INTRODUCTION

Fibrous dysplasia was first described by Lichtenstein in 1938 as a disorder characterized by progressive replacement of normal bone element by fibrous tissue. It is a bone tumour that although benign, has the potential to cause significant cosmetic and functional disturbance particularly in craniofacial skeleton. Its management poses significant challenge to the surgeons. Its compression of the optic nerve with resulting visual impairment is especially alarming.1

Fibrous dysplasia affecting the jaws is an uncommon developmental anomaly. It may be divided into three categories, Monostotic (74%), Polyostotic (13%) and craniofacial (13%).2 The last category, identified by Davis and Yardley, appear to be confined to the face and jaws involving two or more bones.3

Although its actual incidence is not known it accounts for between 2.5 and 10% of all bone tumours, thus a relatively common disease. FD is a developmental dysplastic disorder of bone in which normal bone matrix is replaced by fibroblastic proliferation. Lesions contain irregular trabiculae of partially calcified osteoid. Some believe that the immature woven bone is formed directly from abnormal connective tissue that is unable to form mature lamellar bone, hence the term dysplasia. Others believe that there is underlying abnormal fibroblast proliferation that results in replacement of normal cancellous bone with an immature fibrous tissue that is poorly mineralized.4

Fibrous dysplasia is a sporadic condition that results from a polyzygotic mutation in the GNAS 1 (guanine nucleotide-binding protein, alpha stimulating activity polypeptide 1) gene. Clinically FD may manifest as a localized process involving only one bone, as a condition involving multiple bones, or as multiple bone lesions in conjunction with cutaneous and endocrine abnormalities. The clinical severity of the condition presumably depends on the point in time during fetal and post-natal life that the mutation of the GNAS 1 occurs.5

ABSTRACT

The study was carried out on a total of 13 biopsy confirmed fibrous dysplasia patients reported to Oral and Maxillofacial surgery Unit of Khyber College of Dentistry, Peshawar over a period of 3 years i.e., from January 2004 to December 2005. The aim of the study was to evaluate the clinical and radiographic features and management of the patients with mono-ostotic fibrous dysplasia of the jaw bones. The age range of the patients suffering from mono-ostotic fibrous dysplasia was from 10 to 35 years with the mean age of years 19.78 years SD + 7.53. There were more females (62%) as compared to males (38%), with male to female ratio of 1:1.6. The maxillary bone (69%) was more frequently affected than the mandibular (31%). While the condition has affected most of the patients in their second decade of life i.e. 53.85%. Swelling has been the most common presenting complaint (76.9%). All the cases were treated with surgical shaving or re-contouring. Two cases of recurrent fibrous dysplasia were also reported within the study period.

Key words: Fibrous dysplasia, Mono-osteotic fibrous dysplasia, Maxillofacial Fibrous Dysplasia, FD at Khyber College of Dentistry

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MATERIALS AND METHODS

The study was carried out on 13 patients of fibrous dysplasia in Oral and Maxillofacial Surgery Unit of Khyber College of Dentistry, Peshawar. The criteria of diagnosis were history taking; clinical and radiographic examination (Periapical, Ortho-pantomogram, PNS view and Computed Tomography) followed by biopsy confirmation. The age, gender and frequency of bone involvement by FD were the parameters analyzed. All these patients were managed surgically. Patients below 25 years were told about the possible recurrence. Aesthetic concern was the main indication of the surgery. Surgical re-contouring or paring was the treatment modality. Patients were followed up over a period of three months, six months and one year interval.

RESULTS

According to the present study age range of the patients with fibrous dysplasia was from 10 to 35 years with the mean age of years 19.78 years SD ± 7.53. Fibrous dysplasia has affected most of the patients in their second decade of life (53.85%), followed by third and fourth decades of life i.e., 23.07% and 15.38% respectively (Table 1). Eight patients (62%) were females while five were males (38%) with a male to female ratio of 1:1.6, which means that FD was common in females than in males (Fig 1). Out of a total of 13 patients the maxilla was involved in 9 patients (69%). While in (31%) of cases the mandible was involved (Fig 2). According to the present study 76.9% of patients had a presenting complaint of swelling and 23.9% were complaining of pain. The most consistent radiographic feature in all patients with FD was ground glass appearance on plane radiograph (Fig 4). All the cases were treated with surgical shaving or re-contouring. Two cases of recurrent (FD) were also reported within the study period.

<table>
<thead>
<tr>
<th>Age in years</th>
<th>No of Patients</th>
<th>Percentages</th>
</tr>
</thead>
<tbody>
<tr>
<td>1-10</td>
<td>1</td>
<td>7.69%</td>
</tr>
<tr>
<td>11-20</td>
<td>7</td>
<td>53.85%</td>
</tr>
<tr>
<td>21-30</td>
<td>3</td>
<td>23.07%</td>
</tr>
<tr>
<td>31-40</td>
<td>2</td>
<td>15.38%</td>
</tr>
<tr>
<td>TOTAL</td>
<td>13</td>
<td>100%</td>
</tr>
</tbody>
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TABLE 1: AGE DISTRIBUTION OF FIBROUS DYSPLASIA PATIENTS

![Fig 1. Gender distribution of fibrous dysplasia of the jaws](image1)

![Fig 2. Maxillary Vs Mandibular bone involvement in Fibrous dysplasia](image2)

![Fig 3. Note the buccal cortical expansion right maxilla](image3)

![Fig 4. Ground-glass appearance on the radiograph Note the obliteration of the right maxillary sinus](image4)
DISCUSSION

Monostotic fibrous dysplasia is characterized by a swelling resulting from a poorly circumscribed area of fibro-osseous proliferation. Prior to 1970, fibrous dysplasia was used as an all-inclusive term that encompassed both monostotic and polyostotic forms of fibrous dysplasia and a variety of other fibro-osseous lesions, notably ossifying fibroma, fibrous osteoma and osteoblastoma. Recently, the term fibro-osseous lesion has been frequently used for disorders ranging from fibrous dysplasia to the circumscribed lesions of ossifying or cementifying fibroma and cemental dysplasia. More recently, the WHO classification of fibro-osseous lesion has recognized these conditions as distinct entities.

The diagnosis of fibrous dysplasia is often made in infancy and childhood. The maxilla or mandible may be involved with equal frequency, but the predominance of the maxilla has been documented. Males are less often affected than females. The present study also support Ogunsalu (1998) study as for as the bone involvement and gender distribution is concerned.

The deformity of the jaw results from a progressively slow growing painless swelling, but growth often slows or become arrested at a time coinciding with the onset of puberty. Because of the similarity between fibrous dysplasia and ossifying fibroma at a histological level, the diagnosis of each of these fibro-osseous lesions continues to be a problem. For this reason one can correctly say that the X-ray studies are important for the interpretation of fibro-osseous lesions of the jaw, and more particularly in the differentiation between fibrous dysplasia and ossifying fibroma, which is virtually impossible on the basis of histopathology alone.

Aside from McCune-Albright syndrome, it is usually difficult to diagnose FD on clinical, radiographic or histological criteria alone; one must consider all three factors. McCune-Albright syndrome (MAS) is a sporadic disease classically defined by polyostotic fibrous dysplasia, cafe-au-lait spots, sexual precocity, and other hyperfunctional endocrinopathies.

Monostotic fibrous dysplasia starts in childhood but typically undergoes increasing ossification and subsequent arrest in adulthood. It is clinically seen mainly in young persons, usually in their 20s, as a painless, smoothly rounded swelling, usually in the Maxilla. But the present study shows that the condition is common in patients in their teen age. The mean age of occurrence in the 69 patients reported by Zimmerman and his associates was 27 years, while in the 53 patients with craniofacial fibrous dysplasia reported by Gardner and Halpert, the mean age was 34 years both of which are higher than the mean age of 19.78 years for 13 patients reported in the present study.

The craniofacial form of fibrous dysplasia can be diffuse and may involve multiple bones. When the anatomic spaces and foramina are constricted because of encroachment of the lesions, the patient may experience a variety of symptoms, including headaches, loss of vision, proptosis, diplopia, loss of hearing, anosmia, nasal obstruction, epistaxis, epiphora and symptoms mimicking sinusitis.

With initial development of fibrous dysplasia the patient usually reports facial swellings and asymmetries. Although the lesion is usually asymptomatic, encroachment on canals and foramina, as well as limitations of movement, may engender complaints of pain and discomfort. According to the present study 76.9% of patients had a presenting complaint of swelling and 23.9% complained of pain. In general, males and females are thought to be affected evenly, although recent research has shown a slight female preponderance. However, McCune-Albright syndrome, a form of polyostotic fibrous dysplasia associated with cafe-au-lait pigmentation and multiple endocrinopathies such as precocious puberty, pituitary adenoma or hyperthyroidism, almost always affects females.

The literature suggests that fibrous dysplasia in women can be reactivated during pregnancy. This association is more commonly seen with the polyostotic...
tic form. Cystic lesions resembling aneurysmal bone cysts have been noted in association with the monostotic form.\textsuperscript{15}

Panoramic, reverse Towne, PA and lateral skull views are often adequate to visualize lesions in the mandible. It is desirable to have at least 2 images, exposed at right angles, to assess the extent of the lesion in all dimensions. Because of the complexity of the anatomy, CT is helpful for assessing lesions in the maxilla.\textsuperscript{11,16}

Differential diagnosis of the initial radiolucent stage must include the following: central ossifying fibroma (COF), central giant cell granuloma (CGCG), aneurysmal bone cyst, osteomyelitis and early fibrous lesions.\textsuperscript{17} Because these lesions represent a variety of disease processes with different behaviours, including infection and endocrine dysfunction, prompt diagnosis incorporating clinical, radiographic and, occasionally, histologic findings, is essential.\textsuperscript{18}

Fibrous dysplasia may also mimic Paget’s disease of bone on clinical examination, particularly if a patient with fibrous dysplasia does not present until later in life. In addition to the predilection of Paget’s disease for an older population, certain radiographic and clinical features help to distinguish this lesion from other radiographically similar lesions. These features include thickening of the cortices, cotton wool appearance of the involved bone and increased blood levels of alkaline phosphatase.\textsuperscript{19} The most useful clinical feature for distinguishing Paget’s disease from fibrous dysplasia is that the former tends to occur bilaterally in the jaws, whereas the latter affects only one side. Histologically, Paget’s disease exhibits many osseous dysplasia is that the former tends to occur bilaterally in the maxilla.\textsuperscript{20} The affected bone resides within a well-vascularized fibrous connective tissue stroma. Although osteomyelitis demonstrates sequestra in the later

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), endocrinologist.\textsuperscript{21} The multi-disciplinary approach becomes even more important in polyostotic fibrous dysplasia i.e., carno-facial and syndromic forms (Jaffe-Lichtenstein syndrome and McCune-Albright syndrome).

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