

Original research article**Non-ossifying maxillary fibroma: A case study**

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Abstract

Background and objectives: The metaphysis of children's long bones are where non-ossifying fibromas, also known as NOFs, are most frequently found to be benign, non-neoplastic lesions. Objective is to confirm a diagnosis through investigation into the radiographic, clinical, and histologic characteristics of non-ossifying fibroma of the maxilla.

Method: A 14-year-old male, has been experiencing swelling in his left cheek for the past eight years, and he sought treatment at the government ENT hospital in Vishakhapatnam. It was characterised by a slow, steady progression in the absence of precipitating, ameliorating, or associative influences.

Result: Nasal obstruction, pain, bleeding, impaired eyesight, breathing or swallowing complaints, loss of smell, numbness, appetite loss, or weight loss have not been reported. No significant medical history. No family history of such complaints; no interracial or consanguineous marriages among parents. Oriented, afebrile, CVS-S1, S2+; per abdomen soft, no organomegaly; RS has normal vesicular breath sounds; No pedal edema or generalized lymphadenopathy was found.

Conclusion: The NOF is always eccentric and ovoid, thinning and expanding the underlying cortex radiographically. Periosteal reaction and cortical violation are rare. Histologically, the NOF has spindle-celled fibrous tissue in a storiform pattern, a variable amount of multinucleated large cells, hemosiderin pigment in fibroblasts, and lipid-laden histiocytes (Xanthoma cells). The lack of adult NOF reports suggests that most lesions spontaneously recover. Recurrence in gnathic bone NOF cases is rare after curettage or resection.

Keywords: Non-ossifying fibroma, metaphysis, maxilla

Introduction

The non-ossifying fibroma, which is commonly abbreviated to NOF for short, is a benign and non-neoplastic lesion that is most commonly discovered in the metaphyses of children's long bones. It is also the most common location for this type of lesion. The mandible has shown evidence of the NOF, despite the fact that it is a rather rare variation [1,2,3].

The NOF in the extra-gnathic skeleton has a distinct radiographic appearance, is typically asymptomatic, and has a varied histologic make-up. Moreover, the NOF usually be found in metaphysis of long bones. In addition, the NOF commonly located anywhere in the skeleton that is outside of the gnathic region.

The association that exists between the radiographic appearance of the lesion, the clinical presentation of the lesion, and its histology makes it possible to separate the NOF from odontogenic cysts and tumours as well as nonodontogenic cysts and tumours [4,5].

The NOF has been described in a number of different ways, and many different accurate synonyms have been used to describe it. These words describe the normal stages that occur when the lesion progresses. Some of the names that have been given to this disorder are histiocytic fibrous defect, metaphyseal fibrous defect, fibrous cortical defect, fibrous xanthoma, and histiocytic xantho granuloma. Another name for this condition is fibrous xanthoma [5,6].

At the moment, the terms "fibrous cortical defect" (FCD) and "non-ossifying fibroma" are the two that are utilised the most frequently in order to define these lesions. Both of these terms are abbreviated from the medical term "fibrous cortical defect" (NOF)

We present a rare instance of this intriguing phenomena and carry out an in-depth analysis of the radiographic, clinical, and histologic features of the Non-ossifying fibroma of maxilla [6].

Material and Method

A 14-year-old male, has been experiencing swelling in his left cheek for the past eight years, and he sought treatment at the government ENT hospital in Vishakhapatnam from. It was characterised by a slow, steady progression in the absence of precipitating, ameliorating, or associative influences.

The patients pre-existing conditions, family history were examined, evaluation of swelling and ENT and histopathological finding, and differential diagnosis were done.

Result

No reports of nasal obstruction, pain, bleeding, impaired vision, problems breathing or swallowing complaints, loss of smell, numbness, appetite loss, or weight loss. A medical history without high blood pressure, diabetes, asthma, or epilepsy. No relatives with identical problems; parents did not marry within their race or religion. Cvs-s1, s2+, no murmur, afebrile, alert, orientated; P/A: soft, no organomegaly, RS: normal vesicular breath sounds present; CNS- no focal neurological deficit, no generalized lymphadenopathy or pedal edema is seen. Inspection - A single oval-shaped swelling on the left cheek, no scar, sinuses, or dilated veins, skin above swelling is normal.



Fig 1: ENT examination

Palpation: swelling extends superiorly to infra orbital edge, medially to left lateral wall of nose, and inferiorly to left upper alveolus. Firm, non-tender, no local rise of temperature, no crepitus. NOSE: external bony cartilaginous framework showed slight deviation to right side. On Anterior rhinoscopy - on left side a mass from lateral wall touches the septum; on right side-normal. Cotton wool test-reduced movement on left side compared to right side and Cold spatula test showed reduced mist formation on left side.

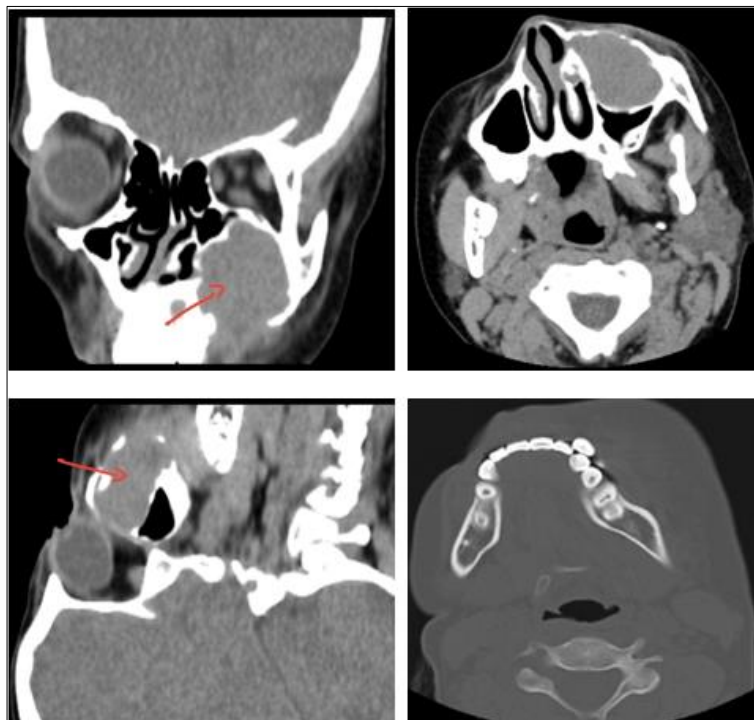


Fig 2: CT Para nasal sinuses

Oral cavity and oropharynx examination: normal, no swelling or development in alveolar region. Ears: right tm intact, left tm intact. Large cystic lytic lesion on left side of maxilla measuring 58 * 9 mm, potentially ameloblastoma, with substantial cortical remodelling. General anaesthesia given, paring done through sublabial incision.

Macroscopically, grey-brown tissue piece. Microscopically, sections showed predominantly spindle cells in fascicles with storiform pattern and occasional multinucleated giant cells.

Mild nuclear atypia, mitotic numbers less than 2/10 hpf. Pathology suggests fibrous cortical defect (non-ossifying fibroma) and no cancer.

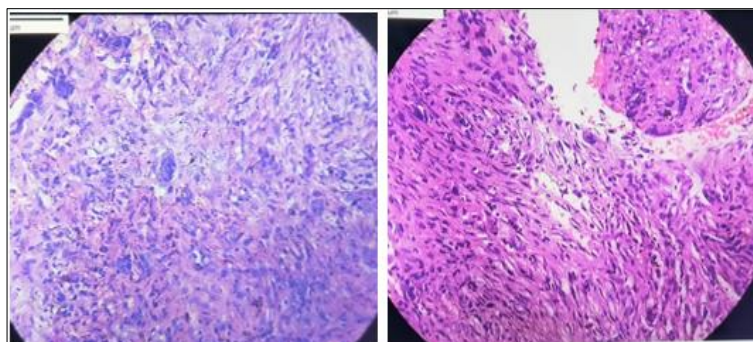


Fig3:Histopathological examination

Differential diagnosis indicated giant cell granuloma is more frequent in children and young adults. Both NoF and benign fibrous histiocytoma of the bone have well-defined, sclerotic borders. BFH is more common in older people, is commonly symptomatic without accompanying fractures, and can recur. Ameloblastoma may look radiographically as a multicystic radiolucency, but the septae in the usual "soap bubble" appearance are rougher and the locules are larger than in the nof.

Discussion

In this case, Nasal obstruction, pain, bleeding, impaired vision, problems breathing or swallowing, loss of smell, numbness, lack of appetite, or weight loss have not been reported. Medical history free of major risk factors. No history of such complaints in the family; no interracial or no consanguineous among the parents. Afebrile, alert, and orientated; CVS-S1, S2+; Per abdomen soft, no organomegaly. Typical vesicular breath sounds audible in RS; central nervous system; NFND, no pedal edema or generalized lymphadenopathy has been detected.

Few instances of metaphyseal fibrous defects were found in the literature. Children and young adults were disproportionately affected by cases in the jaws. Fibrous cortical defect (Nonossifying fibroma) of the metaphysis is typically asymptomatic and may be bilateral. A metaphyseal fibrous defect will typically show up on radiographs as a radiolucent region with a distinct border.

According to the World Health Organization's (WHO) histologic classification of tumours, the lesion demonstrates spindle-celled fibrous tissue with a storiform pattern and contains a variable amount of multinucleated giant cells, hemosiderin pigment, and lipid-bearing histiocytes (xanthoma cells) [6,7].

All of these were present in the presented example with the exception of hemosiderin pigment, which is not typically present. The patient was diagnosed with a metaphyseal fibrous defect (Nonossifying fibroma). Giant cell tumour, central fibroma, bone cyst, solid ameloblastoma, and fibrosarcoma are all potential diagnoses.

Enucleation or curettage is the typical procedure for dealing with a metaphyseal fibrous defect in the jaws. Nonetheless, more complex surgical techniques such as segmental resection may be necessary for large tumours. Although cases of malignant transformation have been observed, most patients recover without complications after surgery. A disruption in bone development may contribute to the pathophysiology of metaphyseal fibrous abnormalities, which are most common in young people. However, at present, the mechanisms responsible for this lesion's development are unknown, and its pathophysiology categorization must wait for more research [7,8].

Examination reveals an oval-shaped swelling on the left side of the cheek; there are no visible scars, sinuses, or dilated veins, and the skin over the swelling appears normal. The left cheek swollen and palpated. The entire swelling was hard and nontender, and there was no increase in temperature or crepitus. The external nasal bone and cartilaginous framework was normal.

The results of an anterior rhinoscopy showed a slight deviation to the right, a mass pressing against the septum from the left side, normal results from a cotton-wool test, and noticeably less movement on the left side than the right. Reduced formation of mist on left-side during cold spatula test. Oral cavity and pharynx examination reveal no abnormalities, such as growth or swelling, in the alveolar region. Examination of the ears reveals that both the right and left ears have intact tympanic membranes. Paranasal sinuses were sectioned serially in an axial, coronal, and sagittal orientation at a thickness of 0.3 mm (1 mm at OMC).

Significant remodelling of the cortex was observed, which may be indicative of neoplastic ameloblastoma, and a large cystic lytic lesion involving the left side of the maxilla was observed (measuring 58 mm by 9 mm). General anaesthesia administered, paring through a sublabial incision, the fibro-osseous bone over the maxilla accessed and removed using lucs forceps [8,9].

Our study denoted, In macroscopic view, it was clear that a solitary piece of greyish-brown tissue. A look under the microscope tumor tissue pieces and a few bone trabeculae were seen in the sections, and the cells were primarily spindle-shaped and grouped in fascicles and a storiform pattern, with the multinucleated giant cells. Mitotic numbers less than 2/10 hpf, and nuclear atypia is modest. There was no sign of cancer in the pathology report, hence the diagnosis was "fibrous cortical defect (non-ossifying fibroma). Giant cell granuloma was the differential diagnosis. Patients of any age can be diagnosed, however those under the age of

30 have the highest incidence. It manifests as a swelling and causes no symptoms in most cases. Benign fibrous histiocytoma is indistinguishable from non-ossifying fibroma (NOF) histologically, and both may exhibit well-defined, sclerotic borders on radiographs. Nonetheless, the bfh is more common in the elderly, can cause symptoms even when no other fractures are present, and can come again ^[9].

While ameloblastoma can also appear on radiographs as a multicystic radiolucency, the septae in the traditional "soap bubble" appearance are frequently coarser, and the individual locules are usually larger than those seen in the NOF.

Conclusion

Most often found in the metaphyses of children's long bones, the non-ossifying fibroma (NOF) is a benign, non-neoplastic lesion. Reports of the NOF in the GNATHIC bones are uncommon, although they do occur. Compared to NOF in the long bones, NOF of the gnathic bones appears to present at a later age, suggesting a possible age-related difference in onset. Twenty instances of NOF in the mandible were studied, and the average age at presentation was 19 years and 7 months. Non-ossifying fibromas in the long bones affect 30–40% of children older than 2 years old, with the peak occurrence between the ages of 4 and 8 years old, according to the orthopaedic literature.

Although its exact cause is unknown, most scientists believe that NOF results from a growth disorder or dystrophic calcification rather than a neoplasm. Nonetheless, an odontogenic origin can be ruled out because the NOF has been found in non-dental tissues. If you look at a radiograph of a NOF, you'll notice that it's always shaped like an eccentric oval and that it causes the underlying cortex to thin and grow. The periosteum does not react and the cortex is not visibly broken. Lesions have a multilocular appearance, and bone sclerosis defines the inner margin of the lesion. Spindle cells, multinucleated giant cells, hemosiderin pigment inside fibroblasts, and lipid-laden histiocytes make up the NOF on a histological slide (xanthoma cells). Adult cases of NOF are uncommonly reported in the literature, suggesting that most of these lesions also heal on their own. Gnathic NOF has been effectively treated by curettage or resection in all reported cases, with no recurrences.

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Conflict of interest

Nil

References

1. Stell, Maran's text book of head and neck surgery and oncology 5th edition.
2. Schajowicz F, Ackerman LV, Sissons HA, *et al.* International histological classification of tumors (histological typing of bone tumors). Geneva: world health organization, 1993.
3. Mizukawa N, Nishijima Y, Nishijima K. Metaphyseal fibrous defect (non-ossifying fibroma) in the mandible. A case report. *Int j oral maxillofac surg.* 1997;26:129-30.
4. Chrcanovic B, Albanese AI, Freire-Maia B, *et al.* Non-ossifying fibroma (metaphyseal fibrous defect) of the mandible. *Oral Maxillofac Surg.* 2011;4:233-7.
5. Fletcher CDM, Unni KK, Mertens F. World health organization: classification of tumours. Pathology and genetics of tumours of soft tissue and bone. Lyon: IARC press, 2002.
6. Dahlin DC. Fibroma (Non-Osteogenic fibroma of bone, metaphyseal fibrous defect), Myxoma, Cortical Desmoid, Fibromatosis and "Xanthoma". In: Thomas CC, editor. Bone tumors. General aspects and data on 6,221 cases, 3rd ed. Springfield, IL: Charles C Thomas, 1978, 122-36.
7. Blau RA, Zwick DL, Westphal RA. Multiple non-ossifying fibromas. A case report. *J bone joint Surg. Am.* 1988;70:299-304.
8. Hudson JW, Livesay KW, McCoy JM. Condylar Lesion. *J Oral Maxillofac Surg.* 2003;61:824-6.
9. Hudson JW, Livesay KW, McCoy JM. Condylar Lesion. *J Oral Maxillofac Surg.* 2003;61:824-6.
10. Aldred MJ, Breckon JJW, Holland CS. Non-Osteogenic Fibroma of the Mandibular Condyle. *Br J Oral Maxillofac Surg.* 1989;27:412-6.