Case Report
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Craniofacial Fibrous Dysplasia: A Rare Disease

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Abstract

Introduction: Fibrous Dysplasia (FD) is a fibro-osseous lesion of the osseous structures of the body. The exact etiology is not known. Monostotic (when one bone is involved), polyostotic (when multiple bones are involved) and craniofacial are the three subtypes of craniofacial dysplasia. When the lesions are limited to contiguous bones of the craniofacial skeleton, it is termed as craniofacial dysplasia. McCune Albright syndrome is the triad of polyostotic fibrous dysplasia, Cafe aulait spots and endocrine pathology. **Case Report:** This case describes craniofacial dysplasia in a 30 year old female patient who had unusual presentation on the right side of the face involving maxilla, sphenoid, ethmoid and orbit. The clinical features, radiological findings and treatment have been discussed.

Keywords: Craniofacial Fibrous Dysplasia, Maxilla, Monostotic, Polyostotic

1. Introduction

Fibrous Dysplasia (FD) is a fibro-osseous lesion of the osseous structures of the body. The exact etiology is not known. Monostotic (when one bone is involved)], polyostotic (when multiple bones are involved) and craniofacial are the three subtypes of craniofacial dysplasia. When the lesions are limited to contiguous bones of the craniofacial skeleton, it is termed as craniofacial dysplasia. Mc Cune Albright syndrome is the triad of polyostotic fibrous dysplasia, Cafe – au lait spots and endocrine pathology.

This case describes craniofacial dysplasia in a 30 year old female patient who had unusual presentation on the right side of the face involving maxilla, sphenoid, ethmoid and orbit. The clinical features, radiological findings and treatment have been discussed.

2. Case Presentation

A 30-year-old female patient presented with the chief complaints of painless swelling of the right maxilla and right nasal obstruction since 3 months. She complained of watering from right eye since 1 month. (Figure 1). Clinically, she presented with facial asymmetry. There was no history of trauma, restricted mouth opening, decreased vision, loosening of teeth or bleeding from nose. No skin lesions or pigmentation or any endocrinal problems such as hyperthyroidism, pituitary abnormalities, renal disease (to rule out Mc Cune Albright syndrome) were noted. External examination of the face revealed well-defined bony hard swelling in the right maxillary region. There was bony remodelling involving the right orbital region which resulted in telecanthus. Intraorally, there was no

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significant abnormally detected. There was no swelling present elsewhere in the body.

On anterior rhinoscopy, pinkish mass was seen obstructing the right nasal cavity. Turbinates couldn't be appreciated. Left gross DNS was present. On nasal endoscopic examination, pinkish soft tissue mass was seen obstructing the right nasal cavity till the anterior end of the inferior turbinate with no areas of inflammation. Turbinates couldn't be appreciated. It was bleeding on touch so probing test to assess the extension of lesion couldn't be performed. Other normal structures couldn't be differentiated from the lesion since it was obstructing the right nasal cavity. Ophthalmic assessment showed no diminished vision, papilloedema. Fundus examination was normal. Extraocular movements were normal. Telecanthus was seen.

Routine investigations such as complete blood count, liver and kidney function tests, serum electrolytes, HIV, HbSAg and serum Alkaline Phosphatase (ALP) were performed. All parameters were within normal limits. The Computed Tomography (CT) scan showed diffuse ground glass expansile lesion within anterior part of clivus, sphenoid bone, right ethmoidal air cells, base of right pterygoid plate, medial wall of right orbit and right maxillary sinus. Lesion was seen protruding into the nasal cavity with gross deviation of the septum to the left side. It showed the lesion to be bulging in the right orbit, indenting over medial and inferior rectus muscle. Right optic nerve was deviated laterally. Mild proptosis if right eye globe was seen (Figure 1). CT scan was suggestive of fibrous dysplasia.

Based on the clinical history and radiographic assessment a diagnosis of craniofacial Fibrous Dysplasia was made. Full anaesthetic check-up was done and patient was posted for endoscopic sinus surgery with resection of the fibrous osseous lesion. Mass was seen engulfing the inferior, middle and superior turbinate with erosion of lateral nasal wall. Debulking of the lesions was done along with the removal of inferior turbinate, middle turbinate and superior turbinate. All the sinuses were wide opened. Ophthalmology opinion was taken again in the operation theatre and the intra operative findings were studied. Since there were no ocular symptoms conservative management with regular follow up was recommended. Haemostasis was achieved throughout the procedure with the bipolar cautery. Surgery was performed with the conventional cold steel nasal instruments endoscopically.

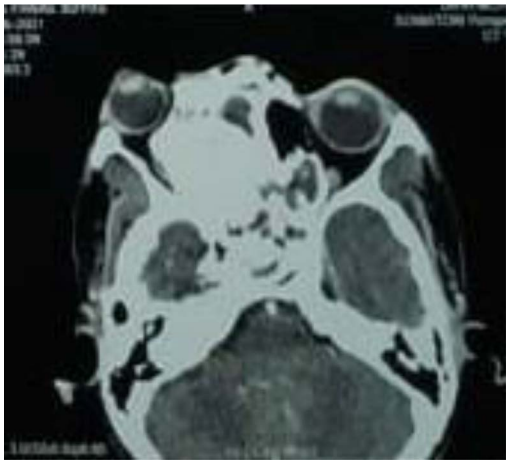

Figure 1. CT scan image.

Microdebrider, coblator and osteotomes were not needed in the surgery as the tissue was friable enough to be removed with instruments and good haemostatis was achieved throughout the procedure. Histopathological examination of the tissue sent was suggestive of fibrous dysplasia. Post operatively, nasal obstruction was completely relieved with no other significant morbidity and no development of visual symptoms were seen. After 3 months of follow up, no recurrence was seen.

Due to difficulty to access the areas surgically and difficulty in reconstructing the defects in this region, conservative management was decided in our case. Also regular follow up was recommended for the orbital involvement as patient had no orbital complaints at that time.

3. Discussion

Fibrous Dysplasia (FD) is a rare uncommon nonhereditary, developmental anomaly of skeletal structure. In this, cellular fibrous connective tissue proliferates the normal bone. This was then intermixed with the irregular bony trabeculae. This mixing of fibrous and osseous tissue causes secondary bony metaplasia, with no osteoblast maturation forming new, immature and weak calcified bone. It is called as monostotic when it arises as a single lesion or polyostotic when it affects many bones forming multiple lesions. Craniofacial FD (CFD) affects craniofacial bones, mandible and maxilla, skull base and vault[1,3]. It affects children and young adults. These conditions are relatively common in females[2]. Maxilla (12%) and mandible (12%) are frequently involved bones

whereas temporal bone, ethmoid, sphenoid, frontal are uncommon. The affected bones are thickened, sclerosed and expanded.

The patients may have visual or hearing abnormalities, loosening of tooth, facial inequality and tooth displacement depending on the bone which is involved[4]. FD is relatively uncommon in the craniofacial region[8]. Frontal, sphenoid, naso-ethmoid and maxillary bones involvement may cause nasal blockage, sinus obstruction (frontal and maxillary sinus) with resultant sinusitis. Other features seen are abnormal and painful sensations in the distribution of the trigeminal nerve, eye watering, and headaches[5].

The classical radiographic feature of FD is a radiolucent, ground-glass or hazy pattern. It is indifferent from the usual radiographic appearance of normal bone[1]. The CT images showed the ground glass appearance which is most commonly seen. It can also be homogeneous dense or sclerotic and radiolucent i.e. cystic. These features characterize fibrous dysplasia[5]. Magnetic Resonance Imaging can help to assess cranial nerve and soft-tissue structures surrounding the lesion. CT and MRI are both excellent imaging techniques to assess the compression effects on the orbit, optic canals and paranasal sinuses[4]. Serum ALP is significantly elevated in FD. It is a reliable marker for predicting the prognosis[7].

There is no definite treatment for FD. Aim of surgery is to prevent pathological fracture, pain control and lower down bone deformities. Surgical treatment aims at correcting facial deformity and restoring the obliterated foramen when they cause symptoms. There are 4 treatment options available. Observation, when there are small asymptomatic lesions which are cosmetically acceptable. Medical treatment consists of Bisphosphonates and IV Pamidronate which inhibits bone resorption and bony pain[9]. Calcitonin, Vitamin D and calcium supplements have also been recommended. Surgical treatment is the mainstay and it comprises of remodelling procedure aimed at achieving good aesthetics. This can result in recurrence and so the procedure is postponed till puberty so that the disease may undergo remission. When skull base is involved with complications, radical resection and reconstruction is recommended[8]. Wait-and-watch policy is indicated if the lesions are stable and stops to grow after the puberty. Reconstructive techniques are applied to achieve adequate aesthetical and functional results. Radical resection is done in aggressive lesions but in pediatric patients bone shaving is tried initially. Aggressive treatment is done in relapse or after skeletal maturity is achieved[8]. Most cases of FD can be treated conservatively. The surgery is indicated after active growth and maturity of bones and a regular follow-up is needed to check for recurrence.

4. Conclusion

Fibrous Dysplasia affecting craniofacial region is uncommon and in most cases can be treated conservatively. Long term follow up of patients is important considering the possible flare up of progressive growth of lesion.

5. References

1. Lietman SA, Levine MA. Fibrous dysplasia. Pediatr Endocrinol Rev. 2013; 2:389–96.
2. Menon S, Venkatswamy S, Ramu V, Banu K, Ehtaih S, Kashyap VM, et al. Craniofacial fibrous dysplasia: Surgery and literature review. Ann Maxillofac Surg. 2013; 3:66–71. PMid: 23662263 PMCid: PMC3645615. https://doi.org/10.4103/2231-0746.110088
3. Abdelkarim A, Green R, Startzell J, Preece J. Craniofacial polyostotic fibrous dysplasia: A case report and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2008; 106:e49–55. PMid: 18585612. https://doi.org/10.1016/j.tripleo.2008.03.023
4. Guruprasad Y, Prabhakar C. Craniofacial polyostotic fibrous dysplasia. Contemp Clin Dent. 2010; 1:177–9. PMid: 22114411 PMCid: PMC3220106. https://doi.org/10.4103/0976-237X.72787
5. Ziadi S, Trimeche M, Mokni M, Sriha B, Khochtali H, Korbi S. Eighteen cases of craniofacial fibrous dysplasia. Rev Stomatol Chir Maxillofac. 2009; 110:318–22. PMid: 19615708. https://doi.org/10.1016/j.stomax.2008.11.005
6. Deshpande A, Naidu GS, Dara BG, Gupta M. Craniofacial fibrous dysplasia: A summary of findings with radiological emphasis. J Indian Acad Oral Med Radiol. 2016; 28:403–8. https://doi.org/10.4103/0972-1363.200631
7. Park BY, Cheon YW, Kim YO, Pae NS, Lee WJ. Prognosis for craniofacial Fibrous Dysplasia after incomplete resection: Age and serum alkaline phosphatase. Int J Oral Maxillofac Surg. 2010; 39:221–6. PMid: 20083386. https://doi.org/10.1016/j.ijom.2009.12.008
8. Valentini V, Cassoni A, Terenzi V, Della Monaca M, Fadda MT, Rajabtork Zadeh O, et al. Our experience in the surgical management of craniofacial Fibrous Dysplasia: What has changed in the last 10 years? Acta Otorhinolaryngol Ital.

2017; 37:436–43. PMid: 29165438 PMCid: PMC5720872. https://doi.org/10.14639/0392-100X-1081

9. Ozdemir Kutbay N, Sarer Yurekli B, Kartal Baykan E, Sahin S, Saygili F. Characteristics and treatment results of 5 patients with Fibrous Dysplasia and review of the literature. Case Rep Endocrinol 2015. 2015. 670809. PMid: 26171261 PMCid: PMC4480246. https://doi.org/10.1155/2015/670809

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