Case report

Intraosseous Solitary Neurofibroma in Ramus of Mandible: A Unique Clinical Case Report

Abstract

Neurofibroma is a rare benign non -odontogenic tumor which may occur predominantly as a feature of Von Recklinghausen's disease affecting the soft tissue. Intraorally, the intraosseous solitary variant of Neurofibroma is a very rare entity thereby intriguing the oral physicians. We report a rare case of solitary Neurofibroma located at the level of Mandibular foramen on the left side without a family history of Von Recklinghausen's disease in a 39- year-old female. The diagnosis was made based on clinical and radiological findings and histopathological report . On Serological investigation the patient was HIV positive. The present case is rare in regard to its location and the immunodeficiency condition of the patient

Introduction:

Although the head and neck is the most common region for peripheral nerve sheath tumors, central involvement particularly in the jaw bones is quite unusual [1]. Intra oral Neurofibroma is common in association with neurofibromatosis (Von Recklinghausen's disease), whereas solitary intraosseous neurofibroma is a rare entity since the bones do not contain myelinated nerves or nerve sheaths within their medullary spaces [2]. Among the benign nerve sheath tumors only about 45% involve the craniofacial region and in that approximately 9% occur in the oral cavity[3] and also, in the craniofacial region the most common occurrence of neurofibroma are, tongue, palate, mandibular ridge/vestibule, maxillary ridge/vestibule, buccal mucosa, lip, mandibular intrabony and gingiva[4]. The term 'solitary neurofibroma of oral cavity' was first coined by Bruce in 1954 [2]. Literature search of intraosseous neurofibroma gave a result of approximately 50 reported cases [5] including the present case, hence it can be said that occurrence of mandibular intraosseous neurofibroma is extremely rare. In this paper, we present an unusual case of intraosseous neurofibroma affecting the right side of mandible at the mandibular foramen in a 39 year old female patient.

Case report:

A 39-year-old apparently healthy female reported to us with a chief complaint of pain and swelling in the right lower one third of the face since one year. The patient experienced tingling sensation of the right half of lower lip continuously during the entire above mentioned period and was not preceded by any trauma or toothache. The swelling was initially small and gradually attained its present size. The patient gave history of pain as dull aching and intermittent in nature. Family history of the patient was non-contributory. General physical examination did not reveal any cafe au lait pigmentation, axial freckling or cutaneous nodules. On clinical examination, visually no gross asymmetry was found extraorally (fig.1a) or intraorally(fig.1b) .

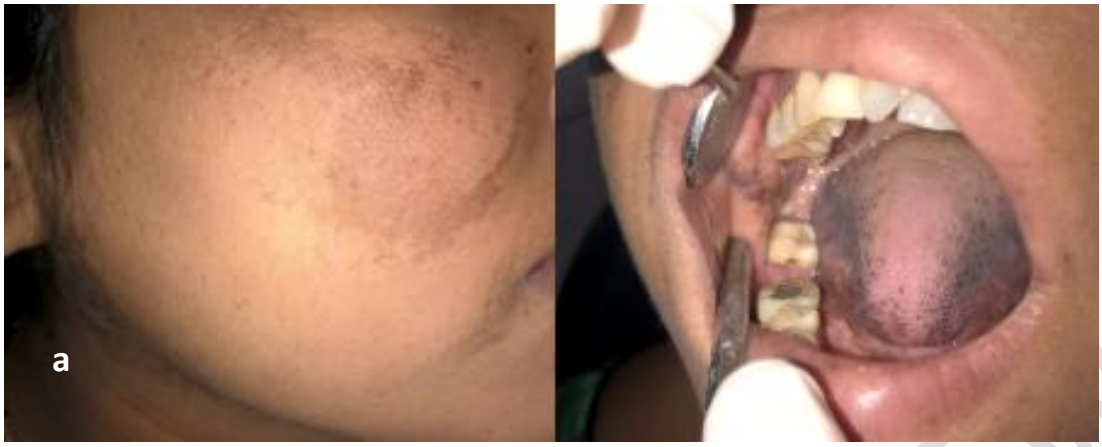

Fig. 1: Extraoral(a) and Intraoral(b) clinical photograph showing no gross asymmetry

The skin over the swelling was apparently normal without any scars, sinus or any drainage. Local temperature of the skin was normal. On palpation, extraorally, a bony hard well defined non tender swelling measuring about 2x3cms was present about 2cms anterior to the pinna of the right ear. Intraorally, swelling was palpated in the right ramus and was non tender, however, the posterior border of the swelling was inaccessible for palpation. There was no lymph node involvement in relation with the lesion.

The patient was advised for all routine hematological and radiological investigations along with HIV screening. The patient's routine haemogram was found to be within normal limits and the patient tested positive for HIV.

A panoramic radiograph, 3DCT and MRI were advised. Panoramic radiograph revealed a well circumscribed homogenously radiolucent area in the ramus of the right mandible (Fig. 2). The radiolucent area was in relation to the mandibular foramen and in continuation with the inferior alveolar nerve canal. The radiolucent area gave an appearance of a cyst within a cyst without any internal calcifications or septae.

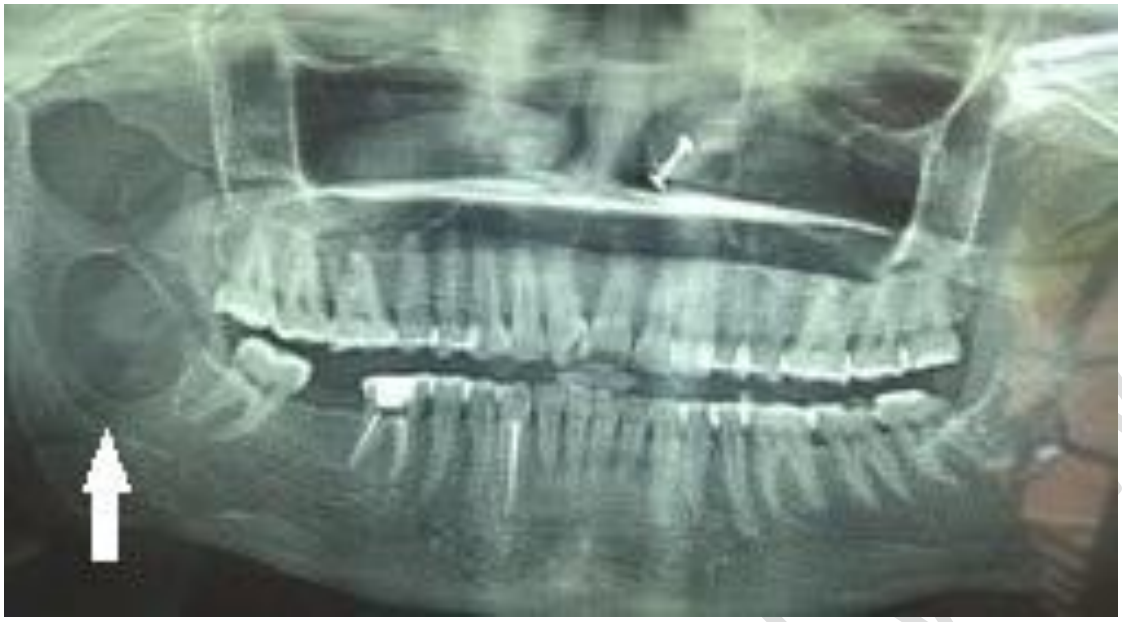

Fig. 2: Orthopantamograph revealing a well circumscribed radiolucent area in the ramus of the right mandible

3D CT scan of mandible (fig.3) revealed a well-defined expansile lytic lesion in the right mandibular ramus measuring about 2.6cmx 2.0cm x 2.6 cm with significant thinning of both medial and lateral cortices of the right mandibular ramus. Soft tissue involvement of masseter and pterygoid muscles attachment was observed.

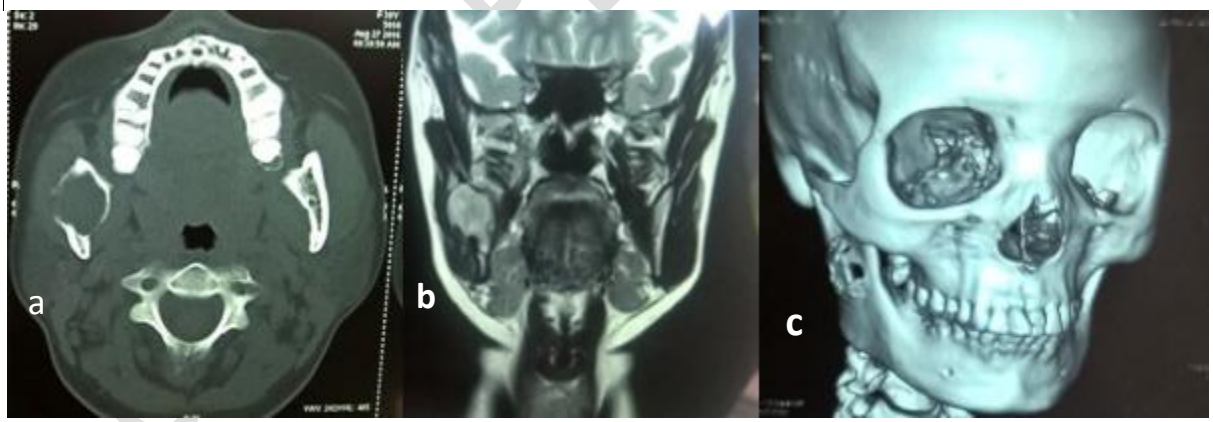

Fig. 3: 3DCT view (3a & 3c) slice of CT scan of mandible showing a well- defined expansile lytic lesion. MRI neck (3b) showing a well-defined T2 hyperintense bone destructive lesion involving the right ramus of the mandible

MRI neck (fig.4) revealed a well-defined T2 hyperintense bone destructive lesion involving the right ramus of the mandible measuring 3.1 x 2.2 x 2.9 cms, medially the lesion abutted medial pterygoid muscle with intact fat planes and laterally the lesion abutted the inner fiber of masseter

muscle with loss of fat planes. Mild widening of right mental foramina with T2 hyperintensity is seen along the lumen in continuous with mass lesion.

After an informed consent, an incisional biopsy was performed and specimen was sent for histopathological evaluation. Hematoxylin and eosin section (fig. 4) revealed a non encapsulated lesion consisting of elongated spindle cells with wavy hypochromatic nuclei dispersed in a collagenous stroma. The tumor cells are set in a fibromyxoid background interspersed with thin wirelike collagen fibres. Few areas showed whorl or fascicular arrangement of cells. Endothelial lined blood vessels along with few mast cells and diffuse chronic inflammatory infiltration was noted. A diagnosis of Solitary Neurofibroma was given.

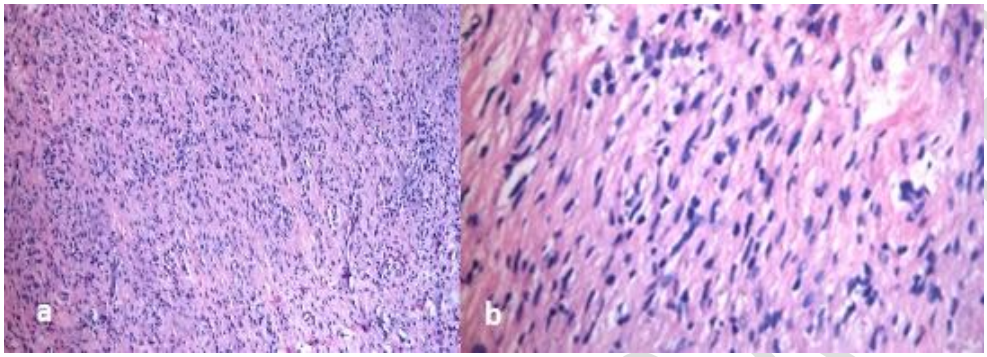

Fig. 4: Photomicrograph shows Hematoxylin and eosin sections with dense cellular lesion showing spindle cells with wavy nuclei and pale eosinophillic cytoplasm. (a-10X, b-40X).

A segment resection of the right side of the mandible was performed and the tumor was completely removed through a cervical approach under general anaesthesia (fig. 5).

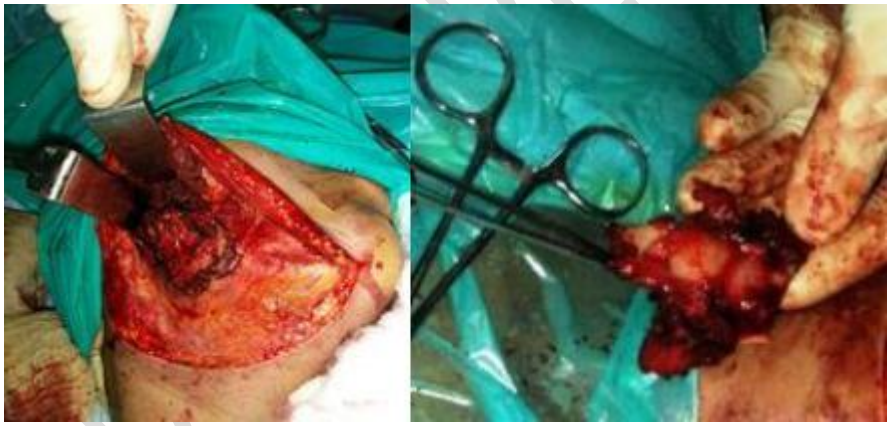

Fig. 5: Intraoperative photograph showing the surgical approach and the excised lesion

The postoperative course was uncomplicated and the patient was discharged. A regular clinical followup is scheduled to check for any indications of any recurrence.

Discussion:

According to Polak [2] and his associates, neurofibroma of the mandible has a predilection for females (2:1) and for the posterior mandible with an average age of occurrence of 27.5 years.

Neurofibroma is an uncommon benign tumor of the oral cavity often involving the trigeminal nerve which arises from the cells of the nerve sheath [6]. The head and neck are commonly involved because of the rich innervations, and also superficial involvement of soft tissues is more frequent than the deeper location [7]. Most of the intraosseous neurofibromas give rise to pain and numbness of lip on the affected side due to the compression of adjacent structures by the increased tumor size. About 90% of the neurofibromas are associated with neurofibromatosis type1, and hence the presence of a solitary lesion requires physical examination and familial history to exclude Von Recklinghausen's disease [8]. The present case presented with a negative family history of neurofibromatosis, a physical examination was done and our patient did not have any clinical features suggestive of Von Recklinghausen's disease. Hence this case is unique due to the sporadic presentation of the lesion without any associated family history. Studies have shown that most intraosseous neurofibroma of the mandible are asymptomatic. So far, few cases of symptomatic intraosseous neurofibroma have been reported [8]. Larrson et al. and Apostolidis et al. reported cases of symptomatic intraosseous neurofibromas of mandible with symptoms like pain, swelling, paresthesia and bone destruction [9]. Similarly, our patient presented with swelling pain and paresthesia of the lower lip on the affected side. Complete surgical excision has been the standard treatment for solitary neurofibroma and recurrence is rare [10].

Conclusion:

Radiolucent lesions in the head and neck region often pose a diagnostic challenge to the oral physians, especially those of tumors of neural origin. Although neurofibromatosis is a common occurrence, intra osseous solitary neurofibroma is a rare finding. Hence a thorough knowledge of the rarities and its varied presentation of a common disorder facilitates early diagnosis and a good prognosis, thereby providing a better quality of life to the affected individual

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