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Case Study

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SOLITARY INTRA-OSSEOUS NEUROFIBROMA OF MANDIBLE- REPORT OF A RARE CASE

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ABSTRACT

Neurofibroma is a benign tumor of the peripheral nervous system affecting the Schwann cells as well as neural sheath cells. Though it is known to be a component of Von Recklinghausen's disease/ Neurofibromatosis-1(NF-1), solitary lesions have also been reported. Its occurrence in the head and neck region is rare; trigeminal nerve and the cervical nerves being frequently affected. In the oral cavity, soft tissue (tongue) is the most commonly affected site. Intra-osseous neurofibromas are even rarer and have been usually reported in the mandible. We hereby report a case of an intra-osseous neurofibroma in a 26 -year-old female who presented with swelling of the left side of the face since one year.

KEYWORDS: Intra-osseous neurofibroma, neural sheath cells, S-100 protein, schwann cells.

INTRODUCTION

Neurofibromas are benign tumors of the peripheral nervous system arising from Schwann and neural sheath cells.[1] Usually, they are pathognomic feature of von Recklinghausen's disease of the skin/ Neurofibromatosis type1 (NF-1), but can occur in solidarity too.[2] Skin is most commonly affected but intraoral sites have also been reported. Bruce in 1954 reported the very first case of neurofibroma occurring in the oral cavity.[3] Very rarely, intra-osseous neurofibromas have been reported in the jaws, with posterior mandible being the most common site.

CASE REPORT

A 26-year old female, reported to the Department of Oral Medicine and Radiology with a chief concern of swelling in the lower left back region of the jaw since one year. Initially, the swelling was barely discernible, approximately the size of a pea, but, gradually, over a period of time, it attained the size of a tennis ball. From the last few months, the patient had been experiencing a sudden, sharp, intermittent pain in the lower left back tooth. The pain occurred on chewing the first bite of the meal and would radiate to the left jaw and ear. Thereafter, it would resolve on its own, only to reoccur during the next meal. She denied any history of food lodgment, sensitivity to cold/ hot beverages or night pain in the tooth. There was no history of intermittent swelling in the mouth prior to meals. She also denied dryness of the mouth. There was no history of trauma, lip paresthesia or pus discharge from the concerned region. History of fever, similar swelling elsewhere in the body and weight loss was also negative. No relevant dental history could be elicited. The patient was a beautician and hence, was extremely concerned with her appearance. She was married and had a two-year-old baby.

Figure: 1. Diffuse swelling on the left side of the face.

On extra-oral examination, a subtle, diffuse swelling was evident on the left side of the face, involving the mandibular body, left submandibular and pre-auricular

region (Figure 1). The overlying skin was of the same color as the contra-lateral side without any evidence of abnormality (sinus tract /fistula/growth). On palpation, the temperature of the swelling was not raised, it was non-tender, non-compressible and smooth. It was approximately 5x4x3cm in size, extending from chin to angle of mandible anteroposteriorly, preauricular region to left submandibular region superoinferiorly and mediolaterally from submandibular region to the border of the mandible. On intra-oral examination, the patient had good oral hygiene. The entire dentition had a healthy periodontium. Third molars were missing. Mild obliteration of the left buccal vestibule was evident distal to 37 (Figure 2a). Floor of the mouth was not raised and the lingual, as well as buccal cortical plate, did not reveal expansion (Figure 2b).

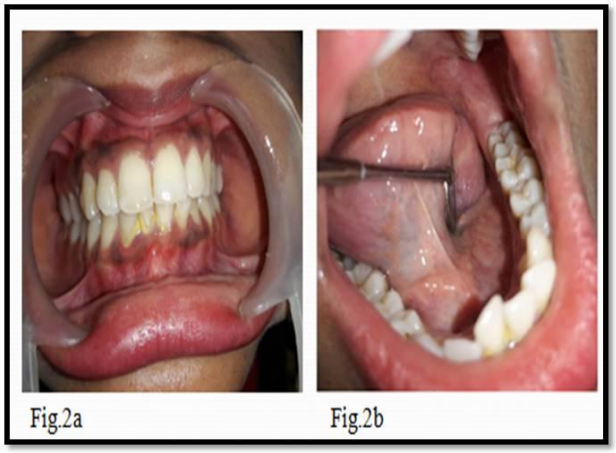

Figure: 2. Intraoral examination.
Figure: 2 a. Mild obliteration of the left buccal vestibule evident distal to 37.
Figure: 2 b. Floor of the mouth is not raised. Lingual cortical plate did not reveal expansion.

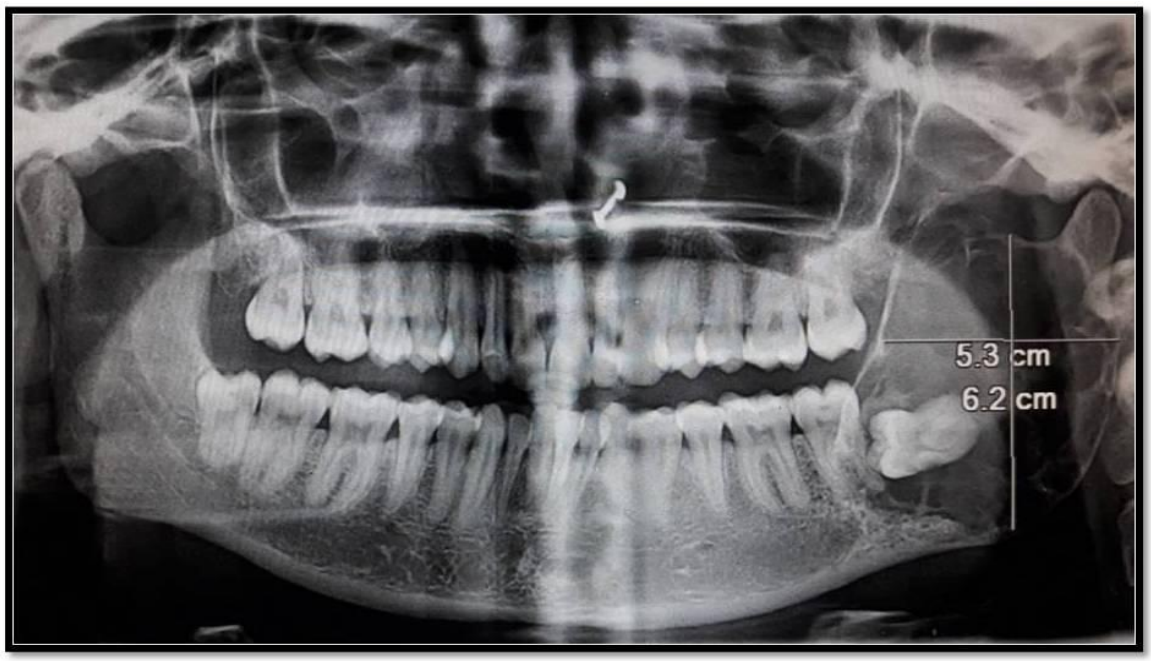

Figure: 3. OPG showing a solitary, well- defined multilocular radiolucent lesion within the left mandible, involving the entire ramus.

A provisional diagnosis of benign odontogenic cyst was made. Orthopantomograph (OPG) (Figure 3) revealed a solitary, well- defined multilocular radiolucent lesion within the left mandible, involving the entire ramus. It was extending from 37 anteriorly to the posterior border of the ramus posteriorly. Supero-inferiorly, it extended from the coronoid to the angle of the mandible. The condyle appeared to be spared. Few thick septa originated at 90^0 to the periphery. Within the radiolucency, horizontally impacted 38 abutted against the distal root of 37. The cortication of the Inferior alveolar nerve (IAN) canal could not be traced. Cone-beam computed tomography (CBCT) revealed extensive mediolateral expansion of the ramus with thinning at multiple sites (Figure 4). Radiographic diagnosis of odontogenic tumor, most likely ameloblastoma was made. Odontogenic myxoma, giant cell lesion of hyperparathyroidism, central haemangioma, dentigerous cyst, odontogenic keratocyst, and neurofibroma was considered in the differential diagnosis.

Aspiration of the lesion was unsuccessful. Biopsy distal to 37 revealed a highly cellular connective tissue with cells arranged in short fascicles showing whorled pattern. Spindle cells with long slender nuclei were present, suggesting a spindle cell lesion. For confirmation, the sample was sent for immunohistochemistry (IHC). The tissue was found to be strongly reactive for S-100 protein, confirming the diagnosis of intra-osseous neurofibroma.

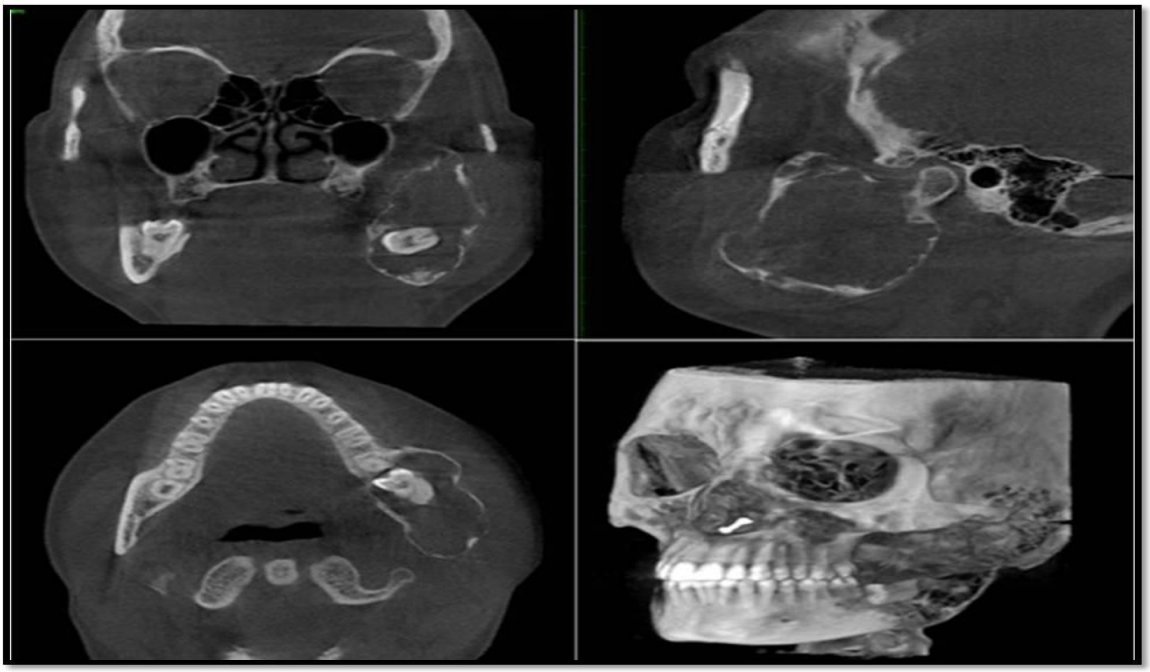

Figure: 4. CBCT images showing the extent of lesion in the coronal, sagittal and axial sections.

Since neurofibromas are known to be a pathognomic feature of NF-1, a thorough careful re-examination was done to rule out café-au-lait macules, Lisch nodules (iris hamartomas) and Crowe's sign (axillary freckles) in the patient. She was then posted for surgical intervention.

Segmental mandibular resection was performed (Figure 5) and the defect was reconstructed with reconstruction plates (Figure 6). The healing was uneventful. Two-year follow-up revealed no evidence of recurrence of the lesion.

Figure: 5. Resected mandible.

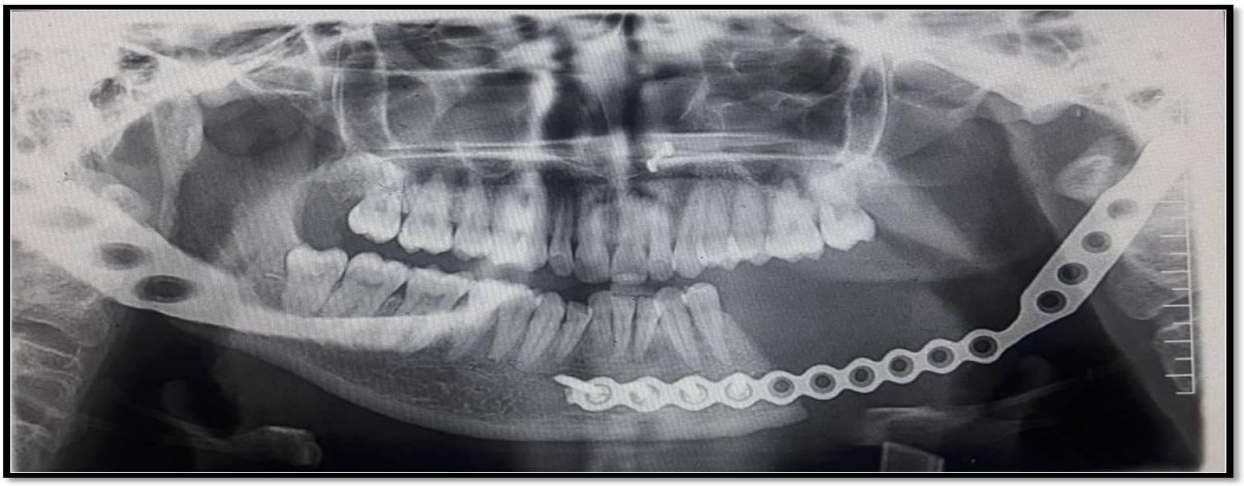

Figure: 6. Post-operative OPG showing reconstruction plate on the left side.

DISCUSSION

Approximately 25% neurofibromas are found in the head and neck region (arising from trigeminal and cervical nerves) owing to the rich innervations of this area.[4] 6% cases have been reported in the oral cavity with the tongue being the most commonly reported site followed by buccal mucosa, and lips.[5] In the jaws, the majority of cases have been reported in the posterior mandible, which can be attributed to the major nerve bundle (IAN) present in this area.[6] Few cases have been reported in the maxilla as well. The average age is 27.5 years with a female to male predilection of 2:1.[7] This predilection is well supported in the present case as the patient was a 26 year old female.

Clinically, the intra-osseous neurofibromas present as a diffuse swelling of variable size. The patient presents with numbness of lip if the inferior alveolar nerve is involved. In the present case, the patient reported with a diffuse swelling without any concomitant paresthesia.

Radiographically, intra-osseous neurofibromas may appear as unilocular or multilocular, ill-defined or well-defined lesions. Usually ill-defined lesions are reported in the maxilla. In the mandible, when involving a major nerve like inferior alveolar nerve (IAN), neurofibromas can cause fusiform enlargement of the nerve canal, also known as the "blunderbuss canal" formation.[8] The present case depicted a multilocular, well-defined lesion in the posterior mandible with the destruction of the entire IAN canal.

Microscopically, neurofibromas can be classified into classical, cellular and myxoid variety. On immunohistochemistry, neurofibromas show strong positivity for S-100 protein. Immunoreactivity for PGP 9.5 has also been seen, but with low specificity.[9] The present case depicted a classical picture with spindle-shaped cells arranged in whorls with a wavy nuclei. It also showed strong positivity for S-100 protein, confirming the diagnosis.Lack of encapsulation makes neurofibromas more prone to recurrence. Hence, radical surgical procedures like en- bloc resection and hemi-mandibulectomy are considered as the management of choice.[10]

CONCLUSION

Neurofibromas of the oral cavity are rare lesions and the intra-osseous lesions are even rarer. Given the wide array of non-specific radiological features, neurofibromas must always be considered in the radiographic differential diagnosis of lesions pertaining to the jaws, especially the posterior mandible. Histopathological examination followed by immunohistochemistry is mandatory to confirm the diagnosis. Surgical excision with wide local margins is the management of choice. Since neurofibromas are not well encapsulated, chances of recurrence are high. The patients should be kept on a follow-up to rule out recurrence as well as malignant transformation.

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