Craniofacial Fibrous Dysplasia: Study of 23 Cases

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Abstract: Objectives: Despite recent advances in the understanding of the natural history and molecular abnormalities, many questions remain surrounding the progression and management of fibrous dysplasia. We aimed to describe the clinical, radiological findings and management in a consecutive series of patients diagnosed with fibrous dysplasia of the craniofacial bones. **Patients and methods**: A prospective and retrospective analysis of collected data for23patients with histopathologically confirmed fibrous dysplasia involving the skull and facial bones managed at the Maxillofacial and Neurosurgical department between February 2011-June 2012. The demographic data, clinical presentation, radiographic characteristics, and the management of these patients were reviewed. **Result**: Age of patients ranged from 7-55 years with higher predilection in female (16 patients, 70%). In the current study, the most common affected fascial bones were maxilla (7 patients), mandible (3patients). As regarding the cranial bones, the frontal and temporal bones were affected in 5 patients. Sphenoidal bone was affected in 3 patients. Most of the patients presented by facial painless swelling of deformity (17 patients) while 6patients presented by swelling and proptosis. Conservative surgical procedures (shaving) were performed in 15 cases while resection and reconstruction were performed in 8 cases. Reconstruction with mesh was used in 7 patients while reconstruct with bone graft was used in one patient**.** All patients were followed up for more than 6 months, by clinical examination, x ray and CT scanning. The frequency of follow up was tailored according disease aggressiveness, optic nerve involvement and type of intervention performed. Marked improvement in the aesthetic aspect in most cases.3 of the patients who were treated by shaving required further creation, and one patient required 3sittings to reach acceptable results. **Conclusions**: Each patient may present with variable symptoms and clinical findings, thus the care of these patients must be customized to their needs and sites of involvement.

1. Introduction

Fibrous dysplasia is a benign dysplastic process of bone in which the normal bone matrix is replaced by fibroblastic proliferation. It may occur within a single bone (monostotic) or multiple bones (polyostotic)[1,2,3]. When polyostotic fibro-osseous lesions areassociated withother anomalies andendocrinopathy, this variant form constitutes the McCune-Albright syndrome (MAS)[4].

Fibro-osseous dysplasiaaccounts for around 7% of all benign boneneoplasms[5].Craniofacial involvement occurring in all of the severe forms and about 30% of the monostotic forms[6]. However, the distinction between monostotic and polyostotic forms can be verydifficult becauseof the intimate connection of the individual craniofacial bones and the fact that the disease does not appear to be limited by the suture lines[7].

Most craniofacial lesions are monostotic, yet when a diagnosis of fibrous dysplasia is rendered a skeletal imaging survey must be ordered to search out lesions in other bones. Physical examination must be performed to uncover accompaniments that support the diagnosis of McCune-Albright syndrome[8-11]. (8-11).

Monotonic fibrous dysplasia of the craniofacial complex is often confused with other benign fibrous osseous lesion, typically ossifying fibroma and diffuse sclerosing osteomyelitis of the mandible, diseases that manifest unique clinic-radiologic features. Monostotic fibrous dysplasia occurs in the jaw, frontal, ethmoidal, temporal and calvarias bones[1,2,3].

Any cranial or facial bone can be affected by fibrous dysplasia and the clinical associated features will depend upon the bone or bones affected. Signs and symptoms can include facial pain, headache, cranial asymmetry, facial deformity, tooth displacement, and visual or auditory impairment[12,13].

Malignant change is rare, roughly 0.5% for the monostotic form and 4% for McCune–Albright syndrome[14]. Osteosarcoma was the most common, followed by fibrosarcoma, and chondrosarcoma. Malignant transformations were found mostly in the third or fourth decade of life. It is important to note that 28% of these transformations were in patients who had had fibrous dysplasia lesions radiated[15].

The clinical behavior and progression of FD vary, thereby making the management of this condition difficult with few established clinical guidelines. This paper provides a clinically-focused comprehensive description of craniofacial FD, its natural progression, the components of the diagnostic evaluation and the multi-disciplinary management.

Table (1) patient demography, site, clinical presentation, and treatment options in 23 patients with fibrous dysplasia

Patient n	Sex	Age	Site	Clinical presentation	Treatment
1	Male	15 years	Rtzygoma	Painless swelling	Shaving
2	Female	25 years	Rt maxilla	Painless swelling	Shaving
3	Female	16 years	Lt mandible	Painless swelling	Shaving
4	Female	7 years	Lt maxilla	Painless swelling	Shaving
5	Female	43 years	Rt temporal and fronto-orbital	Painless swelling proptosis	resection + reconstruct with mesh
6	Female	47 years	Rt temporal and fronto-orbital	Painless swelling proptosis	resection + reconstruct with mesh
7	Female	33 years	Ltzygoma	Painless swelling	Shaving
8	Female	20 years	Rtzygoma and maxilla	Painless swelling	Shaving
9	Female	30 years	Rtfronto-orbital	Painless swelling proptosis	resection+ reconstruct with mesh
10	Male	19 years	Pan facial	Facial deformity	Shaving
11	Male	18 years	Rt maxilla	Painless swelling	Shaving
12	Male	7 years	Lt mandible	Painless swelling	Shaving
13	Male	25 years	Rt maxilla	Painless swelling	Shaving
14	Female	36 years	Rt temporal and fronto-orbital	Painless swelling	resection+ reconstruction by bone graft
15	Male	7 years	Rt maxilla	Painless swelling	Shaving
16	Female	22 years	Rt sphenoidal ridge	Painless swelling proptosis	resection+ reconstruct with mesh
17	Female	45 years	Lt maxilla	Painless swelling	Shaving
18	Female	43 years	Rt temporal and fronto-orbital	Painless swelling proptosis	resection + reconstruct with mesh
19	Female	27 years	Fronto-nasal	Painless swelling	Shaving
20	Male	22 years	Ltparietal	Painless swelling	Shaving
21	Female	15 years	Left zygoma	Painless swelling	Shaving
22	Female	55 years	Rt sphenoidal ridge	Painless swelling proptosis	resection + reconstruct with mesh
23	Female	35 years	Ltsphenoidal ridge	Painless swelling	Shaving resection + reconstruct with mesh

Patients and methods:

This study was performed at Assiut University hospital (Single Tertiary Hospital) with 23 patients of cranio-maxillofacial fibrous dysplasia managed at the Maxillofacial and Neurosurgical departments between February 2011-June 2012.All clinical records, investigations and treatment charts were reviewed and data regarding age, gender, anatomic site, clinical presentation, associated complications, radiographic evaluations including X-ray, computed tomography(CT) and magnetic resonance imaging (MRI) were obtained preoperatively and postoperatively when possible. The indication and sequence of these investigations is best tailored according to the individual situation.Treatment details were analyzed. In all patients, histopathological examination confirmed the diagnosis (table1).All patients were followed up by clinical examination, X ray and CT scanning. The frequency of follow up was tailored according disease aggressiveness, optic nerve involvement and type of intervention performed but not less than 6 months.

3. R*esults:*
Age and SEX

There were total 23 patients who sustainedcranio-maxillofacial fibrous dysplasia. There were 7 males (30%) and 16 females (70%).Theiragerange from 7 – 55 years. The majority of patients were in the second (6 cases) and third decades (6 cases) of life.

Case (1) Pre and post-operativeimage of patient has RT sphenoidal ridge FD and presented with proptosis
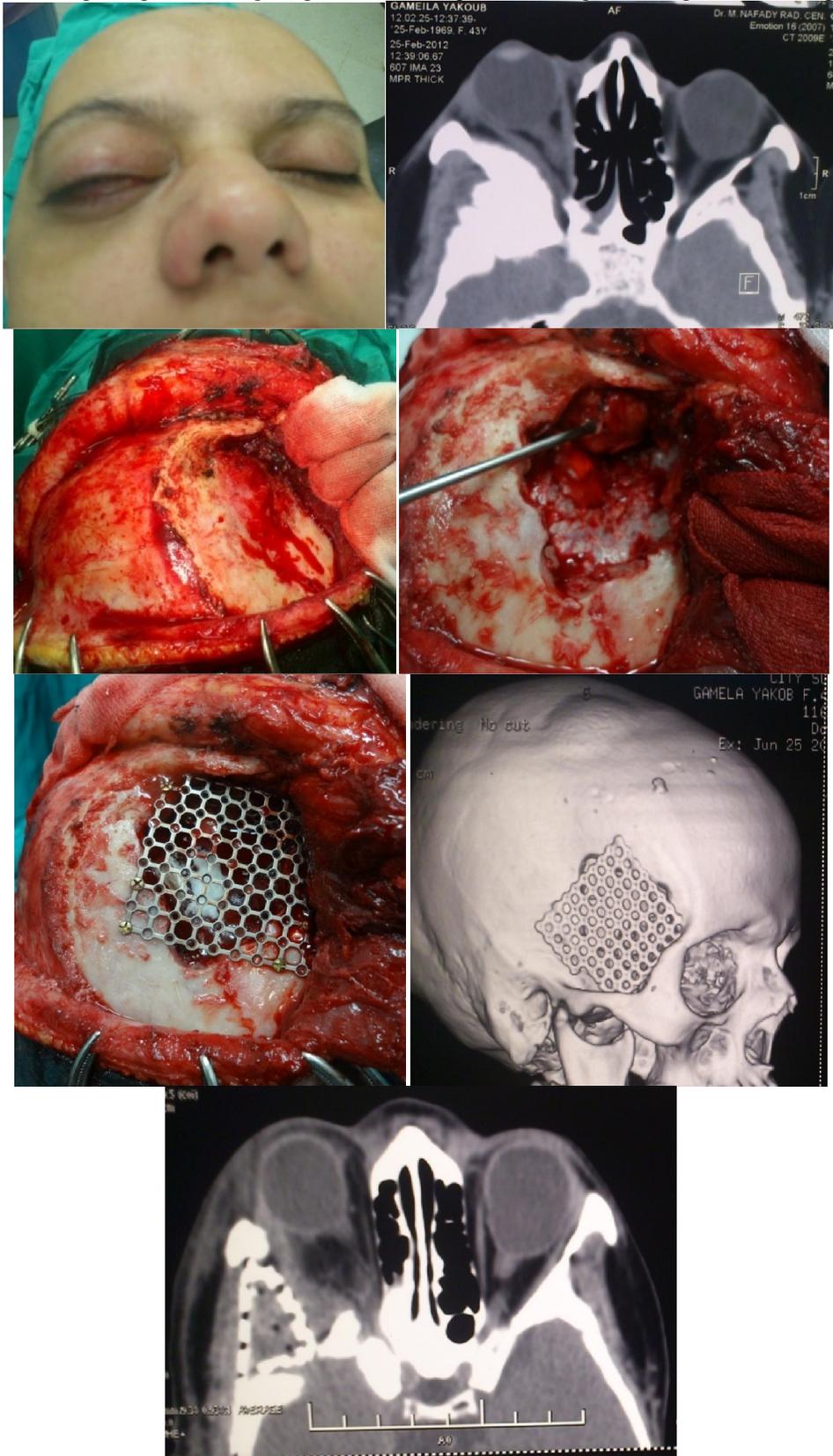
Case (2) Pre and post-operativeimage of patient has LT mandible FD and presented with swelling
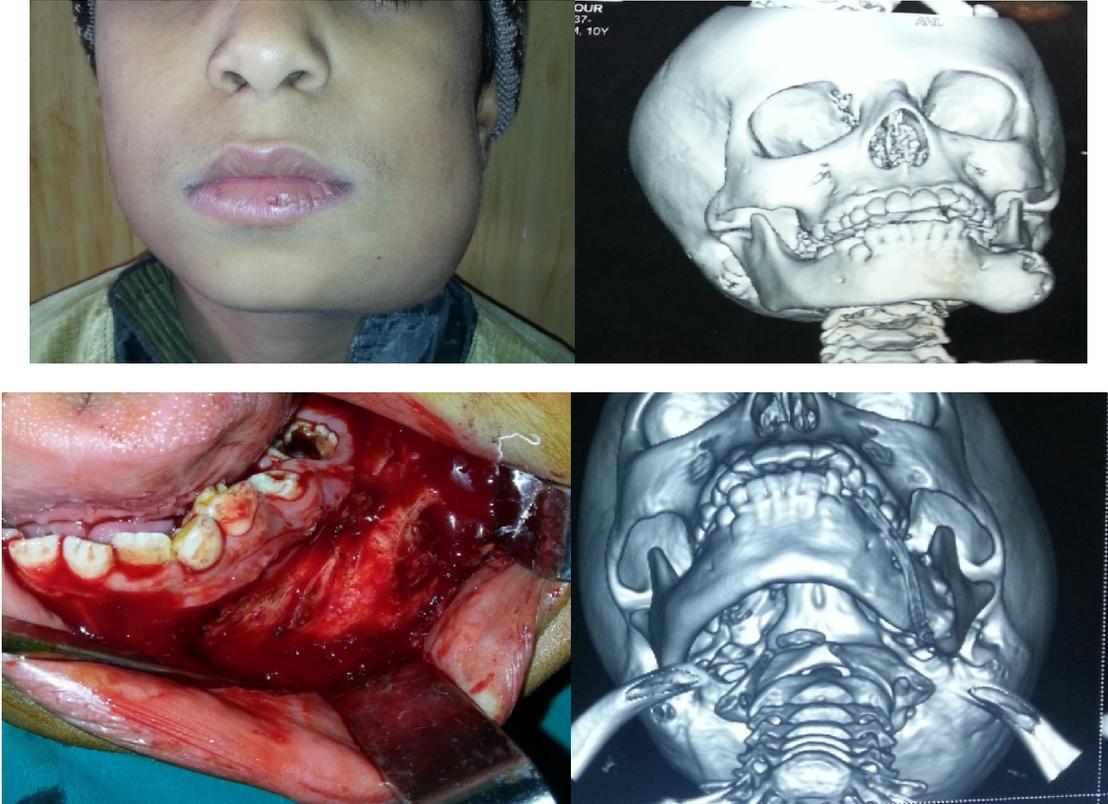
Anatomical site

In the current study, the most common affected bones were maxilla (7 patients), mandible (3 patients). As regarding the cranial bone, the frontal and temporal bones were affected in 5 patients where sphenoidal bone was affected in 3 patients.

Clinical presentation:

The site of cranial involvement in fibrous dysplasia determines the clinical manifestations. Most of them present as a painless, slowly enlarging bony mass. In the current study, most of the patients presented by facial painless swelling of deformity (17 patients) while6patients presented by swelling and proptosis.

Radiological findings

All patients submitted to skull X-ray with serial films to assess disease progress. In total, 23 patients underwent computed tomography (CT), 7 patients had three-dimensional computed tomography (3D-CT), and 10 patients underwent magnetic resonance imaging (MRI). For patients in our series, plain film X-rays were usually sufficient to diagnose most cases of fibrous dysplasia. Radiographic findings demonstrated the classic ground-glass appearance on standard X-ray films. CT findings of the 23 patientsin this study also showed characteristics of fibrous dysplasia (pagetoid, cystic or radiolucent and sclerotic).

Histological finding

Histological confirmation of the disease was performed in all cases that underwent surgery. No case showed malignant changes over the period of observation.

Treatment, and follow up

Conservative surgical procedures (shaving) were performed in 15 cases while resection and reconstruction were performed in 8 cases. Reconstruction with mesh was used in 7 patients while reconstruct with bone graft was used in one patient. All patients were followed up for more than 6 months. Marked improvement in the aesthetic aspect in most cases. 3 of the patients that were treated by shaving required further operation, and one patient required 3 sittings to reach acceptable results. Patient treated by resection showed improvement of proptosis and saving of vision.

4. Discussion

Fibrous dysplasia is a developmental, benign anomaly of bone development occurring in single or multiple bones characterized by the replacement of normal bone by fibro- osseous tissue.

Fibrous dysplasia can be divided into subtypes; roughly 70% are monostotic, and roughly 30% are polyostotic. It is also found in McCune–Albright syndrome (an association of fibrous dysplasia), precocious puberty, endocrine abnormalities, and pigmented cutaneous lesions in female patients. There seems to be no transition from one form to the other. Fries found that the skull was involved in 72% with a polyostotic form, and in 28% with a monostotic form[16]. The fronto-orbital region was affected in 20% of patients[17].

Age and sex

Craniofacial FD typically presents at around 10 years of age and then progresses throughout adolescence[7]. The disease was initially thought to become inactive after childhood but subsequent reports have proved this to be untrue[8]. In our study, age of patients range from 7 – 55 years. The majority of patients were in the second and third decades of life (6 cases for each).Gender prevalence of FD is equal however in our study there is higher predilection in female (16 patients, 70%).

Anatomical site

There is a predilection for membranous bones. In order of frequency, the maxilla, mandible, frontal, ethmoid, and sphenoidal bones are commonly involved whereas parietal, temporal and occipital bones are less commonly affected. In the current study, the most common affected bones were maxilla (7 patients), mandible (3patients). As regarding the cranial bone, the frontal and temporal bones were affected in 5 patients where sphenoidal bone was affected in 3 patients.

Clinical presentation

The clinical presentation depends on the site, duration, extent and nature of the lesion. It ranges from a mild local swelling with little or no pain to a gross deformity with complications such as proptosis, visual disturbance and sensorineural hearing loss[18].

The ocular effects of craniofacial FD are of particular concern. It may cause globe displacement due to the involvement of the sphenoid and/or ethmoid bones. Loss of vision may occur secondary to involvement of the sphenoid bone that compresses the optic nerve. Hence, in cases of optic canal involvement, a complete ophthalmological assessment, including testing for visual acuity, visual field, color perception and visual evoked potential, is necessary. In the current study, most of the patients presented by facial painless swelling of deformity (17 patients) while 6 patients presented by swelling and proptosis.

Radiographic features

Radiographic features vary depending upon the stage of the disease, degree of ossification (the proportion of bone and fibrous tissue in the lesion).

Early onset lesions are radiolucent and later progressively calcify, culminating in a ''ground glass'' or mottled mixed radiolucent/radiopaque pattern. Critical to the diagnosis is the fact that fibrous dysplasia fails to manifest any discrete margins; rather, the lesional bone subtly blends into the surrounding normal appearing bone[19].

The ability of CT to resolve detail of both soft tissue and dense bony structures makes it a very suitable imaging modality for the assessment of craniofacial fibrous dysplasia. Computed tomography (CT) is a better radiological tool, especially for assessing the extent of the tumor in cases of suspected optic canal involvement[20]. FD has characteristic appearances on CT and consists of three varieties: ground-glass pattern (56%), homogeneously dense pattern (23%) and cystic variety (21%)[21]. Various studies have suggested the use of magnetic resonance imaging (MRI) as a diagnostic tool for FD[22,23].

Radionuclide scans, such as bone scintigraphy, have some role in the diagnosis/evaluation of FD[24].Radionuclide scan has high sensitivity but low specificity. Single photon emission computed tomography (SPECT) has been reported to be more sensitive in detecting the areas involved in cases of FD[25].

Management

The management of the condition has undoubtedly become a subject of challenging complexity. Opinions about treatment range from various surgical methods to medical treatment such as pamidronate given intravenously,[26,27] but it is clear that a conservative approach may not be suitable in all cases of craniofacial fibrous dysplasia[28.] Fibrous dysplastic lesions that are not symptomatic, that do not progress, and that do not cause deformities our functional impairment should simply be monitored.

Non-operativetreatments have a role in the management of craniofacial FD. Some authors have reported their experience with the use of steroids, mainly in the treatment of visual symptoms from optic nerve compression[29-31]. Another line of medical treatment is the bisphosphonates, for example pamidronate. This group of drugs inhibits osteoclastic activity. Most experiences have been in patients with polyostotic FD or McCune-Albright Syndrome; there is limited data on patients with craniofacial FD[32].

Unfortunately there are no objective methods to assess or predict the outcome of medical treatment. Subjective criteria have been suggested, such as a decrease in inflammatory symptoms like pain and swelling[32]. Serum alkaline phosphatase, a marker for bone turnover, is consistently reduced in patients treated with pamidronate, making it a good monitor of response to medical treatment[33-35].The use of urinary hydroxyproline as a marker has also been suggested,

although experience with it is more limited. Serial radiographs have been used to assess response but results are not consistent[35]. Local bone mineral density has been found be more consistent than serial X-rays in the monitoring of response to treatment[32].

The difficulties in establishingthe surgical indications result from the benignnature of the lesionand the unpredictable natural history. These lesions can behave in a reactive, dysplastic or neoplastic manner[36].There may be frequently sudden and irreversible symptoms that complicate the planning of surgical treatment beside the disease extent, especially with regard to involvement of the skull base[37].

Adilemma exists about the wisdom of early operative intervention. Resection may clear the disease but the impact of surgery on the maturing craniofacial skeleton must be considered[38].An incomplete extenteration of the lesion may result in up to a 25% local recurrence rate[39]. However, a more radical clearance may compound the difficulties of reconstruction. While an extended resection may help prevent the rare occurrence of malignant transformation in residual dysplastic bone, the extent to which this approach is justified remains uncertain.

Surgical treatment of FD consists of either conservative shaving/contouring or radical excision with immediate reconstruction. The choice of surgical option depends on several factors: site of involvement, rate of growth, aesthetic disturbance, functional disruption, patient preference, general health of the patient, surgeon's experience and the availability of a multi-disciplinary team (neurosurgeon, ophthalmologist, otolaryngologist, orthodontist)[40].

Surgical classification

In thinner bone, such as the orbital plate of the maxilla, ethmoid and frontal bones, the cortex expands more rapidly and to a greater degree than in thicker cortical bone[41]. When the orbit is involved compression and subsequent distortion of the globe can cause errors in refraction, focusing, and accommodation,[42]so immediate removal of the dysplastic process and decompression of the optic nerve canal is necessary. Visual loss in fibrous dysplasia is caused by the progressive compression of venous drainage of the optic nerve, which leads to reduced retinal perfusion[43]. Operations also essential when the foramen magnum is affected to prevent life-threatening conditions.

It was previously considered prudent to wait for completion of skeletal growth as this might coincide with an arrest of further disease progression[44]. However, the growth rate of normal residual tissue and perhaps, more importantly, dysplastic tissue appears unaffected by early surgical intervention[45]. In some clinical situations, early surgery may prevent the onset of irreversible symptoms, such as loss of vision. The possibility of reactivation in later life exists however, especially in pregnancy[46]. Some authors have advocated that sequential radiographic monitoring may be adequate in many cases, with limited surgery for symptomaticcomplications[47]. Clearly these varied perspectives call for clinical judgment in any given case.

We found the classification proposed by Chen and Noordhof[48]to be of practical value, in planning our surgical approach (Table 2).

Zone 1	Frontal, orbital, nasal, ethmoid, zygoma, upper maxilla, (facial involvement amenable to surgery).
Zone 2	Parietal, part of occipital, temporal (lateral cranial base) (Hair covered cranium).
Zone 3	Central cranial base, petrous, mastoid, pterygoid, sphenoid (difficult or dangerous).
Zone 4	Maxillary alveolar bone, mandible (teeth bearing).

For lesions in zone 1, total excision of the dysplastic bone is recommended. For lesions in zones 2, 3 and 4, conservative excision or shaving has been proposed. In the current study, conservative surgical procedures (shaving) were performed in 15 cases while resection and reconstruction were performed in 8 cases.

Reconstruction after excision is important in the management of craniofacial FD. This is particularly true in cases of zone 1 involvement. The use of autologous tissues, namely grafts of calvarial bone and rib, is preferable. Split calvarial grafts are usually obtained from the frontal, temporal or parietal regions. As these bones have diploe between the inner and outer cortices they are easily split. The inner cortex is

used as the graft while the outer cortex is placed back to its original position. Rigid fixation is achieved with mini or microplates[40].

Ribgrafts are also frequently used in a split fashion. One reconstructive technique is the "chainlink fence" technique, useful for the reconstruction of large defects especially in the fronto-orbital region, particularly when calvarial bone graft is not available[49]. Full-thickness rib grafts are useful for the reconstruction of the superior orbital rim as they are effective in re-establishing rim contour[40]. We used it in one patient after excision of fronto-temporal FD.

Micro vascular free flap reconstruction has a role, especially for lesions involving the mandible where segmental excision is necessary[40]. Orthognathic surgery may be necessary in some patients with zone 4 lesions, as these patients have been found to have higher rates of mal-occlusion[51]. orthognathic surgery helps to restore stable occlusion and good facial aesthetics. Results after orthognathic surgery have been maintained long-term, without cases of recurrence after surgery. This demonstrates that fibrodysplastic bone is capable of healing adequately using standard methods of fixation[52]. Routine dental therapies, including orthodontic treatment, have been found not to exacerbate the disease[51]. In our study, reconstruction with mesh was used in 7 patients while reconstruct with bone graft was used in one patient.

Malignant transformation of fibrous dysplasia occurs infrequently, with reported frequency ranged from 0.4 to 4%. None of our cases demonstrated malignant change over the period of observation.

Conclusion

FD, a disorder characterized by the replacement of bone with fibro-osseous tissue with immature woven bones. It results in skeletal distortion with cosmetic and functional problems. The extent and location of the disease must be carefully assessed in each patient, and treatment plan individualized accordingly.

References

1. Waldron CA, Giansanti JS. Benign fibro-osseous lesions of the jaws: a clinical-radiologic-histologic review of sixty-five cases. Oral Surg Oral Med Oral Pathol. 1973;35:190–201.
2. Eversole LR, Sabes WR, Rovin S. Fibrous dysplasia: a nosologic problem in the diagnosis of fibro-osseous lesions of the jaws. J Oral Pathol. 1972;1:189–220.
3. Brannon RB, Fowler CB. Benign fibro-osseous lesions: a review of current concepts. Adv Anat Pathol. 2001;8:126–43.
4. Cohen MM Jr. The new bone biology: pathologic, molecular, clinical correlates. Am J Med Genet A. 2006;140:2646–706.
5. Derome, P.S. and Visot, A. (1983) La dysplasia fibreuse cranienne. Neuro-Chir. 29 (suppl l), l-117.
6. Fries, J.W. (1957) The roentgen features of fibrous dysplasia of the skull and facial bones. A critical analysis of thirty-nine pathologically proven cases. Am. J. Roentgenol. 77, 71.
7. Lello, G.R. and Sparrow, O.C. (1985) Craniofacialpolyostotic fibrous dysplasia. J. Maxillofac. Surg. 13, 267.
8. Lietman SA, Ding C, Levine MA. A highly sensitive polymerase chain reaction method detects activating mutations of the GNAS gene in peripheral blood cells in McCune-Albright syndrome or isolated fibrous dysplasia. J Bone Joint Surg Am.2005;87:2489–94.
9. Weinstein LS, Liu J, Sakamoto A, Xie T, Chen M. Minireview: GNAS: normal and abnormal functions. Endocrinology. 2004;145:5459–64.
10. Perdigao PF, Pimenta FJ, Castro WH, De Marco L, Gomez RS. Investigation of the GS alpha gene in the diagnosis of fibrous dysplasia. Int J Oral Maxillofac Surg. 2004;33:498–501.
11. Lumbroso S, Paris F, Sultan C, European Collaborative Study. Activating Gsalpha mutations: analysis of 113 patients with signs of McCune-Albright syndrome–a European collaborative study. J Clin Endocrinol Metab. 2004;89:2107–13.
12. Alam Wg, Cdr A, Chander Gp, Capt BN: Craniofacial fibrous dysplasia presenting with visual impairment. MJAFI 2003,59:342-343.
13. Lustig LR, Holliday MJ, McCarthy EF, Nager GT: Fibrous dysplasia involving the skull base and temporal bone. Arch Otolaryngol Head Neck Surg 2001, 127:1239-1247.
14. Hoshi M, Matsumoto S, Manabe J, Tanizawa T, Shigemitsu T, Izawa N, et al. Malignant change secondary to fibrous dysplasia. Int J Clin Oncol 2006;11(3):229–35.
15. Yabut Jr SM, Kenan S, Sissons HA, Lewis MM. Malignant transformation of fibrous dysplasia. A case report and review of the literature. Clin Orthop Relat Res 1988;(228):281–9.
16. Fries JW. The roentgen features of fibrous dysplasia of the skull and facial bones; a critical analysis of thirty-nine pathologically proved cases. Am J Roentgenol Radium Ther Nucl Med 1957;77:71–88.

17. Jackson IT, Hide TA, Gomuwka PK, Laws Jr ER, Langford K. Treatment of cranio-orbital fibrous dysplasia. J Maxillofac Surg 1982;10:138–41.

18. Smith-Kirwin SM, Funanage VL. Glucocorticoids decrease interleukin-6 levels and induce mineralization of cultured osteogenic cells from children with fibrous dysplasia. J Bone Miner Res 1999;14:1104-14.

19. Roy Eversole Æ Lan Su Æ Samir ElMofty Benign Fibro-Osseous Lesions of the Craniofacial Complex, A Review, Head and Neck Pathol (2008) 2:177–202

20. Chen YR, Wong FH, Hsueh C, Lo LJ. Computed tomography characteristics of non-syndromic craniofacial fibrous dysplasia. Chang Gung Med J 2002;25:1-8.

21. Brown EW, Megerian CA, McKenna MJ, Weber A. Fibrous dysplasia of the temporal bone: imaging findings. AJR Am J Roentgenol 1995;164:679-82.

22. Yano M, Tajima S, Tanaka Y, Imai K, Umebayashi M. Magnetic resonance imaging findings of craniofacial fibrous dysplasia. Ann Plast Surg 1993;30:371-4.

23. Jee WH, Choi KH, Choe BY, Park JM, Shinn KS. Fibrous dysplasia: MR imaging characteristics with radiopathologic correlation. AJR Am J Roentgenol 1996;167:1523-7.

24. Zhibin Y, Quanyong L, Libo C, Jun Z, Hankui L, Jifang Z, Ruisen Z. The role of radionuclide bone scintigraphy in fibrous dysplasia of bone. ClinNucl Med 2004;29:177- 80.

25. Nakahara T, Fujii H, Hashimoto J, Kubo A. Use of bone SPECT in the evaluation of fibrous dysplasia of the skull. ClinNucl Med 2004;29:554-9.

26. Harrison, D.F.N. (1984) Osseous and non osseous conditions affecting the craniofacial bones. Ann. Otol. Rhinol. Laryngol. 93, 199-203.

27. Higashi, T., Iguchi, M., Shimura, A. and Kruglik, G.D. (1980) Computed tomography and bone scintigraphy in polyostotic fibrous dysplasia. Report of a case. Oral Surg. 50, 580.

28. Camilleri, A.E. (1991) Craniofacial fibrous dysplasia. J. Laryngol. Otol. 105(8), 662-666.

29. Osguthorpe JD, Gudeman SK. Orbital complications of fibrous dysplasia. Otolaryngol Head Neck Surg 1987;97:403-5.

30. Weisman JS, Hepler RS, Vinters HV. Reversible visual loss caused by fibrous dysplasia. Am JOphthalmol 1990;110:244-9.

31. Bland LI, Marchese MJ, McDonald JV. Acute monocular blindness secondary to fibrous dysplasia of the skull: a case report. Ann Ophthalmol 1992;24:263-6.

32. Kos M, Luczak K Godzinski J, Klempous J. Treatment of monostotic fibrous dysplasia with pamidronate. J Craniomaxillofac Surg 2004;32:10-5.

33. Collins MT, Kushner H, Reynolds JC, Chebli C, Kelly MH, Gupta A, Brillante B, Leet AI, Riminucci M, Robey PG, Bianco P, Wientroub S, Chen CC. An instrument to measure skeletal burden and predict functional outcome in fibrous dysplasia of bone. J Bone Miner Res 2005;20:219-26.

34. Chapurlat RD, Hugueny P, Delmas PD, Meunier PJ. Treatment of fibrous dysplasia of bone with intravenous pamidronate: long-term effectiveness and evaluation of predictors of response to treatment. Bone 2004;35:235-42.

35. Plotkin H, Rauch F, Zeitlin L, Munns C, Travers R, Glorieux FH. Effect of pamidronate treatment in children with polyostotic fibrous dysplasia of bone. J Clin Endocrinol Metab 2003;88:4569-75.

36. Eversole, L.R., Sabes, W.R. and Rovin, S. (1972) Fibrous dysplasia: A nosologic problem in the diagnosis of fibrous lesions of the jaws. J. Oral Pathol. 1, 189-220.

37. Derome, P.S. and Visot, A. (1983) La dysplasia fibreuse cranienne. Neuro-Chir. 29 (suppl l), l-117.

38. Di Rocco, C., Marchese, E. and Velardi, F. (1992) Fibrous dysplasia of the skull in children, Paediatr. Neurosurg. 18, 117-126.

39. Jackson, I.T., Hide, T.A.H., Gomuwka, P.K., Laws, E.R. and Langford, K. (1982) Treatment of crania-orbital fibrous dysplasia. J. Maxillofac. Surg. 10, 138.

40. Chen YR, Kao CC. Craniofacial Tumors and Fibrous Dysplasia. In: Achauer BM, Eriksson E, Guyuron B, Coleman JJ III, Russell RC, Vander Kolk CA, eds. Plastic Surgery: Indications, Operations, and Outcomes, Vol 2: Craniomaxillofacial, Cleft, and Pediatric Surgery. St Louis: Mosby, 2000:729-39.

41. Ricalde P, Horswell BB. Craniofacial fibrous dysplasia of the fronto- orbital region: a case series and literature review. J Oral Maxillofac Surg 2001;59:157–68.

42. Goisis M, Biglioli F, Guareschi M, Frigerio A, Mortini P. Fibrous dysplasia of the orbital region: current clinical perspectives in ophthalmology and cranio-maxillofacial surgery. Ophthal Plast Reconstr Surg 2006;22:383–7.

43. Bland LI, Marchese MJ, McDonald JV. Acute monocular blindness secondary to fibrous dysplasia of the skull: a case report. Ann Ophthalmol 1992;24:263–6.

44. Harrison, D.F.N. (1984) Osseous and non osseous conditions affecting the craniofacial bones. Ann. Otol. Rhinol. Laryngol. 93, 199-203.

45. Van Horn, P.E. Jr., Dahlin, D.C. and Bickel, W.H. (1963) Fibrous dysplasia: a clinical study of orthopaedic cases. Proc. Mayo Clin. 38: 175-189.

46. Hall, M.B., Sclar, A.G. and Garner, D.F. (1984) Albright's syndrome with reactivation of fibrous dysplasia secondary to pituitary adenoma and further complicated by osteogenic sarcoma. Report of a case. Oral Surg. 57, 616.

47. Higashi, T., Iguchi, M., Shimura, A. and Kruglik, G.D. (1980) Computed tomography and bone scintigraphy in polyostotic fibrous dysplasia. Report of a case. Oral Surg. 50, 580.

48. Chen, Y.R. and Noordhoff, MS. (1990) Treatment of craniomaxillofacial fibrous dysplasia: how early and how extensive? Plast. Reconstruct, Surg 86(5), 835-842; discussion 8433844.

49. Munro IR, Chen YR. Radical treatment for fronto-orbital fibrous dysplasia: the chain-link fence. Plast Reconstr Surg 1981;67:719-30.

50. Feingold RS, Argamaso RV, Strauch B. Free fibula flap mandible reconstruction for oral obstruction secondary to giant fibrous dysplasia. Plast Reconstr Surg 1996;97:196-201.

51. Akintoye SO, Lee JS, Feimster T, Booher S, Brahim J, Kingman A, Riminucci M, Robey PG, Collins MT. Dental characteristics of fibrous dysplasia and McCune-Albright syndrome. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2003;96:275-82.

52. Yeow VK, Chen YR. Orthognathic surgery in craniomaxillofacial fibrous dysplasia. J Craniofac Surg 1999;10:155-9.

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