Case Report

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Dentistry Section

Intraosseous Neurofibroma of the Mandible: A Case Report and Review of Literature

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ABSTRACT

Neurofibroma (NF) is a benign tumor of the peripheral nervous system. Head and neck NF are generally located in the soft tissue. The tumour is rarely seen intraosseously and most commonly such tumours are seen as solitary lesions, rather than part of neurofibromatosis. The following report describes a case of an intraosseous neurofibroma in a 45-year-old male located in the left posterior mandible. The diagnosis was made based on the clinical findings, radio graphical features, histopathology, and immunohistochemistry. A literature review has been done on intraosseuos neurofibromas located in the jaws, with a discussion on the possible differential diagnosis.

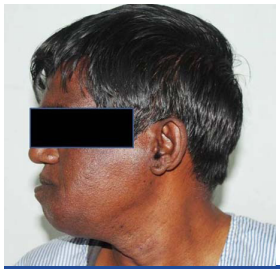
Keywords: Benign tumour, Immunohistochemistry, Nerve sheath, Peripheral nervous system

CASE REPORT

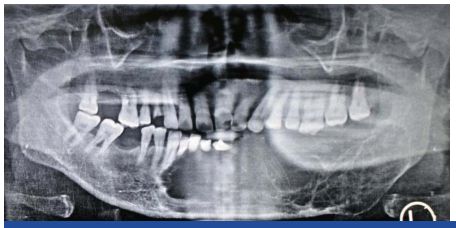
A 45-year-old male patient presented with a swelling in the left posterior region of the mandible extending from tragus of the ear and crossing the midline towards the right side of the face [Table/Fig-1]. The swelling was first observed one to one and half years back which started as a small swelling and has reached to the present size. Extra orally, a diffuse swelling was seen on the left side of the face extending from the angle of the mandible to the mid symphysis. There were no lesions seen on the skin. On intraoral examination, the swelling was seen from the left third molar region to the central incisor of the opposite side crossing the midline. The swelling was irregular in shape, firm in consistency, with irregular borders and a rough texture [Table/Fig-2]. There was no associated pain or tenderness. The patient gave history of exfoliated teeth due to mobility in the same region of the jaw.

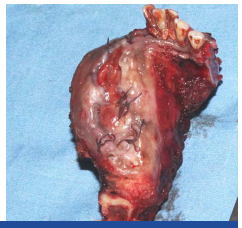
Orthopantomograph (OPG) showed irregular radiolucency in the area of lesion involving the mandibular canal with displacement of the anterior teeth and missing teeth #33-38 [Table/Fig-3].

An incisional biopsy was done and the specimen was sent with a provisional diagnosis of ameloblastoma. The haematoxylin and eosin sections of the incisional biopsy tissue showed hyper cellular connective tissue stroma consisting of spindle shaped cells, with wavy nuclei arranged in fascicular and storiform patterns. Hence, the diagnosis of NF was given which was followed by left hemi-mandibulectomy along with the dissection of supraomohyoid lymph nodes [Table/Fig-4]. Similar histopathological picture was seen in excisional biopsy tissue [Table/Fig-5]. Immunohistochemistry staining with S-100 showed strong positivity signifying the neural origin of the tumour [Table/Fig-6]. Mind bomb E3 Ubiquitin Protein Ligase 1 (MIB-1), which was negative, revealed a low proliferative index of the tumor thus, excluding its malignant nature.

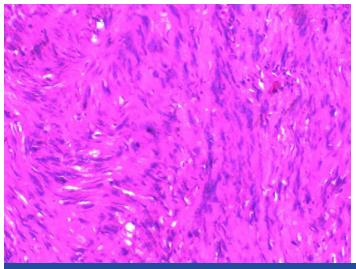


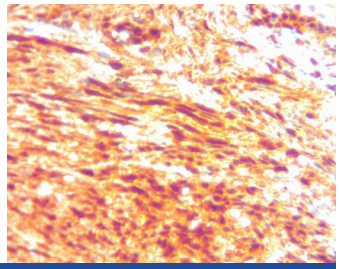
[Table/Fig-1]: Photomicrograph showing diffuse swelling on the left side of the mandible. **[Table/Fig-2]:** Intraoral view of the tumour.





[Table/Fig-3]: Panoramic radiography showing a diffuse radiolucency on the left side of the mandible along with displaced teeth. **[Table/Fig-4]:** Resected tumour mass.





[Table/Fig-5]: Photomicrograph showing spindle shaped cells with wavy nuclei (H&E x20). **[Table/Fig-6]:** Photomicrograph showing S-100 positive spindle shaped cells with wavy nuclei (x10).

DISCUSSION

NF is a benign tumor of the peripheral nervous system. In most of cases, they are found as solitary tumors, though in few cases multiple lesions are seen in persons with neurofibromatosis, an autosomal dominant genetically inherited disease [1]. Bruce described solitary NF of the oral cavity in 1954. NFs are most commonly seen on the skin and only 6% of the cases have been reported in the oral cavity [2]. NFs can also occur intraosseously, the major location being the posterior region of mandible. A few cases have been reported in the maxilla as well. Literature search reveals very few cases of NF of oral cavity. Approximately 25% of the NFs are seen in the head and neck region, and 5.6% of them occur in the oral cavity. Among the craniofacial region, NF often involve 5th cranial nerve and upper cervical nerves, with common sites being, tongue, lip, palate, gingiva, major salivary glands and jaw bones [2]. A literature review has been done of reported cases of intraosseous NFs based on clinical examination, radiographic findings, histological features and immunohistochemical reactivity [Table/Fig-7].

NF is caused by a germline mutation in the NF1 gene, a tumor suppressor gene located at 17q11.2 chromosome. This gene encodes for a protein known as neurofibromin, which plays a role in neural cell signaling.

The mean age of occurrence is 27.5 years with a slight predilection towards females [2,16].

Author	Age	Gender	Location	Symptomatology	Radiographic Features	Histological Features	Immunohistochemistry Features
Larsson et al., [3]	46 Yr	M	Mandible	Intermittent pain	Bone destruction with slightly radio opaque areas	Spindle cells with elongate or oval nuclei forming cords	Not perfomed
Larsson et al., [3]	25 Yr	F	Mandible	No	Extense bone resorption	Irregular nerve fiber strands intermigled with collagen fibers and abundant cells	Not perfomed
Skouteris et al., [4]	16 Yr	F	Maxilla	No	Poorly-defined radiolucent lesion	Spindle cells and abundant myxomatous stroma	Not perfomed
Polak et al., [5]	60 Yr	M	Mandible	No	Unilocular radiolucency	Cords of fusiform or ovoid cells intermixed with a fibrillary stroma	Anti S-100 Positive Anti- Lai 7 Positive
Mori et al., [6]	18 Yr	F	Maxilla	Tooth mobility	Well circumscribed multilocular radiolucency	Wavy growth of tumor cells in a myxomatous matrix	Anti S-100 Positive
Poupard et al., [7]	14 Yr	M	Maxilla	No	Poorly defined radiolucency	Spindle and stellate cells with a mucoid extracellular material with some condensation of fibrous tissue	Anti S-100 Positive
Apostolidis et al., [8]	67 Yr	F	Mandible	Paresthesia and hyperesthesia	Circumscribed elliptical radiolucency with expansion of the mandibular canal	Numerous spindle cells in a myxoid matrix	Not perfomed
Vivek et al., [9]	39 Yr	F	Mandible	No	Well circumscribed radiolucent area with continuity loss of the mandibular canal	Spindle cells with wavy nuclei arranged in the form of booklets	Anti S-100 Positive
Narwal A et al., [10]	5 months	F	Maxilla	No	Radiolucent mass enclosed by radiopaque border	Densely arranged collagen fibers, plum fibroblasts, nerve bundles, wavy nuclei	S-100 positive, EMA negative
Depprich R et al., [11]	64 Yr	M	Mandible	No	No	Proliferative spindle cells in myxoid stroma	S-100 Positive
Sharma et al., [12]	5 months	F	Maxilla	No	No	Cords of dense collagen fibers intermixed with strands of nerve tissue with wavy nuclei	Anti S-100 Positive EMA Negative
Dalili Z and Adham GH [13]	16 Yr	M	Maxilla	Hypoglobus and proptosis of left eye	Oval radiolucency, destruction of overlying bone	Spindle and ovoid cells, wavy nuclei in fairly lose matrix	S-100 Positive
Bharat TS et al., [14]	30 Yr	F	Palate	Mild intermittent dull aching pain	No	Spindle shaped cells, wavy nuclei in Myxomatous areas	S-100, NSE, Vimentin Positive
Jain D et al., [15]	65 Yr	F	Maxilla	Exfoliation of teeth, dysphagia	Slight haziness in maxillary sinus	Myxoid stroma, spindle cells, few areas of plexiform pattern	Not performed
Jangam SS et al., [1]	62 Yr	F	Mandible	Occasional sharp pain, parasthesia, of lower lip	Well defined radiolucency	Admixture of nerve fibers and fibrous tissue	S-100 Positive
Gujjar PK et al., [2]	28 Yr	F	Mandible	Intermittent pain	Homogeneous radio opacity surrounded by thin radiolucent border	Proliferation of spindle cell with wavy nuclei with in myxoid stroma	S-100 Positive

[Table/Fig-7]: A review on clinical, histopathological and immunohistochemical features of intraosseous neurofibroma of the oral cavity.

Most of the intra-osseous NFs do not show any symptoms in the initial stages. Later on pain and numbness of the affected side of the lip may occur [2]. On radiograph, it appears as a well circumscribed or poorly demarcated radiolucency which usually involves the mandibular canal as seen in our case [8].

As a rule, the gross appearance of the tumours is not encapsulated and has a softer consistency [8] though the presentation may vary from case to case.

Microscopically, these tumours are composed of all the elements of peripheral nerve. The Schwann cells usually represent the predominant cellular element which is seen as small round to spindle shaped cells with wavy nuclei showing a serpentine configuration and pointed ends. The stroma is composed of a rich network of collagen fibers which are arranged in a characteristic "shredded carrot pattern". Few NFs, especially the cellular one's show fascicular or storiform growth pattern of cells [8], myxoid areas and scattered mast cells are found in the connective tissue.

NFs may exhibit cells with large hyperchromatic nuclei. These NFs with atypia may also have increased cellularity but mitotic activity is scanty or absent and the MIB-1 index is very low. The immunoreactivity for MIB-1 was negative in the present case.

On immunohistochemistry, NFs show a strong positivity for S-100 protein, variable numbers of EMA positive perineural cells and CD34 positive fibroblasts may also be seen. Immunoreactivity for PGP 9.5 is often seen but its degree of specificity is very low [10].

NFs should be differentiated from other spindle cell lesions like schwannoma, traumatic neuroma, desmoplastic melanotic melanoma, benign fibrous histiocytoma, spindle carcinoma and amelanotic melanoma [2]. Absence of verocay bodies, presence of mast cells, and fine fibrillar collagen matrix differentiates NF from Schwannoma [17]. A positive history of trauma in traumatic neuroma is necessary for its diagnosis. Desmoplastic melanoma demonstrates junctional melanocytic proliferation, neurotropic growth and the presence of lymphoid aggregate in the deep dermis and subcutis. The cells tend to grow together in small packets which is not seen in NF [18]. NFs need to be differentiated from other neural tumors as the malignant transformation rate is around 5% to 16% [8].

Radical surgery which includes hemimandibulectomy or enbloc resection of the mandible is often advocated for the treatment of intraosseous neurofibroma [9]. Local recurrence of NFs has been reported which is attributed to absence of complete capsule making complete surgical removal of the tumor difficult [1,8].

CONCLUSION

It is important to recognize and diagnose these tumors, as they could be the first manifestation of neurofibromatosis. These tumours are known to recur and thus, it is essential to conduct a thorough clinical and radiographical examination along with long term follow up. As the follow up information is not available in the literature search, the transformation into neurofibromatosis cannot be commented on.

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