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INTRA-OSSEOUS NEUROFIBROMA IN THE MAXILLA OF A YOUNG GIRL- A REPORT OF A RARE CASE

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ABSTRACT

The neurofibroma is a benign tumor of nerve tissue origin derived from cells that constitute the nerve sheath. It is seen as a solitary lesion or as part of a generalized syndrome of neurofibromatosis (von Recklinghausen disease of the skin). The skin is the most common site for neurofibromas. Oral lesions though found in a minority of patients with the generalized syndrome are not uncommon. The tongue and buccal mucosa are the most common sites. On rare occasions, the tumor can arise centrally within the bone. The intraosseous maxillary location is even rarer. We present a case of neurofibroma that presented in the maxilla of a young girl.

INTRODUCTION

The neurofibroma is the most common type of peripheral nerve neoplasm. [1] It is a benign, slow growing, relatively circumscribed but nonencapsulated nerve sheath tumor composed of a variable mixture of Schwann cells, perineural-like cells, fibroblasts and cells with features intermediate between these cells. Neurofibromas can arise as solitary tumors or as a component of neurofibromatosis.

Solitary tumors usually occur at a young age. Neurofibromas of the oral cavity present as submucosal, non-tender, discrete masses. The tongue, buccal mucosa, and vestibular area are the common sites, and posterior mandible is the most common intraosseous location [2]. Occurrence of this tumor is very rare in the maxillary jaw region. A case of neurofibroma in posterior maxilla of a young girl is reported here.

Case report

A 12-year-old girl was referred from the Department of Paediatrics, K.M.C. Manipal to the Department of Oral medicine & Radiology, M.C.O.D.S., Manipal in 2002 for opinion regarding non-eruption of 16 and 17. There was no associated pain in the region. Her medical history revealed that she was a known case of neurofibromatosis and was born with multiple café-au-lait spots (>6 nos) on her body. Patient's mother revealed a history of similar skin lesions in the maternal side. The girl had a moderate built and showed normal vital signs. No fresh nodules other than already mentioned in her history were present. On extra oral examination there was slight fullness of the right side of the face. Intra oral examination revealed unerupted 16 and 17. Alveolar ridge corresponding to 16 and 17 was broad facio-palatally. It was firm in consistency and revealed indentations from the opposing 46 and 47. There was no tenderness, no paraesthesia or any other symptoms of the nerve involvement. Based on the clinical findings, neurofibroma or a benign fibroosseous lesion like fibrous dysplasia and ossifying fibroma were taken into consideration.

Radiographic examination

Intraoral periapical radiographs (I.O.P.A.) w.r.t. 15 and 16 showed slight fuzziness of trabaculae in the periapical region. Further I.O.P.A. w.r.t. 16 and 17 region revealed impacted 16 and 17 with fully formed roots. The third molar was absent. Orthopantomogram demonstrated similar findings with the addition that the 3rd molar tooth buds were present in all the 3 quadrants except the first. Maxillary sinus morphology was altered on the right side. PNS view showed a reduction in size of the maxillary sinus on the right side. Since the reduction in size was evident only one side and was accompanied with slightly altered bony architecture with respect to 16 and 17 so, a localized lesion in the maxilla associated with neurofibromatosis or a benign fibroosseous lesion was again thought of. CT scan was reported to be normal except for right sided massetric hypertrophy. Serum phosphate and alkaline phosphate were found to be towards the higher limit. The radiographic picture did not provide a clear picture so the histopathological evaluation became mandatory.

Treatment

The lesion was proved to be benign through the above features, so the surgeon went ahead with the treatment and the patient was operated under G.A. The lesion found over the unerupted 16, 17 was excised. Impacted 17 was extracted and unerupted 16 was uncovered and left in situ to erupt in

the oral cavity. The patient's recovery was uneventful and the patient was discharged and kept on a regular follow up. The excised specimen was sent for histopathological examination.

Macroscopic Examination

The soft tissue specimen measured 3.5 X 2 X 2 cm in dimension. The tissue was creamish white in colour, smooth in texture and firm in consistency.

HISTOPATHOLOGICAL FEATURES

On microscopic examination it was seen that the stroma was composed of proliferating spindle cells with thin wavy nuclei. Numerous nerve bundles were present arranged in an irregular pattern. The stroma was interspersed with few inflammatory cells among which mast cells were the predominant type. Delicate connective tissue fibres were evident throughout the section. Muscle tissues showing some degenerative changes, strands of odontogenic rests and areas of haemorrhage were also evident. Correlating with the clinical findings, the final diagnosis of neurofibroma of the maxilla was arrived at. Special stains including silver stain and toluidine blue were employed to check the neural origin of the tumor and the characteristic mast cells respectively.

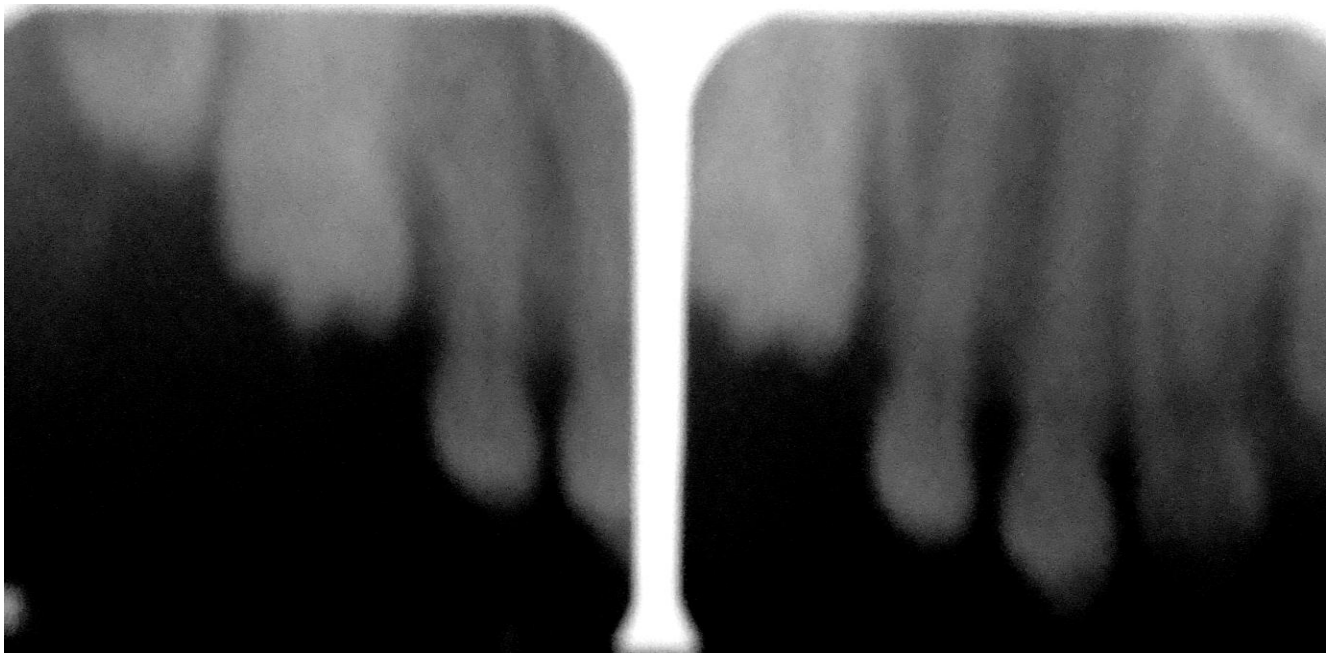

Fig 1a. Intra Oral Periapical Radiograph showing impacted 16, 17
Fig 1b: Intra Oral Periapical Radiograph showing slight fuzziness of trabeculae in 15, 16 region.

DISCUSSION

Neurofibromatosis is a relatively common hereditary condition. Among the various reported forms of this disease, neurofibromatosis type I (NF1) is the most common form and accounts for 85 – 97 % of the cases [1]. Neurofibromas of the oral cavity may occur as solitary lesions or as a part of NF1 [3]. The common site in the oral cavity is the tongue and buccal mucosa and rarely the tumor can arise centrally within the bone. In this case, the lesion presented on the posterior maxilla.

Fig 2: PNS view showing smaller sized maxillary sinus on right side

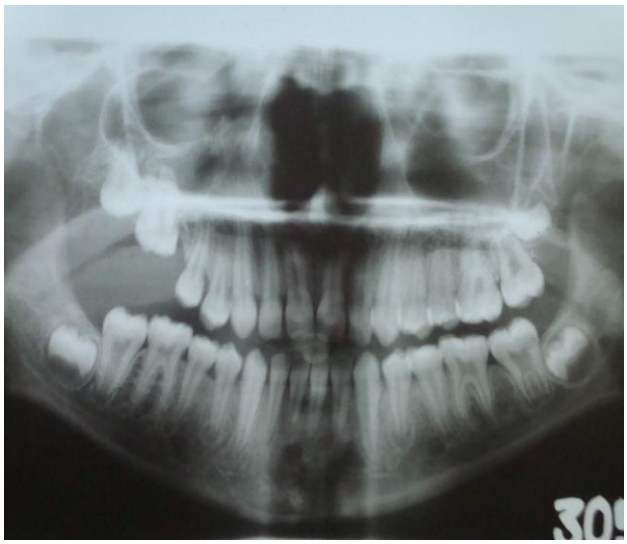

Fig 3: OPG showing fully formed 16, 17 - impacted almost 1cm above (apical) to the occlusal plane. Tooth bud of 18 is absent. Right maxillary sinus morphology is altered.

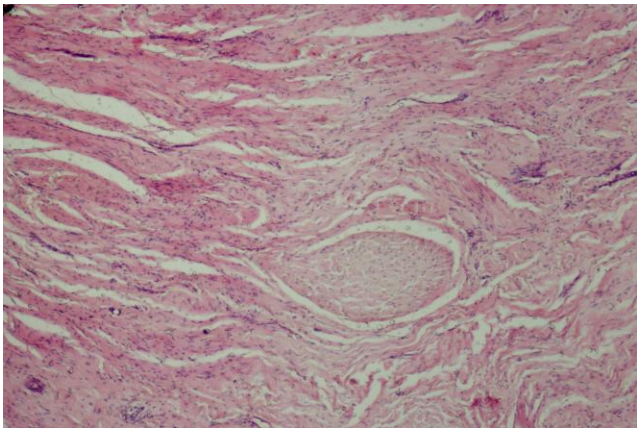

Fig 4 : Spindle shaped cells with (H&E 10x)

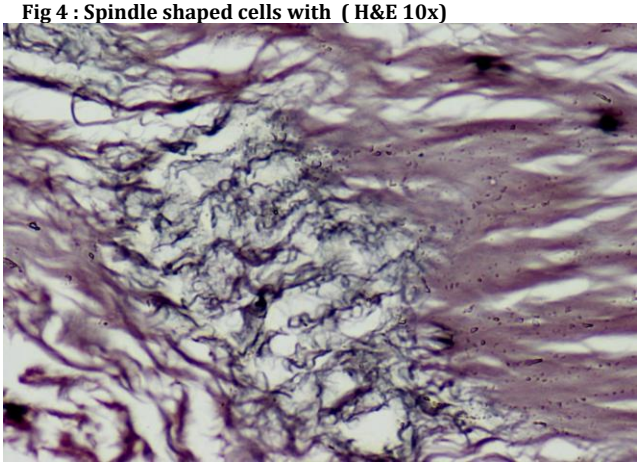

Fig 5. Nerve elements in the tumor (Silver stain 10X)

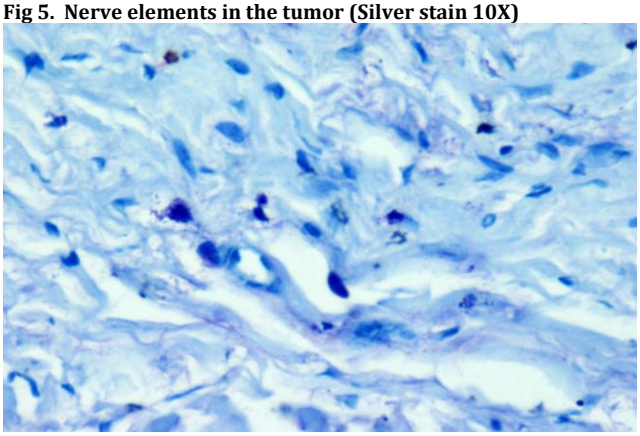

Fig 6. Many Mast cells in the stroma (Toluidine blue stain 10X)

Microscopically, it presents as widely separated irregular spindled or stellate cells with a wavy dark staining nucleus and long thin cytoplasmic processes. Our case presented with an identical histopathological picture. The stroma is usually fibrous or myxoid in nature rich in mucopolysaccharides such as hyaluronic acid and chondroitin-6-sulfate, and N-acetyl-galactosamine, N-acetyl-neuraminic acid, D-glucose or D-mannose. The amount of cells & fibromyxoid stroma varies [4]. The stroma contains mast cells & lymphocytes, which were also seen in this case [5]. Less frequently, neurofibromas may be cellular and composed of fascicles of Schwann cells in a uniform collagen matrix devoid of myxoid substance.

Proliferating nonmyelinating Schwann cells secrete chemoattractants such as the KIT ligand, and angiogenic factors such as the heparin-binding growth factor midkine. These chemicals promote the migration of different kinds of cells that are heterozygous for the NF1 gene into the hyperplastic lesions created by the nonmyelinating Schwann cells. These cell types include fibroblasts, perineurial cells, endothelial cells, and mast cells. The mast cells then secrete mitogens or survival factors that alter the developing tumor microenvironment and result in the formation of neurofibroma. Dermal and plexiform neurofibromas do differ in later development stages, but the details are unclear at this point [6]. Focal atypia or rare mitoses is common. Neurofibromas demonstrate S100 positivity, which can be variable in a given lesion. Variable staining for CD34 is also seen. Collagen IV stains many cells in pericellular pattern [3]. Special stains like silver stains and toluidine blue can aid in an early diagnosis by demonstrating the neural element and mast cells respectively. This was also evident in the current case.

Clinically benign neurofibromas, both the usual and cellular types, can have significant cytologic atypia that can be accompanied with low mitotic activity. Conservative surgical excision for these tumors is adequate [7]. If a marked degree of the above is noted, malignant transformation should be considered.

Here a rare case of intraosseous neurofibroma of the maxilla is reported with characteristic features and diagnosis of which has been aided by the use of special stains. The patient has been followed up for ten years now and there has been no recurrence or malignant transformation till date.

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