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Fibrous Dysplasia of Skull and Facial Bones: A Hospital Based Survey Based on Computed Tomography (CT) Findings

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Abstract
Introduction: *Fibrous dysplasia is a benign disease characterised by a slow progressive replacement of normal bone elements with fibrous tissue. It represents a bone developmental disorder specially a defect in osteoblastic differentiation and maturation. There are two primary categories of the disease: monostotic fibrous dysplasia [70- 85%] that involves a single bone and polyostotic fibrous dysplasia [20- 30%] that involves multiple bones. In the head and neck region, craniofacial involvement in fibrous dysplasia occurs in nearly 100% of polyostotic and 30%of monostotic form.*
The diagnosis is mainly based on radiological findings and the modality of treatment is mainly conservative.
Aims and Objectives: *The aim of the study is to evaluate the involvement of skull and facial bone in diagnosed case of fibrous dysplasia based on CT features.*
Materials and Methods: *The study was carried out during the period of December 2013 to November 2015 (period of two year). 40 patients of fibrous dysplasia involving skull and craniofacial bones were included in the study.*
Results and Observations: *The skull and facial bones are in majority involved by polyostotic variety of fibrous dysplasia with maxilla and mandible involvement seen in majority of them.*
Conclusion: *skull and facial bone involvement is more common in polyostotic form than in monostotic form of fibrous dysplasia. maxilla and mandible involvement seen in majority of polyostotic variety. less common involved bone was lacrimal, temporal, vomer and inferior nasal concha*
Keywords: *fibrous dysplasia, temporal bone, skull, craniofacial bones, monostotic, polyostotic.*

INTRODUCTION

The term fibrous dysplasia was coined by "Lichtenstein" in 1938, although, it was first described by McCune and Bruch in 1937[1.]

Fibrous dysplasia is a benign disease characterized by a progressive replacement of normal bone elements with fibrous tissue. It tends to develop at early childhood with preference for caucasians. There are two primary categories of the disease: monostotic fibrous dysplasia that involves a single bone and polyostotic fibrous dysplasia that involves multiple bones.[2]

The disease can involve any bone in body. In the head and neck region, craniofacial involvement in fibrous dysplasia occurs in nearly 100% of polyostotic and 30% of monostotic form [3]. The bones commonly involves the mandible and maxilla, involvement of the ethmoid sphenoid and frontal and temporal bones are less common[.2]. These lesion cause expansion, thickening and sclerosis of the involved bones with resultant facial asymmetry and swelling, hearing disturbances depending on the bone involved. The diagnosis is mainly based on radiological findings and the modality of treatment is mainly conservative.

AIMS AND OBJECTIVES

The aim of the study is to evaluate the involvement of skull and facial bone in diagnosed case of fibrous dysplasia based on CT features and their relative proportion.

MATERIALS AND METHODS

The study was carried out in Department of Radiodiagnosis, Patna Medical College and Hospital, Patna, during the period of December 2013 to November 2015 (period of two year). 50 patients of fibrous dysplasia invoving craniofacial bones were included in the study. Serial CT sections of cranium was obtained in each case using fourth generation spiral CT scanner GEE Bright speed elect (16 slice). Thinner sections were taken for the temporal region and at the region of interest. Reconstructed images (Sagittal and Coronal) were generated from the data set.

RESULTS & OBSERVATIONS

The present study consists of 40 cases of fibrous dysplasia wherein the principal radiologic study was computed tomography. Although the final diagnosis was established by histopathological evaluation in many instances effort was made to predict the diagnosis on CT characteristics.

Table : 1 Age incidence in present study (n=40)

Age group (in years)	Number of cases	Percentage (%)
0 -10	4	10
11-20	16	40
21-30	10	25
31-40	6	15
41-50	2	5
>50	2	5
Total	40	100

Observation: The majority of cases were in the range of second to third decade of life, with peak incidence (40%) in age group of 11-20 years, followed by age group of 21-30 years.

Table : 2 Sex incidence in present study

Sex	Number of cases	Percentage (%)
Male	16	40
Female	24	60

Observation: The above table shows the sex incidence, where 40% (16 cases) were males and 60% (24 cases) were females. The ratio between male and female cases was 2:3 in the study.

Table : 3 Monostotic vs polyostotic cases (n=40)

Group	No of cases	Percentage(%)
Monostotic variety	14	35
Polyostotic variety	26	65

Observation: Out of these 40 cases of FD involving skull and facial bones, the polyostotic variety constituted largest category of 26 cases (65%) as shown in Table 3. The monostotic cases were lesser in the present study, constituting 14 cases (35.0%) of the total

Table : 4 Involvement of skull and facial bones in monostotic cases detected by CT (n=14).

skull and facial bones involved	Total number of cases	Approx. Percentage (%)
Frontal	4	29
Parietal	3	21
Sphenoid	2	14
Ethmoid	1	7
Temporal	1	7
Occipital	0	0
Mandible	2	14
Maxilla	1	7
Zygomatic	0	0
Nasal	0	0
Lacrimal	0	0
Vomer	0	0
Palatine	0	0
Inferior nasal concha	0	0

Observation: Table 4 further illustrates the distribution of cases of monostotic variety of fibrous dysplasia. In monostotic variety, frontal bone (29%) is most commonly involved followed by parietal bone(21%).

Table : 5 Involvement of skull and facial bones in polyostotic cases detected by CT (n=26).

skull and facial bones involved	Total number of cases	Approx. Percentage (%)
Frontal	6	23
Parietal	10	38
Sphenoid	7	27
Ethmoid	3	12
Temporal	2	8
Occipital	4	15
Mandible	20	78
Maxilla	18	70
Zygomatic	3	12
Nasal	4	15
Lacrimal	1	4
Vomer	2	8
Palatine	3	12
Inferior nasal concha	2	8

Observation: Table 5 further illustrates the distribution of cases of polyostotic variety of fibrous dysplasia. In polyostotic variety, mandible (78%) is most commonly involved closely followed by maxillary bone (70%). Least commonly involved bone was lacrimal (4%). Other less common involved bone was temporal, vomer and inferior nasal concha, all separately constituting 8% of total polyostotic cases.

DISCUSSION

Fibrous dysplasia is non familial genetic disorder. It develop as a result of abnormal growth and differentiation of marrow stromal cells[5]. It is mainly of two types: monostotic (70-85%) and polyostotic (20-30%). It may be associated with variety of syndromes. McCune Albright's syndrome consists of Polyostotic FD (typically unilateral), ipsilateral café au lait spots and endocrine disturbance, most commonly precocious puberty in girl. Mazabraud's syndrome consists of FD (most commonly polyostotic) and soft tissue myxomata.

Craniofacial involvement in fibrous dysplasia occurs in nearly 100%of polyostotic and 30%of monostotic form [3]. The bones commonly involves the mandible [12%] and maxilla [12%], involvement of the ethmoid, sphenoid and frontal and temporal bones are infrequent[4]. Involvement of external auditory canal is the most common manifestation of FD of temporal bone, occurring in approximately 85% of patient resulting in conductive hearing loss[5]. Sometimes severe involvement of craniofacial bones lead to patient's face resembling that of lion (leontiasis ossea).

CT scan is the primary mode for radiologically evaluating FD and it is the best way to display the bony changes. Plain films and MR imaging are useful adjuncts.

At present time, there is no conservative treatment to control and prevent the progression of fibrous dysplasia. The simple presence of the lesion does not justify surgical intervention[6].If followed by significant clinical symptoms the surgery is recommended. Radiotherapy should be avoided owing to high incidence of malignant transform-ation (44%)[2].

Prognosis is good in most cases, depending on disease severity. Malignant change in FD is rare, being reported in 0.5% of cases. It is more common in polyostotic disease and may follow prior radiotherapy.[2]Clinical assessment associated with periodical CT scan may be useful in follo-wing up the patient to assess disease progression and the need for further surgical interventions[7].

Limitation of the Study

Population of the study is a selected one which does not represent the general population. The subjects were assessed on one occasion only. The assessment is not blind due to study constraint therefore bias is possible. Despite its limitations, the present study confirms skull and facial bones are in majority involved by polyostotic variety of fibrous dysplasia, with maxilla and mandible involvement seen in majority of polyostotic variety.

CONCLUSION

Fibrous dysplasia is a benign pathology of unknown etiology. This disease may affect skull and facial bones, causing deformities and dysfunctions. From the present study we can conclude that the majority of cases were in the range of second to third decade of life, with slightly higher incidence in female as compared to male. The skull and facial bone involvement was more common in polyostotic form than in monostotic form of fibrous dysplasia. Maxilla and mandible involvement were seen in majority of polyostotic variety. Less common involved craniofacial bone were lacrimal, temporal, vomer and inferior nasal concha.

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