Head & Face Medicine

Review

Solitary submucous neurofibroma of the mandible: review of the literature and report of a rare case

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Abstract

Solitary neurofibroma is a rare benign non-odontogenic tumor. Particularly in the oral cavity, neurogenic tumors are rare, especially if they are malignant. Neurofibromas may present either as solitary lesions or as part of the generalised syndrome of neurofibromatosis or von Recklinghausen's disease of the skin. Clinically, oral neurofibromas usually appear as pediculated or sessile nodules, with slow growth and mostly without pain. The diagnosis can be confirmed by histological examination. Neurofibromas are immunopositive for the S-100 protein, indicating its neural origin. Treatment is surgical and the prognosis is excellent. For illustration a rare case of a solitary neurofibroma in the mandible is presented.

Introduction

Neurogenic tumors are rare in the oral cavity, particularly so when malignant. Traumatic neuroma, although usually included with neurogenic tumors, is a reactive process rather than a true neoplasm [1]. In contrast, neurofibroma and schwannoma derive from nerve fibers, the perineurium, the endoneurium and the neurolemmomal cells [1]. Neurofibromas may present either as solitary lesions or as part of the generalised syndrome of neurofibromatosis or von Recklinghausen's disease of the skin [4-8]. Since the first description of solitary neurofibroma (neurilemmoma, schwannoma) of the oral cavity in 1954 by Bruce only few cases have been reported in the literature [2].

Epidemiology

Although neurofibroma represents one of the most common neurogenic tumors it is an uncommon intraoral tumor [3] like some other intraoral tumours [4]. Neurofibromas can be multiple or solitary. The tumor's most frequent location is the skin and its multiple appearance is highly associated with von Recklinghausen's disease and poliglandular syndrome MEN III [5-9]. It mainly appears in the third decade of life although occurrence between 10 months and 70 years old has been described. Any preference of sex is reported contradictorily [6]. There are no correlations reported with immunocompromising diseases [10].

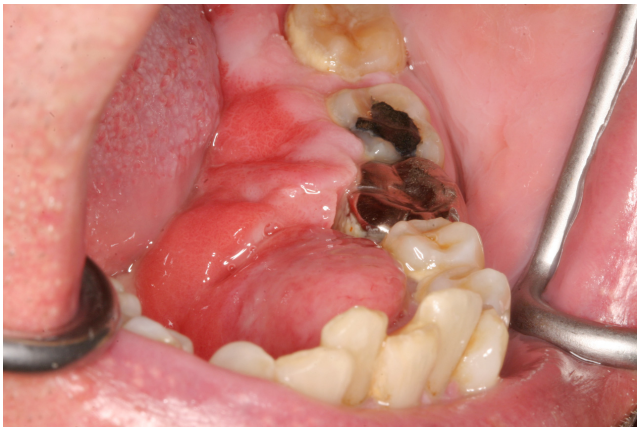

Figure 1
Preoperative view: an exophytic tumor extending all along the lingual aspect of the left mandible.

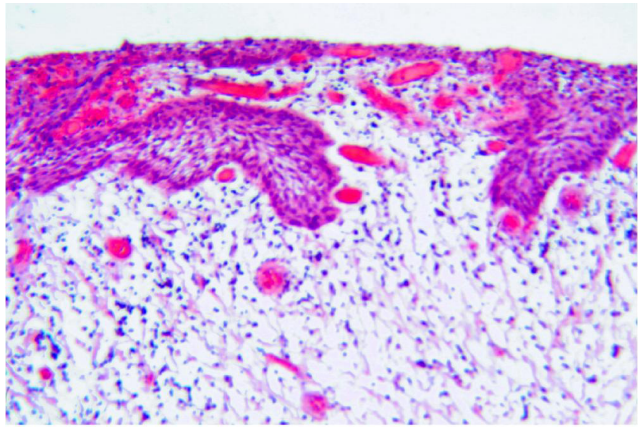

Figure 3
Histological findings showing a tumor of proliferative spindle cells with a stroma composed of irregular collagen fibers (HE, × 100).

Clinical features

Clinically, oral neurofibromas usually appear as pediculated or sessile nodule, with slow growth. They are usually painless, but pain or paresthesia may occur due to nervous compression. The most frequent location is the tongue, although they may occur at any site, especially on the palate, cheek mucosa and floor of the mouth [1,11-14]. Even intraosseous location of the mandible has been described [15-18]. The definitive diagnosis is due to histological examination.

Pathohistological features

The macroscopic appearance of the oral neurofibroma is characterized by a whitish consistent mass with shiny surface. Microscopically the tumor is composed of an irregular pattern of proliferative spindle cells. The stroma is composed of collagen fibers and mucoid masses. Small axons all over the tumoral tissue are demonstrated with silver staining. Neurofibromas are immunopositive for the S-100 protein in 85 to 100% of the cases, indicating its neural origin [19-23].

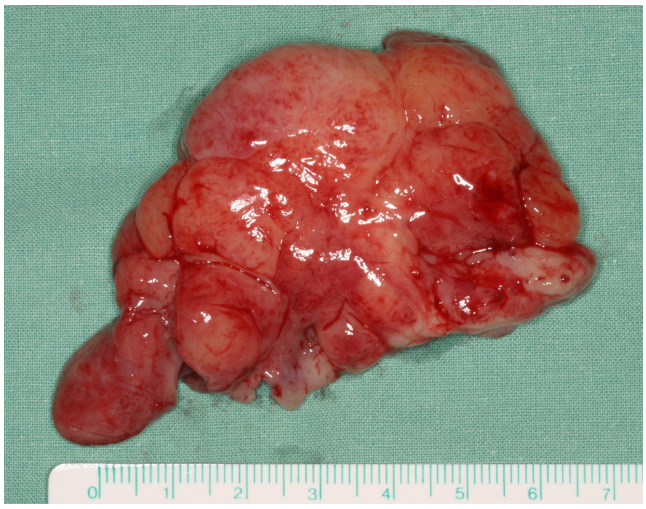

Figure 2
Tumor mass after resection.

Treatment and prognosis

Treatment of choice is surgical excision of the solitary lesions, trying to conserve the nerve from which the tumor originates [5]. Malignant transformation of solitary neurofibroma is extremely rare. Recurrence is also rare although some authors suggest higher rate of recurrence at head and neck location of solitary neurofibromas [24-28]. Therefore, the prognosis is quite excellent.

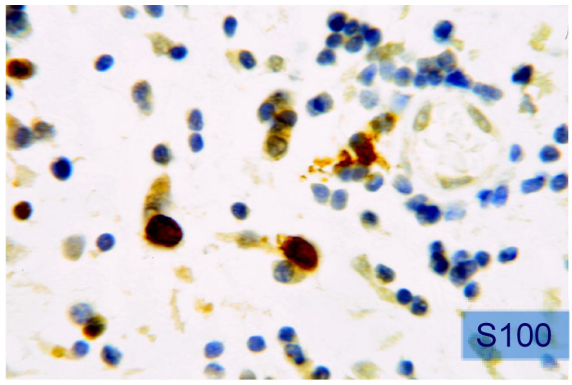

Figure 4
Immunopositive staining for the S-100 protein (×320).

Case report

A 64-year-old male patient with a history of somewhat alcohol but no nicotine or any other diseases attended the department for Cranio- and Maxillofacial Surgery. Clinical examination revealed an exophytic tumor in the oral cavity extending all along the lingual aspect of the left mandible (fig. 1). Panoramic radiographs showed little to moderate interdental loss of bone between teeth 37 and 38 but no other abnormalities. Several biopsies from the oral cavity revealed a submucous benign mesenchymale proliferation with no signs of malignancy and thus, the tumor was completely excised under general anaesthesia (fig. 2, fig. 3). Surgical treatment also included extraction of teeth 37 and 38 and a modelling osteotomie. Immunohistochemical findings showed a solitary submucous neurofibroma with a predominate fibromatous component (fig. 4).

In the presenting case the lesion occured at the lingual site of the left mandible presenting as painless pediculated exophytic tumor with slow growth. The localisation supports the lingual nerve as origin for the neurofibroma. The patient presented no signs of von Recklinghausen disease or poliglandular syndrome. Preoperative panoramic radiographs showed no serious abnormalities. Definitive diagnosis was based upon histological and inmunohistochemical findings. A clinical follow-up has been regularly performed for one year, there was no evidence of recurrence until now.

Competing interests

All authors disclaim any financial or non-financial interests or commercial associations that might pose or create a conflict of interest with information presented in this manuscript.

Authors' contributions

DS, JH, RD and NK made substantial contribution to the conception and design of the manuscript. PR carried out the pathohistological investigations and participated in creating this part of the manuscript.

All authors were involved in revising the manuscript critically and have given final approval of the version to be published.

Consent statement

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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