

Asymptomatic Massive Fibrous Dysplasia Obliterating Maxillary Sinus Extending to the Skull Base: a Case Report and Review of Literature

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ABSTRACT

Fibrous dysplasia is a rare bone condition in which normal bone is replaced by aberrant fibrous tissue. The patient may come to evaluate with other diseases and while doing investigations, it is observed. Here we present a case of 24 years male with fibrous dysplasia who came to our OPD with swelling around the left cheek region for 5 days. The patient underwent a Computerised tomography scan which revealed an expansile, sclerotic, cystic ground-glass appearance that extended to the maxillo-alveolar ridge, right sphenoid sinus wall, right pterygoid plate, and zygomaticofacial suture. The right maxillary sinus was almost obliterated. These radiographical features of the lesion were indicative of fibrous dysplasia in the maxilla.

Keywords: computerised tomography scan; fibrous dysplasia; ground glass appearance.

INTRODUCTION

Fibrous dysplasia is a rare pathological condition characterized by the replacement of normal osseous tissues by abnormal fibrous tissue. This condition was first described by Lichtenstein in 1938 but published in the medical literature with Jaffe in 1942.¹ The patient may be asymptomatic however some patients present with unspecific symptoms such as nasal obstruction, headache, and facial pains, in one case, the lesion was discovered incidentally.² However, CT scan help to reach the diagnosis although biopsy is needed for confirmation. The abnormal expansile growth of lesions can often be identified as either

radiolucent or radiopaque or a mixture of both.³

Fibrous dysplasia is divided into monostotic, in which only one bone is affected, and polyostotic where two or more bone is affected.⁴ The most common sites are ribs, femur, and craniofacial region. In the craniofacial region, mandible, maxilla, and zygoma can be affected. Monostotic bone replacement is twice more common in the maxilla than mandible.⁵

We report a case of 20 years male who present with left cheek abscess and On CT scan, massive fibrous dysplasia of right maxillary sinus was found.

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CASE REPORT

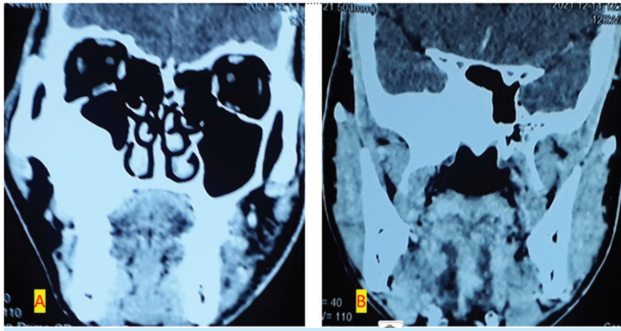


Figure 1. CT scan Coronal view at cribriform plate level showing hyperdense ground glass opacity at infero-lateral wall of right maxillary sinus (A) and extending to sphenoid sinus (B).

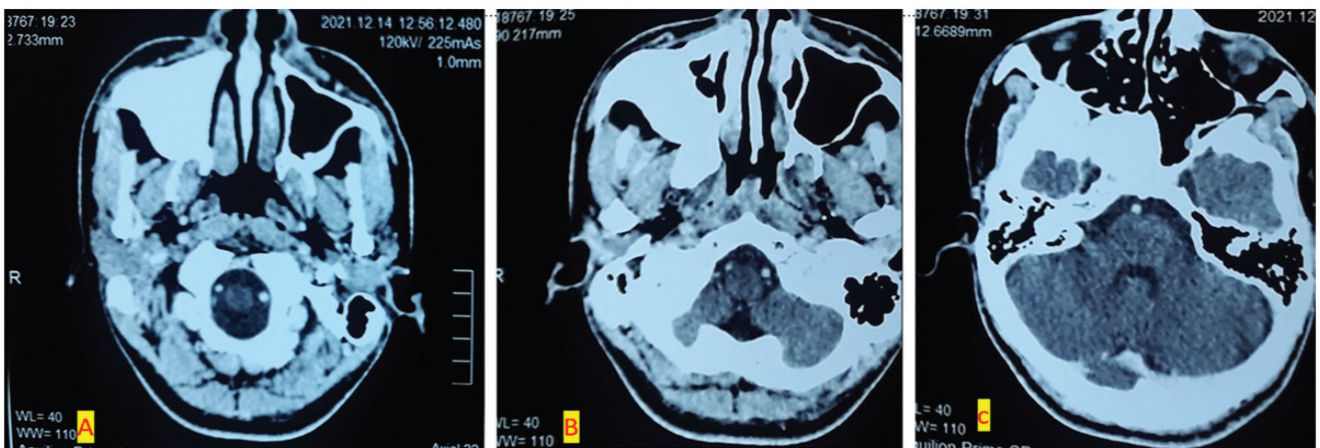


Figure 2. CT scan axial view at level of lower end of inferior turbinate showing hyperdense ground glass opacity occupying right maxillary sinus (A) at level of sphenopalatine foramen encroaching pterygoid plate (B) and extending to skull base (C).

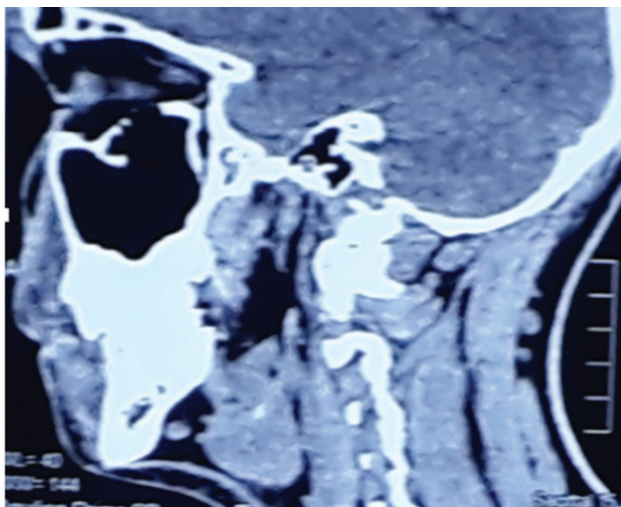


Figure 3. CT sagittal view at right paramedian region showing tumor involving the floor of maxillary sinus and alveolar crest.

A 16-year-old male presented in Otolaryngology OPD at Gandaki Medical College with pain in left cheek region for five days. Examination revealed a tender swelling at the left cheek. Patient was referred to the imaging department. CT scan -coronal view revealed a radio-opacity in the right side of the maxilla extending into maxillary sinus extending to right sphenoid sinus Figure 1 (A,B and C) CT-Axial view revealed that the lesion nearly obliterated the right maxillary sinus and extending to the skull base. Figure 2 (A and B). CT scan Sagittal view showed a radio-opaque lesion characterized by homogenous ground-

glass appearance at involving floor maxillary sinus and alveolar crest (figure 3) and it blended with surrounding normal bone. In our case, we didn't plan for aggressive surgical intervention as the patient was asymptomatic.

DISCUSSION

Fibrous dysplasia is a rare bone disorder that commonly occurs in the craniofacial region typically located in zygomatic and maxillary bone.² The age of our patient was 24 years which was similar to the case reported by LC Keijser et al.⁶ Monostotic form is commonly found in the older group compared to polyostotic form which is common in children

under 10 years old.³ It starts at younger age however it is not diagnosed late. Patient comes with symptoms like facial asymmetry, facial pain, enlargement of craniofacial regions. It may cause displacement of permanent teeth, interfere with the eruption of new teeth, and contribute to malocclusion.⁷ Our case was asymptomatic and had no facial asymmetry and he came for treatment of left buccal abscess. Confirmation is achieved by plain radiographs, computed tomography scan, Magnetic resonance imaging and biopsy. Abnormal growth patterns which contains either radiopaque or radiolucent or a mixed are found. Various distinct bone patterns include granular, cotton-wool and, orange-peel, fingerprint-like patterns are described.³ CT scan was suggestive of fibrous dysplasia in our cases. As he was asymptomatic, we counselled him for regular follow up. There is no cure of fibrous dysplasia, however treatment is able to reduce the effects of symptoms. Bisphosphonates are given to prevent further bone loss.⁴ Role of steroids has been reported mainly in case of

optic nerve compression.⁸

Surgery can be done if facial asymmetry is present, however, it is unable to reverse the complications. Surgery could be performed either via conservative approach i.e. shaving or radical excision followed by reconstruction.⁹ Indication of surgery depends on site of involvement, choice of patient and availability of multidisciplinary team.

CONCLUSIONS

Fibrous dysplasia is a rare bone disorder characterized by the replacement of normal osseous tissue by abnormal fibrous tissue. Maxillary bone fibrous dysplasia is most common among the fibrous dysplasia in craniofacial region. CT scan is able to pick up the asymptomatic cases incidentally. Treatment can be done, if patient has symptoms

Conflict of interest

The Authors have declared no competing interests.

REFERENCES

1. Belsuzarri TA, Araujo JF, Melro CA, et al. McCune-Albright syndrome with craniofacial dysplasia: Clinical review and surgical management. *Surg Neurol Int* 2016; 7: S165-169. 2016/04/09. DOI: 10.4103/2152-7806.178567.
2. Lee JS, FitzGibbon EJ, Chen YR, Kim HJ, Lustig LR, Akintoye SO et al. Clinical guidelines for the management of craniofacial fibrous dysplasia. In: *Orphanet journal of rare diseases* 2012; S2:1-19. <https://doi.org/10.1186/1750-1172-7-S1-S2>
3. White S and Pharoah M. *Oral Radiology: Principles and Interpretation* by Stuart White and Michael Pharaoh. Elsevier, St. Louis, Mo, 1982. URI: <http://vlib.kmu.ac.ir/kmu/handle/kmu/88736>
4. Anitha N, Sankari SL, Malathi L, Karthik R et al. Fibrous dysplasia-recent concepts. *Journal of pharmacy & bioallied sciences* 2015; S 7: S171-S172. doi: 10.4103/0975-7406.155892
5. Menon S, Venkatswamy S, Ramu V, Banu K, Ehtaih S, Kashyap VM. Craniofacial fibrous dysplasia: Surgery

- and literature review. *Annals of maxillofacial surgery*. 2013 Jan-Jun; 3(1): 66–71. doi: 10.4103/2231-0746.110088
6. Keijser LC, van Tienen TG, Schreuder HB, Lemmens JA, Pruszczynski M, Veth RP. Fibrous dysplasia of bone: management and outcome of 20 cases. *Journal of surgical oncology*. 2001 Mar;76(3):157-66. <https://doi.org/10.1002/jso.1028>
 7. Alves N, de Oliveira RJ, Takehana D, Deana NF. Recurrent monostotic fibrous dysplasia in the mandible. *Case reports in dentistry*. 2016 May 31;2016. <https://doi.org/10.1155/2016/3920850>
 8. Guruprasad Y, Prabhakar C. Craniofacial polyostotic fibrous dysplasia. *Contemporary clinical dentistry*. 2010 Jul;1(3):177 doi: 10.4103/0976-237X.72787
 9. Markov P, Syed AZ, Markova C, Mendes RA. Maxillofacial fibrous dysplasia: a diagnostic challenge. *Case Reports*. 2016 Jun 29;2016:bcr2016215874 <http://dx.doi.org/10.1136/bcr-2016-215874>

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