

原文題目(出處)：	A rare malignant peripheral nerve sheath tumor of the maxilla mimicking a periapical lesion. Case Rep Dent Volume 2016, Article ID 4101423
原文作者姓名：	Arruda JA, Álvares P, Silva L, dos Santos Neto AP, Rodrigues CD, Caubi A, Silveira M, Sayão S, Sobral A P
通訊作者學校：	Universidade de Pernambuco, Brazil
報告者姓名(組別)：	溫維爾 Intern F 組
報告日期：	2017/1/10

內文：

Introduction

Malignant peripheral nerve sheath tumor (MPNST) is a malignant neoplasm that is rarely found in the oral cavity. About 50% of this tumor occurs in patients with neurofibromatosis type 1 (NF-1) and comprises approximately 10% of all soft tissue sarcomas of both head and neck region. Its incidence in the oral region is extremely low, roughly 0.001%. Its occurrence in head and neck regions involves soft and hard tissues. However, intraosseous maxillary MPNST presenting as a periapical lesion is exceptionally unusual. This tumor type occurs in peripheral nerves and may exhibit differentiation in nerve sheath elements such as Schwann cells and perineural cells, as well as fibroblasts. The differential diagnosis of this neoplasm may be facilitated by additional information obtained from immunohistochemistry findings

This article reports the unusual clinical case of intraosseous MPNST with no association with NF-1, presenting as a periapical lesion on nonvital endodontically treated teeth

Case Presentation

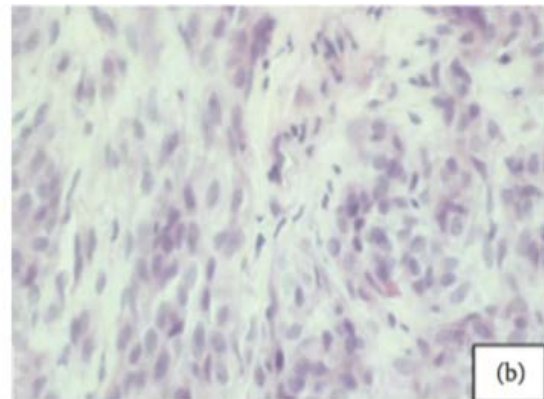
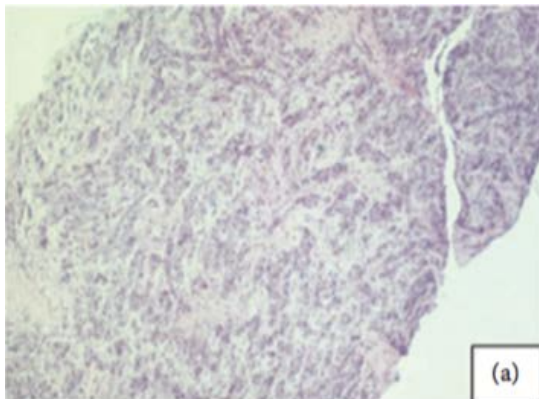
Age: 16	Sex: Female	Location: Upper left incisors region
Chief Complaints: Referred with pain involving the upper left incisors region.		
Present Illness: She accepted Endodontic treatment of the maxillary upper left incisors with necrotic pulp and periapical radiolucency 6 months earlier. A few months after the treatment, she experienced discomfort pain		
Extraoral Examination: An integrate mucosa with normal appearance		
Intraoral Examination: The upper left lateral incisor presented chromatic alterations and mobility.		

Periapical film findings:

A defined radiolucency lesion between the upper left incisors of the endodontically treated teeth with a size of 2.5 × 1.0 cm in a high diameter

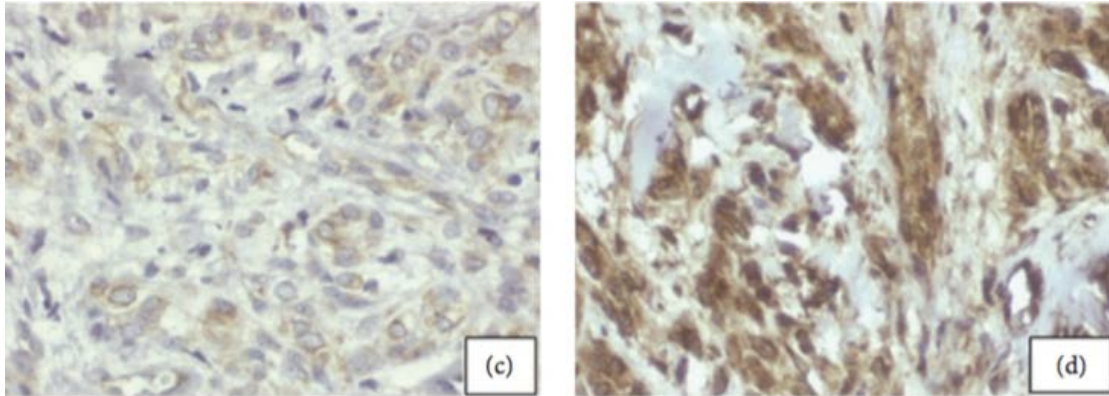
**Incisional Biopsy:**

- Under local anesthesia
- Sent to the Oral Pathology Laboratory
- Fixation: 10% neutral formalin
- Routinely prepared for light microscopy
- Stained with Hematoxylin and Eosin
- Histology Examination:
Malignant neoplasm fragments consisting of fusiform cells with comma-shaped nuclei



- Immunohistochemistry analysis:
 - using streptavidin-biotin technique with monoclonal antibodies

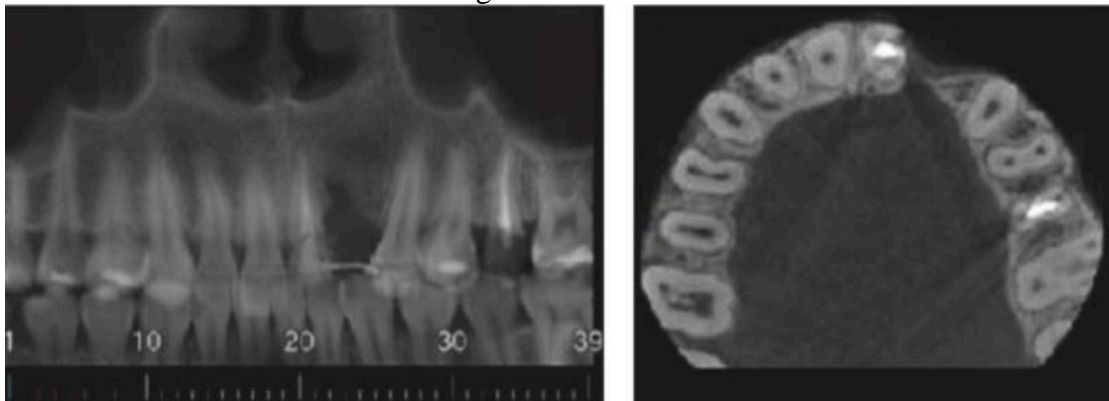
- positive for both the S-100 protein and the glial fibrillary acidic protein (GFAP)

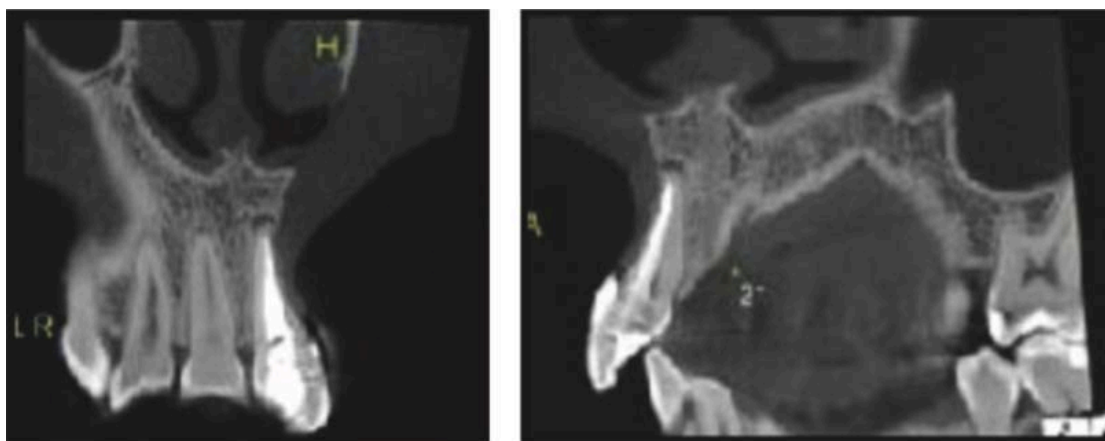


- The immunohistochemical expression of protein S-100 was weak at less than 50% of the tumor cells, and a diagnosis of MPNST of the maxilla was made

Treatment:

- Referred to the Oncology Department:
Surgical excision of the upper left lateral incisor as well as the total removal of the remaining lesion and adjuvant chemotherapy
- Examination of the surgical part:
Stage as low grade of malignancy
- Research of metastasis (through the exams for images):
Reveal no secondary tumors
- Clinical Oncology team:
Not using Radiotherapy
- Adhesive prosthesis in the region of the removal tooth
- Last exam:
CBCT revealed the absence of the upper left lateral incisor, a loss of bone matter, and the absence of vestibular and lingual cortices in the inferior and central thirds of the alveolar ridge





The patient was tumor-free at the nine-year follow-up consultation

Discussion

- An unusual case of intraosseous maxillary MPNST mimicking a periapical lesion -> Several different diseases are able to mimic endodontic periapical lesions
- The presence of periapical images and dental resorption are characteristic of endodontic and/or odontogenic origin lesions. So tissue tumor diagnoses are extremely difficult, and diagnosis of MPNST is particularly difficult because it lacks diagnostic criteria
- Malignant peripheral nerve sheath tumors were most commonly located on the extremities (45%), then trunk (34%), and head and neck region (19%). In head and neck region, they occur most frequently in both nasopharynx and the nasal cavity
- Radiographic findings of intraosseous MPNST may show a great variation, from unilocular to multilocular, with or without well-defined borders of the lesion, and cortical expansion
- Despite imaging features of MPNST being nonspecific and noncharacteristic, the role of radiologic diagnosis is to ensure that MPNST are preoperatively suggested as a diagnosis and are differentiated from malignant lesions through a series of specific imaging findings
- Although no consensus exists regarding the classification of sarcomas as a type of MPNST, the reported lesion could be placed in this category because its onset occurred in a peripheral nerve or neurofibroma, and it fulfilled one of the three established diagnosis criteria, which include
 - (1) The tumor originating in a peripheral nerve that does not exhibit lines of aberrant or heterologous differentiation
 - (2) The tumor originating from a preexisting benign nerve sheath tumor, usually a neurofibroma
 - (3) The tumor exhibiting characteristics that are commonly observed in tumors originating from the aforementioned situations or malignant Schwann cell tumors

The present case fulfills the diagnostic criterion of MPNST onset occurring in a peripheral nerve

- Establishing a diagnosis requires immunohistochemistry technique because MPNST may be similar to fibrosarcoma. And a plethora of other bone tumors are included in differential diagnoses such as central giant cell lesion, fibrous dysplasia, ameloblastoma, myxoma, haemangioma, neurofibroma, leiomyoma, sarcoma, or simply periapical lesions

- The recommended treatment option for MPNST is surgical excision. However, it is a difficult procedure due to poor anatomical accessibility and its probability of recurrence varies approximately between 40 and 65% in head and neck region and approximately between 40 and 68% elsewhere. Surgical approaches to malignant soft tissue tumor varies based on the extent of the disease, age of the patient, and pathological findings. Yet, surgical excision with wide margins is the first treatment choice for the majority of patients
- Radiation and/or chemotherapy may also be used due to the high recurrence rates or in the case of an incomplete lesion excision. Adjuvant radiation and/or chemotherapy treatment must always be considered and guided by the clinical stage

題號	題目
1	Malignant peripheral nerve sheath tumor is most common on/in: (A) Oral Cavity (B) Neck (C) Trunk (D) Extremities
答案(D)	出處：Oral and Maxillofacial Pathology, Nevielle, Saunders W. B. Co. 3 rd edition. Chapter 12: Soft tissue tumor, p.555
題號	題目
2	The treatment of malignant peripheral nerve sheath tumors: (A) radical surgical excision, possibly along with adjuvant radiation therapy and chemotherapy. (B) radiation therapy only (C) Chemotherapy only (D) No need for treatment
答案(A)	出處：Oral and Maxillofacial Pathology, Nevielle, Saunders W. B. Co. 3 rd edition. Chapter 12: Soft tissue tumor, p.555