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A Rare Case of Fibrous Dysplasia of Ethmoid Sinus – A Case Report & Review of Literature

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Abstract

Introduction: Fibrous dysplasia is a benign skeletal lesion where the normal medullary bone is replaced by immature and fibrous stroma due to abnormal osteoblastic differentiation of unknown etiology.

Case report : A 43 year old female with complains of nasal obstruction and anosmia found to have a bony hard mass between septum and middle turbinate. CT scan showed a fibroosseous lesion of ethmoid with ground glass appearance. Biopsy confirmed fibrous dysplasia. Endoscopic removal of mass was done**.**

Discussion: Fibrous dysplasia mostly occurs in the long bones, craniofacial bones are second common site of involvement. Fibrous dysplasia of paranasal sinus is rare. Conservative surgery is indicated for symptomatic patients.

Conclusion: Fibrous dysplasia is a rare disease of ethmoid sinuses. In limited lesion, an endoscopic sinus surgery could serve as an optimal treatment of choice.

Keywords: Fibrous dysplasia, Ethmoid

Introduction

Fibrous dysplasia is one of the fibrous osseous lesions of bone. Among fibrous dysplasia of head and neck, the maxilla and mandible are the most frequent sites. It is rarely found in paranasal sinus. The 'groundglass' appearance on CT scans with bone window is the most useful radiographic sign for diagnosis. Excision of the mass is the treatment of choice. The present report aims at describing a case of fibrous dysplasia of ethmoid sinus treated by endoscopic sinus surgery.

Case Report

A 43 year old female presented with complaints of nasal obstruction for past one year. It was associated with sneezing, mucoid nasal discharge and loss of smell. There was no significant past history of trauma or surgery. She was under treatment with steroid inhalers for bronchial asthma since young age.

On examination she was thin built and under nourished having only 30kg body weight. General inspection revealed no lesions anywhere else in the body. Physical examination found neither facial asymmetry nor limitation of eye movements. Anterior nasal examination revealed pale polypoidal smooth mass seen between septum and middle turbinate in left nasal cavity which was sensitive to touch but was not bleeding on touch and it was bony hard in consistency (fig.1). There was no tenderness of paranasal sinuses.

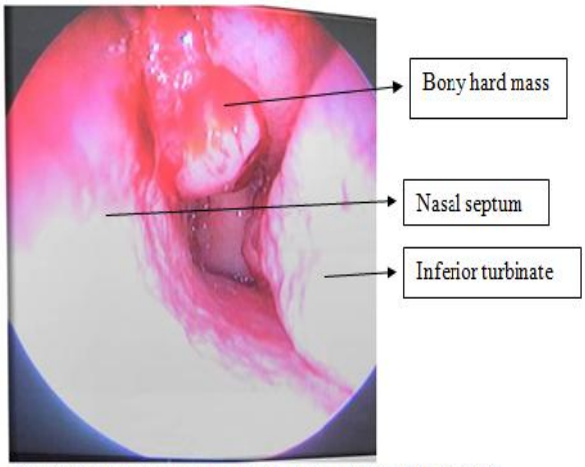

Fig.1 examination of nose revealing mass between septum and middle turbinate.

Computed tomography of sinuses revealed well defined expansile 3.5x3.5x3cm lesion having ground glass matrix with peripheral new bone formation, involving left side of ethmoidal sinus with extension into sphenoidal sinus and left nasal cavity also extending to cribriform plate (fig.2a, 2b).

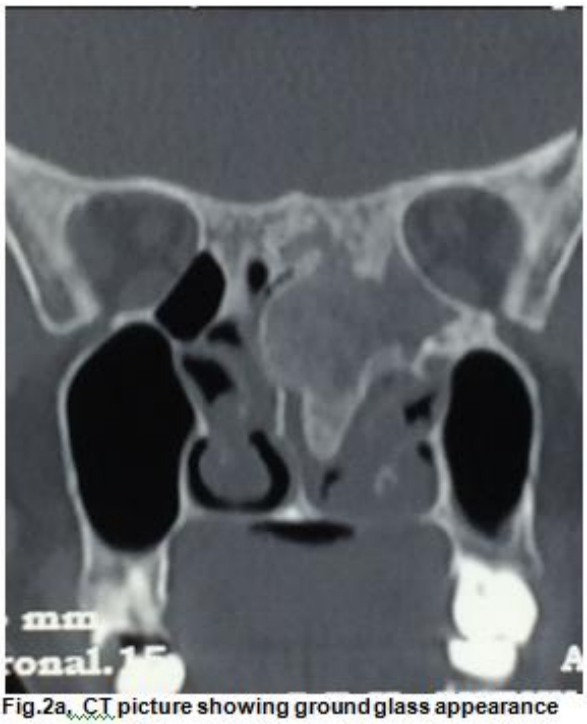

Fig.2a. CT picture showing ground glass appearance

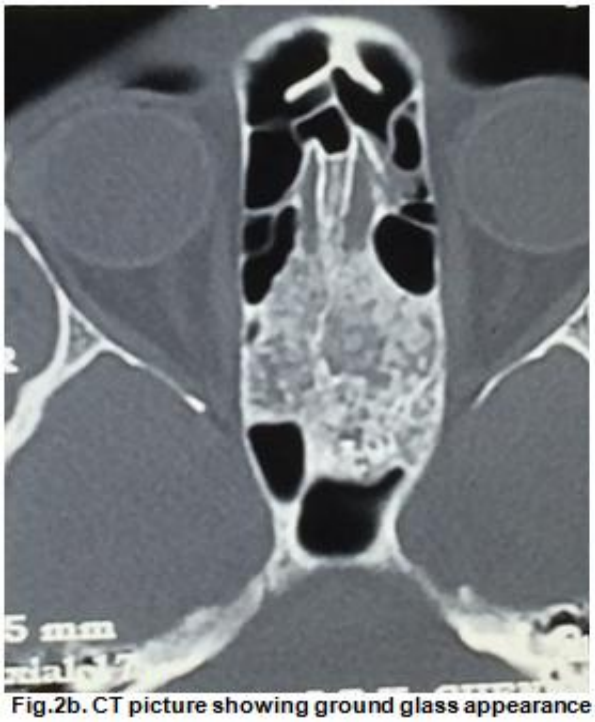

Fig.2b. CT picture showing ground glass appearance

Diagnostic nasal endoscopy done and biopsy was taken and sent for histopathological examination.

Histopathology revealed irregular seams of woven new bone in a cellular stroma without osteoblastic rimming, features confirmatory of fibrous dysplasia (fig.3).

Fig.3 Histopathology showing fibrous dysplasia

A complete workup was done. There was no intracranial extension of tumor. Ocular examination was normal. Skeletal survey showed no similar bony lesions.

Endoscopic excision of the mass was planned. Mucosal flap removed from lesion (fig.4a). Using micromotor and burr bony hard mass over left middle turbinate, anterior ethmoid, posterior ethmoid, anterior wall of sphenoid was removed (fig.4b). Posterior bony septum removed and ethmoid remnants were smoothened using diamond burr (fig.4c). Mucosal flap separated from middle turbinate repositioned over raw bone (fig.4d). Complete hemostasis obtained. Nasal packing was done.

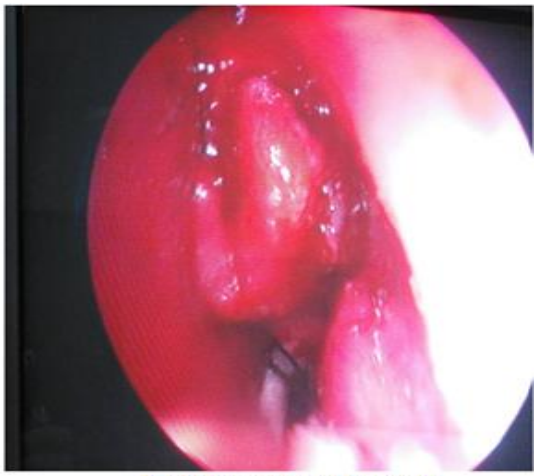

Fig 4a flap separated from lesion

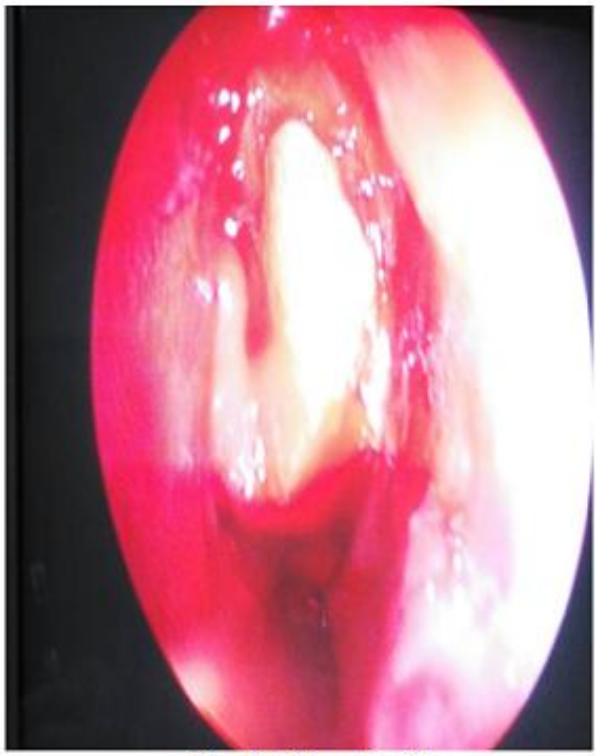

Fig. 4b drilling ethmoid

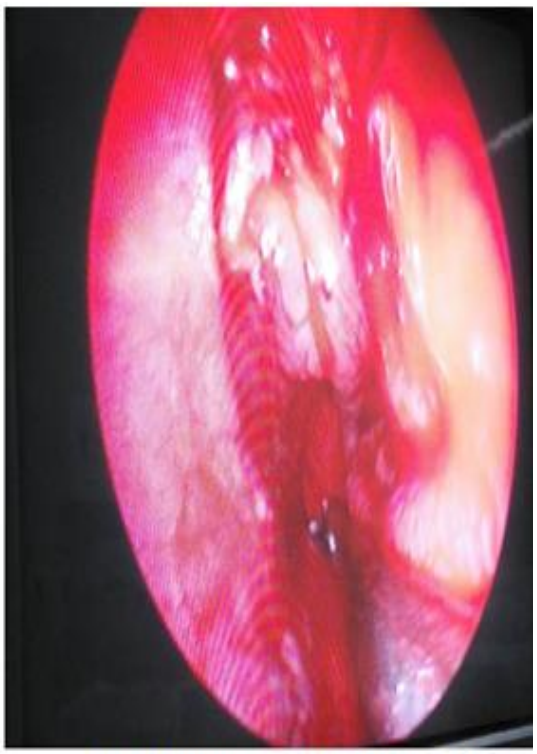

Fig. 4d post op choana after flap reposition

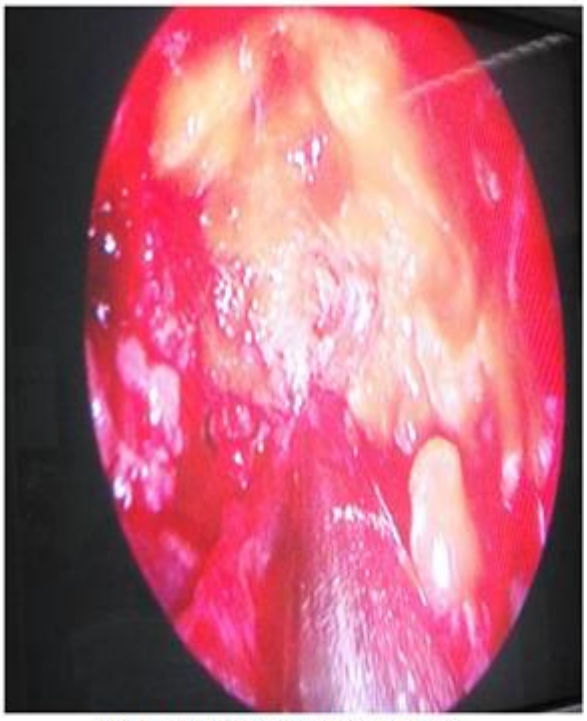

Fig. 4c after removal of bony septum

Postoperative period was uneventful. Suction clearance was done using endoscope. Patient was completely free of symptoms. Postoperative histopathology confirmed fibrous dysplasia. Diagnostic nasal endoscopy done one month after the procedure showed no recurrent lesion (fig.5).

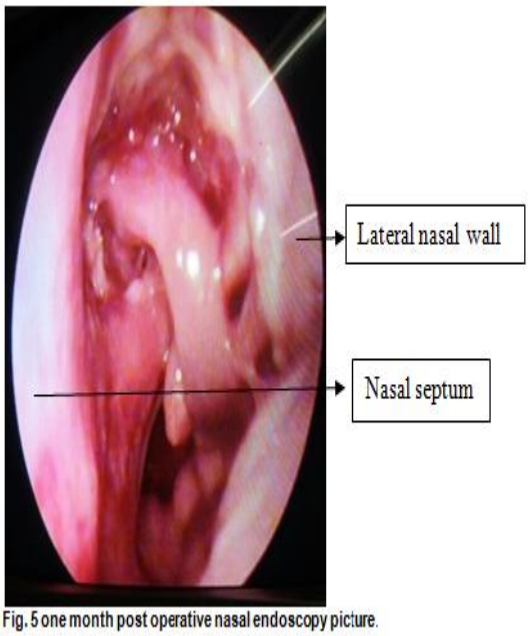

Fig. 5 one month post operative nasal endoscopy picture.

Discussion

Fibrous dysplasia is a benign pathological condition of the bone[1]. It was originally termed in 1938 by Lichtenstein[2] although McCune and Bruch first described the disease in 1937. The normal medullary bone is replaced by immature and fibrous stroma.

Fibrous dysplasia of paranasal sinuses is very rare. It mostly occurs in the long bones, Craniofacial bones are second common site of involvement. Fibrous Dysplasia of ethmoid is a rare entity and often presents a diagnostic challenge.[4] Tsai TL noted six reported cases of fibrous dysplasia confined to ethmoid sinus until 2003[4] in his presentation.

The disease develops in adolescents and young adults. In the majority of cases, fibrous dysplasia is diagnosed before the second decade of life[4], rarely after the fifth. Our patient presented in fourth decade.

Facial asymmetry is the most common presentation followed by pain, ocular proptosis, headache, sinusitis, neurological changes. In our patient none of the mentioned symptoms were present.

Fibrous dysplasia is due to abnormal osteoblastic differentiation of unknown etiology, but different hypothesis have been discussed.[3] Mutation in the gene that encodes the subunit stimulatory G protein(Gs) located at 20q13 is proposed as an etiology.[9] They are divided as Monostotic if one bone is involoved and polyostotic when more bones are affected. McCune-Albright syndrome presents as a combination of polyostotic fibrous dysplasia, skin hyperpigmentation and endocrine dysfunction which occurs in one of 30-40 cases of fibrous dysplasia. The skull is involved in about 15 percent with the majority being monostotic. The maxilla and mandible are most frequently involved[5] in the head–neck zone. Malignant transformation is rare (0.5%) and is usually seen only in polyostotic cases[4]

Fibrous dysplasia on radiology will have groundglass[6], not so well circumscribed lesion and histopathology will show bony trabeculae not lined by osteoblasts[10] having only spicules of woven bone among highly cellular connective tissue as seen in our patient.

Fibrous dysplasia and ossifying fibroma are encompassed within the term FIBRO-OSSEOUS LESION[12,13]. There is difficulty in differentiating fibrous dysplasia from ossifying fibroma. Ossifying fibroma on radiology will show egg shell rim[6] with sharply circumscribed lesion and histopathologically appear with bony trabeculae lined by osteoblasts and there is both lamellar and woven bone among the connective tissue[10]

Surgery of fibrous dysplasia is indicated if the patient is symptomatic causing loss of function and cosmetic deformities. Our patient had nasal obstruction with loss of smell.

Open surgical techniques like external ethmoidectomy and endoscopic removal are commonly employed[4]. Endoscopic sinus surgery techniques are safe as we did in our case and avoid serious complications that shall be caused by radical surgery.

Conclusion

Fibrous dysplasia is a rare disease in ethmoid sinus. Conservative surgery for symptomatic patients are indicated and usually it is self limiting needing no further intervention in monostotic patients.

References

1. Hyams VJ , Batsa kis JG, Michaelis L. Tumors of the upper respiratory tract and ear. Atlas of tumor pathology. Second series, Fascicle 25. Washington DC: Armed Forces Institute of Pathology, 1 988.

2. Lichtenstein L. Polyostotic fibrous dysplasia. Arch Surg 1938; 36: 874–898.

3. Abdel-Wanis M, Tsuch iya H. Melatonin deficiency and fibrous dysplasia : IVI ight a relation exist? Medical Hypotheses. 2002 ; 59 : 552.

4. Tsai TL, Ho CY, Guo YC, et al. Fibrous dysplasia of the ethmoid sinus. J Chin Med Assoc 2003; 66: 131–133.

5. Dominok GW, Knoch HG. Knochengesch wuelste undgeschwulstaehnliche ochenerkrankungen, 3 rd edn. Jen a :V E B Gustav Fischer, 1982.

6. Engelbrecht V, Preis S, Hassler W, et al. CT and MRI of congenital sinonasal ossifying fibroma. Neuroradiology 1999; 41: 526–529.

7. Som PM, B ra ndwein IVI. Sinonasal cavities: Inflammatory diseases, tumors, fractures, a n d postoperative findings. I n :Som PM, Ca rton H D (eds). Head and neck imaging, 3rd ed n. St Lou is: Mosby-Year Book, 1 996: 233-43.

8. Jaffe H L Fibrous dysplasia of the bone. Bulletin of the New York Academy of Medicine. 1 946; 2 2 : 588-604

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9. Diaz A, Danon M, Crawford J. McCune-Albright syndrome and disorders due to activating mutations of GNAS1. J Pediatr Endocrinol Metab. 2007;20(8): 853–880

10. FuYao-Shi, Perzin Karl H. (1974) : Non-epithelial tumors of the Nasal cavity, Paranasal sinuses and Nasopharynx : A Clinico-Pathologic study. II. Osseous and Fiobrosseous Lesions including Osteoma, Fibrous Dysplasia, Ossifying Fibroma, Osteoblastoma, Giant Cell Tumour and Osteosarcoma. Cancer 33; 1289-1305.

11. Lustig LR, Holliday MJ, McCarthy EF, et al. Fibrous dysplasia involving the skull base and temporal bone. Arch Otolaryngol Head Neck Surg 2001; 127: 1239–1247.

12. Commins DJ, Tolley NS, Milford CA. (1998) : Fibrous dysplasia and ossifying fibroma of the paranasal sinuses. J.Laryngology. Otology, 112(10) : 964-968.

13. Dornhoffer J, Schwager K. (1995) : Fibrous dysplasia and ossifying fibroma. 2 unusual fibro-osseous lesions of the paranasal sinuses. HNO, 43(3) : 193-196.

14. Von Rompaey D, Schmelzer B, Verstraete W, et al (1994) :Fibrous dysplasia in the fronto-ethmoidal complex :diagnosis and surgical aspects. Acta Otorhinolaryngology. Belgium 48(1): 37-40.

15. Pinsolle V, Rivel J, MicheletV, etal (1998) : Treatment of fibrous dysplasia of the cranio-facial bones. Report of 25 cases. Ann. Chir. Plast. Esthet. 43(3) : 234-239.