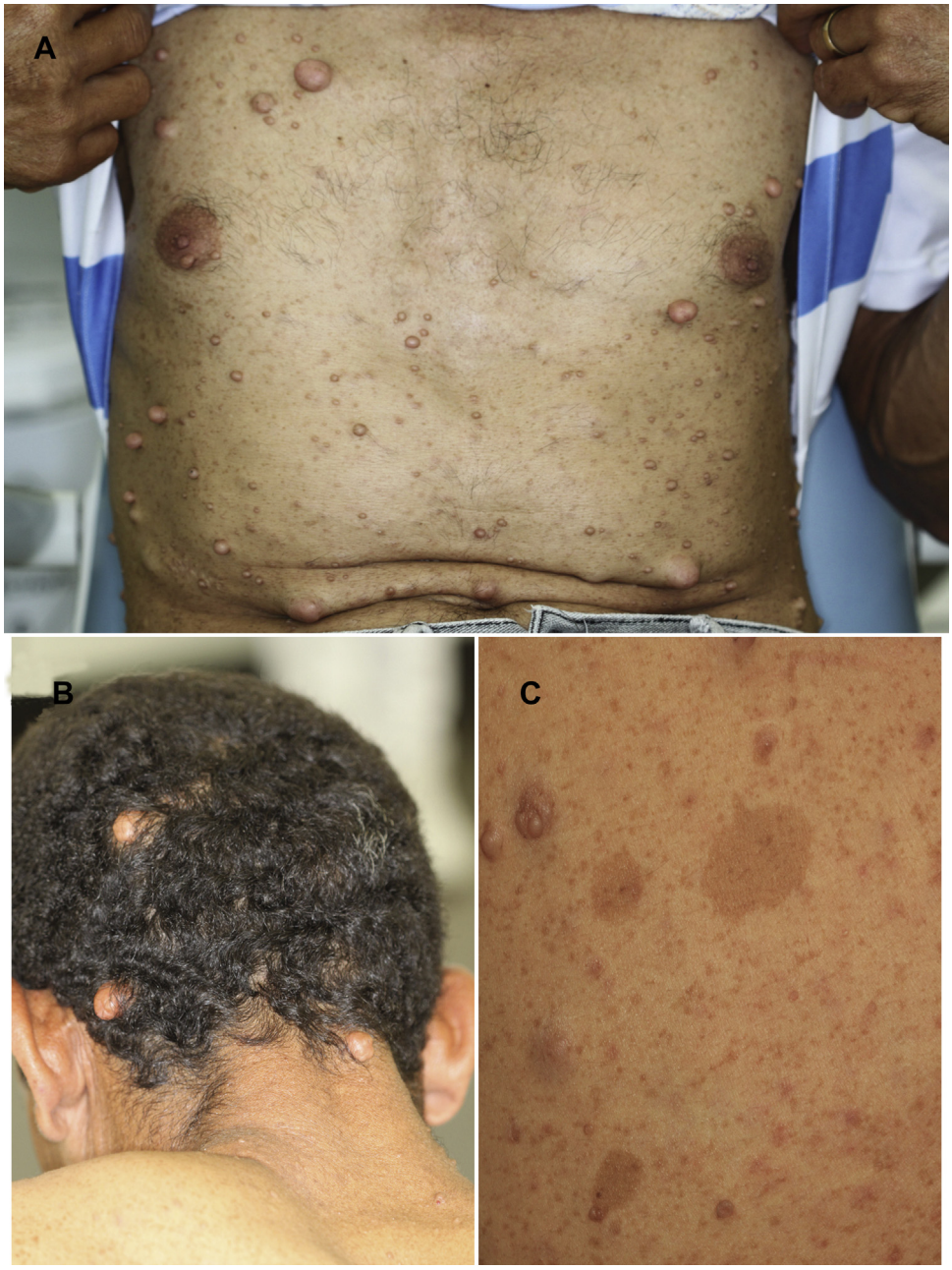

Fig. 1. Extra-oral physical exam. Many nodules of soft consistency were observed in several areas of the skin of the upper body (A) and head (B). Café-au-lait skin macules were found on several parts of the upper body (C).

on the posterior right dorsum of the tongue (Fig. 2). Panoramic radiograph didn't show any significant bone changes.

Based on the clinical characteristics, the diagnostic hypotheses were focal fibrous hyperplasia and benign mesenchymal neoplasia. An excisional biopsy was performed, and the lesion was sent to the Oral Pathology Laboratory at the Pontifícia Universidade Católica de Minas Gerais.

The histological sections stained with hematoxylin and eosin showed oral mucosa fragment covered by parakeratinized stratified squamous epithelium (Fig. 3). On the lamina propria, entangled bundles of fusiform cells exhibiting wavy nuclei associated with delicate collagen bundles were observed (Fig. 4). Mast cells were observed between the fusiform cells (Fig. 4). The conclusive diagnosis was neurofibroma. The patient was sent to the dermatologist to monitor the skin neurofibromas and remains in clinical follow-up. Two years after the surgical treatment, there were no signs of lesion recurrence (Fig. 5) or appearance of new lesions in the oral mucosa.

3. Discussion

Neurofibromas are benign tumors of the peripheral nerves with low occurrence in the population, affecting one in 3000 people [4,8]. They appear as an area of increased volume that may range from a small nodular lesion to a large pedunculated or sessile tumor mass and are characterized by slow growth and a firm consistency. They are usually painless but can cause some pain as a consequence of involved nerve compression. Histological analysis presents fusiform cells with wavy nuclei associated with delicate collagen bundles, in a variable myxoid stroma. The presence of many mastocytes is also common and can aid the diagnosis [9]. Neurofibromas can be found alone but are frequently associated

Please cite this article in press as: Almeida JM, et al. Tongue neurofibroma: An oral manifestation of neurofibromatosis type 1 – case report. J Oral Maxillofac Surg Med Pathol (2017), http://dx.doi.org/10.1016/j.ajoms.2017.05.009