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CASE REPORT

Neurofibroma of the mandible masquerading as temperomandibular joint ankylosis: Report of a unique case

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Abstract

Neurofibroma (NF) is a benign neoplasm of non-odontogenic origin arising from peripheral nerves. Its appearance in the oral cavity is rare and accounts for only 6% of all benign oral cavity tumors. We present an uncommon case report of a 4-year-old boy who reported to us with the complaint of reduced mouth opening for the age of 1½ years, which gave an initial clinical impression of temperomandibular joint ankylosis. Radiographic examination showed an osteolytic lesion of the mandible with soft-tissue extension into the infratemporal fossa and fusion to the pterygoid plates. As a result, the mandible was fused to the base of the skull giving rise to extra-articular or pseudoankylosis of the mandible to the maxilla. Histopathological examination of incisional biopsy was reported as benign fibromyxoid lesion. The patient was taken up for surgical resection of the tumor under general anesthesia. The excisional biopsy specimen was reported as NF of mandible which was confirmed by immunohistochemistry. We report a case of pediatric NF of the mandible which gave the clinical picture of temporomandibular joint ankylosis. A systematic and thorough investigation is, therefore, mandatory to arrive at correct diagnosis and appropriate treatment planning. The clinical presentation, diagnostic challenges, and treatment plan of this case have been addressed in this report. This case serves to emphasize the need to recognize such conditions early, especially in children to reduce the functional and esthetic morbidity.

Introduction

Neurofibroma (NF) is a benign tumor of the peripheral nervous system and is derived from nerve sheath cells. It can present in association with neurofibromatosis Type 1 disease or as a solitary lesion often involving the 5th cranial and upper cervical nerves. Head-and-neck region accounts for approximately 25% of the solitary NFs, 5.6% of which occur in the oral cavity.[1]

The first description of a solitary NF in the oral cavity was given by Bruce in 1954.[1,2] In the oral cavity, they occur as a non-tender, submucosal, discrete mass ranging from a few millimeters to several centimeters in size. The most common sites are tongue, buccal mucosa, labial mucosa, floor of the mouth, and palate. Intraosseous NFs are associated with neurovascular bundle within the intrabony canals. However, they are very rare because bones do not contain myelinated nerves or nerve sheaths within their medullary spaces and even if it does occur, mandible is the most common site; although few cases in the maxilla have also been reported. Localized NFs present in late childhood or during teenage years and are rare in infants. The presentation in adults is in the third and fourth decades of life.[2,3]

A rare case of solitary NF of the mandible in a 4-year-old boy causing false ankylosis is reported.

Case Report

A 4-year-old boy reported along with his parents to the department of oral and maxillofacial surgery with a chief complaint of reduced mouth opening and diffuse swelling on the right side of the face for the age of 3 years. This gave the

initial clinical impression of temperomandibular joint (TMJ) ankylosis. He had difficulty in eating; having to push food through the small gap between his upper and lower anterior teeth. However, there was no history of pain, difficulty in swallowing or in speech. The parents gave no history of forceps delivery, trauma to the face, ear infection, or childhood fever. There was no relevant medical and family history. Furthermore, there was no significant abnormality noted on general physical examination.

Extraoral examination revealed a diffuse swelling extending superiorly from the level of the ala-tragal line and extending inferiorly up to 1.5 cm below the lower border of mandible on the right side. The swelling extended anteriorly from the right corner of the mouth to approximately 2 cm, in front of the tragus posteriorly; measuring about 30 × 45 × 15 mm [Figure 1]. On intraoral examination, an expansile swelling of the mandible was noted in the molar-retromolar region, extending from 85 region, posteriorly along the anterior border of ramus. There was expansion of the buccal cortical plate with obliteration of the vestibular sulcus in this region. On palpation, the inspectory findings were confirmed. The swelling was firm and non-tender and there were no pulsation, bruit, fluctuation, or crepitus felt over the site. The deciduous dentition was intact, the patient was asymptomatic with no evidence of missing, mobile, or displaced teeth.

Orthopantomogram (OPG) revealed a well-defined, homogeneous, multilocular radiolucent lesion in relation to the dental follicle of 45, 46, and 47 measuring about 4 × 3 cm in greatest dimension. The continuity of the inferior alveolar canal could not be traced. The lower border of the mandible appeared intact [Figure 2]. CECT revealed an expansile lytic lesion of the ramus and angle of mandible on the right side measuring about 25 × 15 × 28 mm. An outgrowth was noted arising from the right pterygoid plate and extending laterally into the mandible with expansion of the ramus and adjacent soft-tissue extension into pterygoid muscles with loss of fat plane [Figure 3]. Based on the clinical and radiographic findings, a provisional diagnosis of an odontogenic tumor causing false ankylosis was made. Odontogenic myxoma, unicystic ameloblastoma, ossifying fibroma, odontogenic keratocyst, and aneurysmal bone cyst were considered in the differential diagnosis.

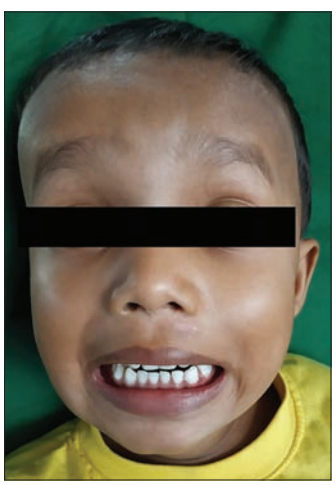

Figure 1: Pre-operative mouth opening of approx. 2 mm

Incisional biopsy was done from the right body region by creating a bony window posterior to the tooth follicle of 46, under general anesthesia (GA). The tissue specimen was firm in consistency and was removed by sharp dissection. The hematoxylin and eosin (H&E) sections of the tissue gave the impression of a benign fibromyxoid tumor. Based on the investigations and incisional biopsy report, wide local excision of the tumor with segmental mandibulectomy was planned followed by reconstruction with rib graft under GA. A submandibular incision was used to expose the tumor. There was cortical expansion seen in the body of the mandible with thinning of the buccal cortical plate [Figure 4a]. The deciduous second molar was extracted on the right side, the first osteotomy cut was made on the buccal aspect along the molar socket. The second osteotomy cut was made at the subcondylar level approx. 1.5 cm away from the superior extent of the tumor with preservation of the condyle. The tumor was seen bulging into the pterygomandibular space, extending superiorly into the infratemporal fossa and was attached to the lateral pterygoid plate and pterygoid fossa causing pseudoankylosis. The entire tumor was resected in toto along with the coronoid process.

The defect was reconstructed with a 1.8 mm angled reconstruction plate and autogenous 5th rib bone graft [Figure 4b]. Primary mucosal and skin closure were achieved. Intraoperatively, a passive mouth opening of 20 mm was achieved. External skin sutures were removed on the 10th day postoperatively and surgical site healing was satisfactory.

The resected specimen of the mandible along with the tumor measured 5 × 4 × 3 cm, white and brown in color, having a glossy surface. The tumor tissue was firm in consistency and was sent for histopathological examination. The H&E stained sections showed proliferation of plump spindle- and stellate-shaped cells with thin wavy nuclei

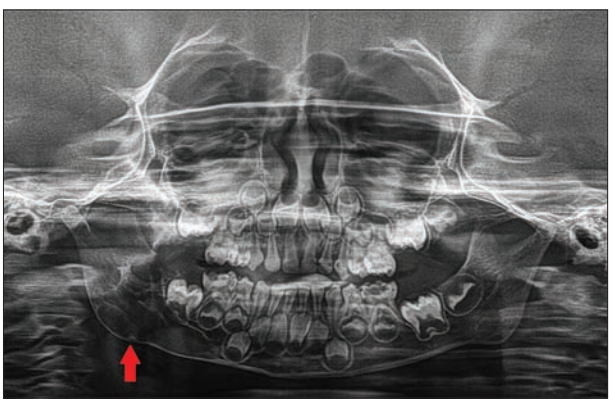

Figure 2: Orthopantomogram showing radiolucent lesion in relation to dental follicle of 45 and 46

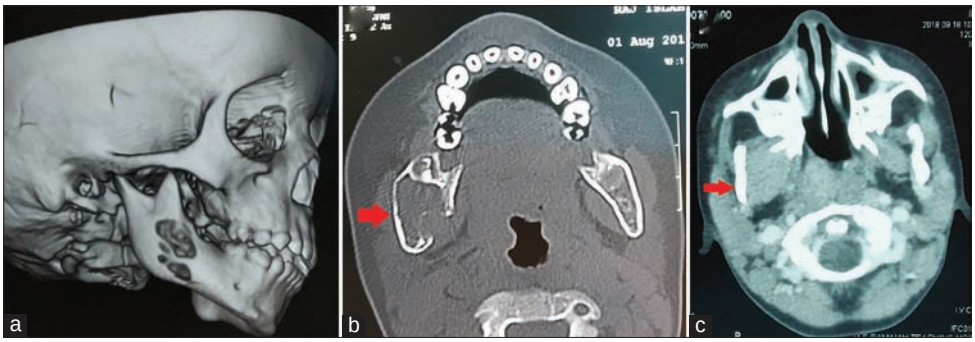

Figure 3: (a and b) 3D CT showing osteolytic lesion of the right ramus of mandible with cortical perforation on medial aspect of the ramus; (c) CECT showing expansile lytic lesion of the ramus of mandible with bony outgrowth arising from the right pterygoid plate extending laterally into the mandible

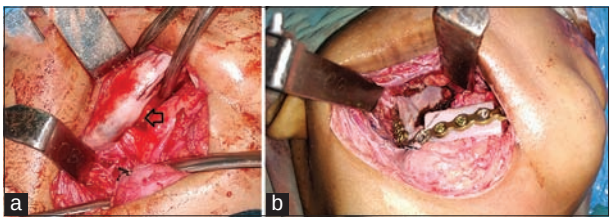

Figure 4: (a) Tumor exposed through submandibular approach; (b) reconstruction done with 1.5 mm angled reconstruction plate along with autogenous rib graft

intermingled with connective tissue fibrils [Figure 5a]. Hypocellular zones with numerous areas of myxoid changes were seen. Areas of hyalinized collagen fibers and numerous proliferating endothelial lined blood vessels of varying sizes that were engorged with RBCs were seen. Toluidine blue stained sections showed the presence of numerous mast cells distributed throughout the lesion which is pathognomonic of NF [Figure 5b].

On immunohistochemical studies, the tumor tissue showed focal positivity for S-100 confirming the diagnosis of NF [Figure 5c]. The post-operative period was not associated with any complications and the patient was discharged after 8 days. During the follow-up visits, a mouth opening of 20 mm was maintained and dental rehabilitation (restoration of decayed teeth, pulpectomy, and stainless steel crowns) was done [Figure 6]. The patient is still under periodic follow-up.

Discussion

The World Health Organization defines NF as "A benign tumor of the peripheral nerve sheath phenotype with mixed cellular components which includes Schwann cells, perineural hybrid cells, and intraneural fibroblasts."[1] According to Regezi *et al.*, neurofibromatosis type 1 accounts for 25% of intraoral NFs, therefore, a thorough family history and physical examination in a case of NF are necessary to exclude the possibility of neurofibromatosis.[4] In our case, it was a solitary lesion with no clinical signs or family history suggestive of neurofibromatosis.

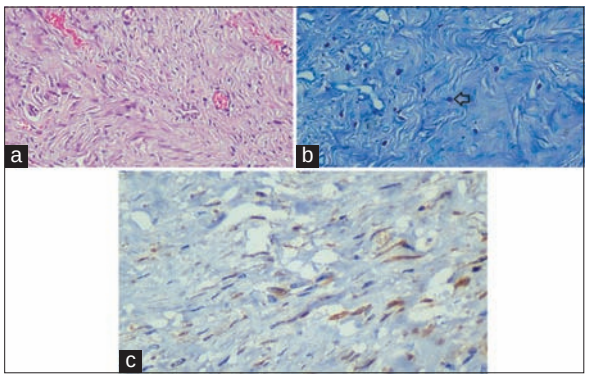

Figure 5: (a) H&E stained section shows proliferation of plump spindle- and stellate-shaped cells with thin wavy nuclei intermingled with connective tissue fibrils; (b) toluidine blue stained section shows the presence of numerous mast cells; (c) S-100 positive cells showing wavy nuclei

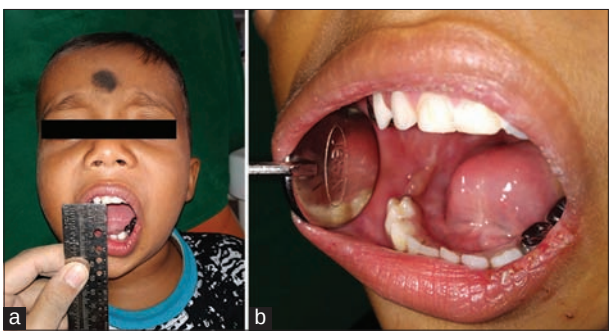

Figure 6: (a and b) Post-operative mouth opening of 20 mm after 2 months

Hubner and Lewis in their study on animal models investigated the factors responsible for the development of the lesion and found that the peripheral nerve section resulted in the formation of an expanded connective tissue cap at the end of the proximal segment. In an attempt to repair and reestablish continuity of the nerve with the distal segment, the nerve fibers penetrated into and beyond the cap, becoming entangled and

entrapped within the soft tissues.[5] The occurrence of NF has been reported between the age group of 10 months and 70 years, with a female-to-male ratio of 2:1, however, some clinical studies report that there is no gender predilection.[1,2,5] In our case, the patient is a 4-year-old boy.

Polak *et al*. analyzed 66 cases of NF in the head-and-neck region, and the site of occurrence was as follows: Tongue – 12, palate – 12, mandibular ridge/vestibule – 15, maxillary ridge/vestibule – 9, buccal mucosa – 10, lip – 4, mandibular intrabony – 2, gingival – 1, and floor of the mouth – 1. In this series, only 29 cases of solitary NF of the mandible were found.[6] In the present case, the tumor was intrabony involving the right posterior body and ramus, sparing the condyle with extension into the infratemporal region, and fusion with the pterygoid plates.

Majority of the cases in the literature are found in the posterior mandible, with a few occurring in the maxilla. The predisposition of the lesion in the posterior mandible is due to the thicker inferior alveolar nerve bundles.[5,7] Intraosseous NFs present with varying clinical symptoms ranging from a small asymptomatic to an extensive lesion causing cortical expansion with or without destruction of the cortical plates. They are usually painless but pain, paresthesia, or anesthesia of the affected area may occur due to nerve compression.[2] In our case, the patient presented with a lesion in the right posterior region of the mandible, causing bicortical expansion. The tumor perforated the lingual cortical plate and was extending into the infratemporal fossa, where it was attached to the pterygoid plates. The patient did not have any symptoms of pain and paresthesia could not be elicited.

Radiographically, intraosseous NF in the mandible can show features such as a cyst-like lesion with fusiform enlargement of the foramen and inferior alveolar canal, a branched mandibular canal, deep sigmoid notch, a decreased mandibular angle, deformed condyle, or presence of unerupted teeth.[6] Enlargement of the mental foramen has also been reported.[8] In our case, the OPG showed a well-defined multilocular radiolucency in relation to the dental follicles of 45 and 46, which gave an impression of a tumor having odontogenic origin such as odontogenic myxoma [Figure 2].

On non-contrast CT, the NF appears isodense with bone erosion of the adjacent bone. Contrast enhancement is generally homogenous and clearly demonstrates the margins of the lesion.[8] In our case, the CECT revealed an expansile lytic lesion in the right ramus and angle of the mandible with a soft tissue out growth from the medial surface of the mandible, involving the pterygoid muscles and was fused to the right pterygoid plate, causing an impression of false ankylosis of the mandible to the maxilla. The TMJ appeared normal. Differential diagnosis of pseudoankylosis of TMJ includes tetanus, inflammatory conditions, trigeminal neuralgia, and pericoronitis causing restricted mouth opening.[9]

Histologically, solitary NFs are not encapsulated and comprise a mixture of Schwann cells, perineural cells, and endoneural fibroblasts. The nerve fibers are arranged in a haphazard manner in the collagen matrix, with a few areas showing spindle cells. Mast cells are frequently present.[5] Special stains such as van Gieson and silver nitrate stains are used to differentiate NFs from other connective tissue tumor such as myofibromatosis and desmoplastic fibroma. Histochemical staining like silver nitrate with gold chloride will show more number of thick black nerve bundles and a few yellow stained collagen fibers, confirming the diagnosis of NF.[10]

NF is immunoreactive for S-100 protein.[10] This immunological marker was positive in the present case. S-100 protein and EMA are known as specific markers for Schwann cells and perineurial cells, respectively. Proliferation of all the elements of the peripheral nerve – Schwann cells, axons, perineural cells (plexiform type), and fibroblasts is seen in NFs. Hence, 30–40% of tumor cells are positive to S100 protein. Axons are neurofilament positive.[10]

Since NFs are not encapsulated, there is a high chance of recurrence if complete surgical removal is not performed. Thus, it would be appropriate to perform a more radical resection.[8] In our case, we performed a segmental mandibulectomy retaining the condylar stump which was free of the tumor. The dissection was carried into the infratemporal fossa where the tumor was attached to the pterygoid plates. The tumor was successfully excised in toto preserving the pterygoid plates. Malignant transformation of NF is rare, however, it is reported to occur in 5–16% of patients with neurofibromatosis.[10]

Conclusion

Solitary intraosseous NF is a rare entity and it might present with diagnostic difficulties. The intraosseous occurrence of solitary NF in the head and neck is comparatively rare, with the most common site being the mandible. Complete surgical removal is the ideal treatment for solitary NFs. It is important for oral and maxillofacial surgeons to consider the intraosseous NF in the differential diagnosis of the jaw radiolucencies and in false ankylosis.

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