

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) .



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

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

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

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

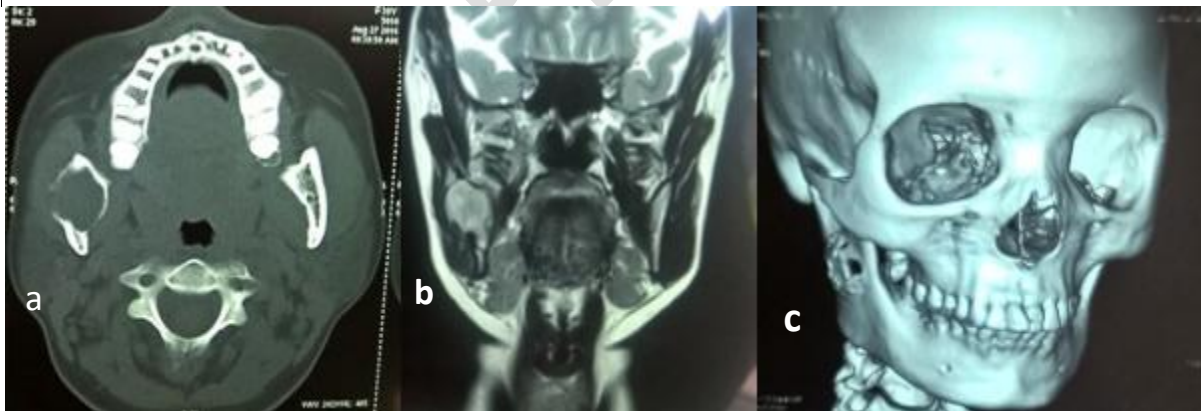


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56 Fig. 2: Orthopantomograph revealing a well circumscribed radiolucent area in the
57 ramus of the right mandible
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59 3D CT scan of mandible (fig.3) revealed a well-defined expansile lytic lesion in the right
60 mandibular ramus measuring about 2.6cmx 2.0cm x 2.6 cm with significant thinning of both
61 medial and lateral cortices of the right mandibular ramus. Soft tissue involvement of masseter
62 and pterygoid muscles attachment was observed.

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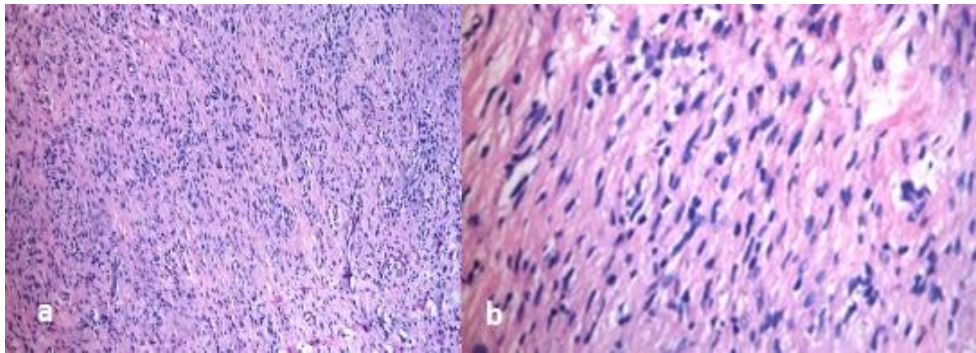
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65 Fig. 3: 3DCT view (3a & 3c) slice of CT scan of mandible showing a well- defined expansile
66 lytic lesion. MRI neck (3b) showing a well-defined T2 hyperintense bone destructive lesion
67 involving the right ramus of the mandible
68

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

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

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



81
82 Fig. 4: Photomicrograph shows Hematoxylin and eosin sections with dense cellular lesion
83 showing spindle cells with wavy nuclei and pale eosinophilic cytoplasm. (a-10X, b-40X).

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



86
87 Fig. 5: Intraoperative photograph showing the surgical approach and the excised lesion

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

90 Discussion:

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

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

110 Conclusion:

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

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117 References :

- 118 1. Che Z, Nam W, Park WS, Kim HJ, Cha IH, Kim HS, et al: Intraosseous nerve sheath
119 tumors in the jaws. *Yonsei Med J.* 2006;47: 264-270.
- 120 2. Polak M, Polak G, Brocheriou C, Vigneul J. Solitary neurofibroma of the mandible: case
121 report and review of the literature. *J Oral Maxillofac Surg* 1989; 47:65-8.
- 122 3. Das Gupta TK, Brasfield RD, Strong EW, Hajdu SI. Benign solitary Schwannomas
123 (neurilemomas) *Cancer.* 1969;24:355–366.
- 124 4. Bisher HA, Kant R, Aldamati A, Badar AA. Plexiform neurofibroma of the
125 submandibular gland in patient with Von Recklinghausens disease. *Rare*
126 *Tumors.* 2011;3(1):e4.
- 127 5. Vivek N, Manikandhan R, James P C, Rajeev R. Solitary intraosseous neurofibroma of
128 mandible. *Indian J Dent Res* 2006;17:135
- 129 6. Borges AH, Correia RD, Borba AM, Guedes OA, Estrela CD, Bandeca MC. Unusual
130 solitary neurofibroma on the lower lip of a child. *Contemp Clin Dent* 2013;4:512-4
- 131 7. Gujjar PK, Hallur JM, Patil ST, et al. The Solitary Variant of Mandibular Intraosseous
132 Neurofibroma: Report of a Rare Entity. *Case Rep Dent.* 2015;2015:520261.
- 133 8. Deichler J, Martínez R, Niklander S, Seguel H, Marshall M, Esguep A. Solitary
134 intraosseous neurofibroma of the mandible. Apropos of a case. *Med Oral Patol Oral Cir*
135 *Bucal.* 2011;16:e704–7..

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9. A. Larsson, F. Praetorius, and E. Hjörting-Hansen, intraosseous neurofibroma of the jaws. *International Journal of Oral Surgery*. 1978;7(5):494–499.
10. Neville DW, Damm DD, Allen CM and Bouquot JE. *Oral and maxillofacial pathology*. 3th ed. Philadelphia: W.B. Saunders; 2009: 528-529.

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