นิพนธ์ต้นฉบับ

Fibrous dysplasia of bone : A study of 44 cases.

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A retrospective study of fibrous dysplasia was conducted in 44 patients. All lesions were histopathologically verified. The lesion was composed of immature bony spicules and fibrous stroma. Only two instances of polyostotic variant were encountered; both were in male. The age of the patients ranged from 4 to 42 years. The average age was 21 years. Males were slightly more affected than females. Craniofacial bones were most commonly affected, and the maxilla was the most frequently involved bone. The Frontal bone, mandible, femur, tibia, and ribs were affected in decreasing frequency. Patients who had lesions in craniofacial regions usually presented with progressive swelling at the affected sites. However, lesions in frontal bones also caused pain and deformity. Lesions in long bones commonly produced mass and pain or even pathological fracture. Recurrences developed in six patients.

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พงษ์ศักดิ์ วรรณไกรโรจน์, สุมาลี ศิริอังกุล. ไฟบรัสดิสเพลเซียของกระดูก : การศึกษาใน 44 ราย. จุฬาลงกรณ์เวชสาร 2530 พฤศจิกายน; 31 (11) : 895-903

ไฟบรัสดิสเพลเซียของกระดูกเป็นความผิดปกติอย่างหนึ่งที่กระดูก ถูกแทนที่ด้วยเนื้อเยื่อไฟบรัสและกระดูกที่ไม่เจริญเต็มที่ รอยโรคมีขอบเขตค่อนข้างชัดเจนและโตขึ้นช้า ๆ พบได้ในกระดูกทุกแห่ง

*ผู้รายงานได้รวบรวมผู้ป่วยที่วินิจฉัยว่าเป็นไฟบรัสดิสเพลเซียในภาควิชาพยาธิวิทยา โรงพยาบาลจุฬาลงกรณ์ ตั้งแต่ปี พ.ศ. 2516 ถึง 2528 ได้ผู้ป่วยทั้งหมด 44 ราย พบเป็น **polyostotic fibrous dysplasia** 2 ราย เป็นชายทั้งคู่ บริเวณที่เป็นโรคมากที่สุด คือ กระดูกใบหน้า และกะโหลก (65.91%) กระดูกที่พบมากที่สุดคือ กระดูกขากรรไกรบน (29.55%) รองลงมาคือ กระดูกฟรอนตัล (13.64%) กระดูกขากรรไกรล่าง (11.36%) กระดูกฟีเมอร์ (11.36%) กระดูกทิเบีย (11.36%) และกระดูกซี่โครง (9.09%) อายุที่ป่วยมาหาแพทย์อยู่ระหว่าง 4 ปี ถึง 42 ปี โดยมีอายุเฉลี่ย 21 ปี พบโรคนี้ในผู้ชายมากกว่าผู้หญิงเล็กน้อย ผู้ป่วยที่มีโรคของกระดูกใบหน้าและกะโหลก มักจะมาหาแพทย์ด้วยมีก้อนในบริเวณที่เป็น ยกเว้นคนที่มีในกระดูกฟรอนตัล ซึ่งจะมีอาการปวดร่วมด้วย และทำให้หน้าตาเสียรูป ส่วนคนที่มีโรคในกระดูกยาวเกือบทั้งหมด มาพบแพทย์เพราะปวดในบริเวณที่เป็นโรค บางครั้งปวดมากจนเดินไม่ได้ และมี 1 ราย ที่มีกระดูกที่เป็นโรคนั่นหัก พบโรคเกิดซ้ำหลังจากรักษาแล้ว 6 ราย*

Fibrous dysplasia of bone is an infrequent skeletal disorder in which bone is replaced by fibro-osseous tissue. The lesion is well circumscribed and slowly progressive. It may affect any bone, but commonly in the craniofacial region or the lower extremities. It may be solitary (monostotic) or multiple (polyostotic). The latter is often associated with endocrine abnormalities (Albright's syndrome).

We have collected cases of fibrous dysplasia of bones diagnosed at Chulalongkorn Hospital between 1973 to 1985 to study their statistics concerning the age at presentation, the sex of the patients, the bones involved and manifestations of the disease in Thailand.

Materials and methods

All 44 instances of skeletal fibro-osseous tumor-like lesions in the pathological file at Chulalongkorn Hospital between 1973 to 1985 were reviewed. The radiographs were revised in some cases. The features are characterized by well-defined expansion of the involved bone with radiolucency or ground-glass appearance depending on the amount of osseous material within the lesion (Figures 1-3). Tissues were routinely processed and stained with hematoxylin and eosin (H & E). Polarized filters were also employed in examining the sections. Histopathology was re-evaluated in all cases. Microscopically, the disease is characterized by randomly distributed bony spicules embedded in fibrous stroma (Fig. 4). The bone is immature and formed by metaplastic process (Fig. 5). It contains irregularly distributed fibers that are weakly birefringent. This differs from normal bones that have lamellated, brightly refringent fibers. Osteoblastic rimming of bony spicule is not present in fibrous dysplasia. This differs from usual metaplastic bones which are formed by initial deposition of woven bones and followed by lamellar bone replacement synthesized by osteoblasts.

The stroma contains cellular fibrous tissue arranging in interlacing fashion with occasional storiform patterns (Fig. 6). Blood vessels and giant cells may be abundant in some areas (Fig. 7). Hemorrhage with hemosiderin deposition is occasionally found. Clusters of foamy histiocytes are also observed (Fig. 8).

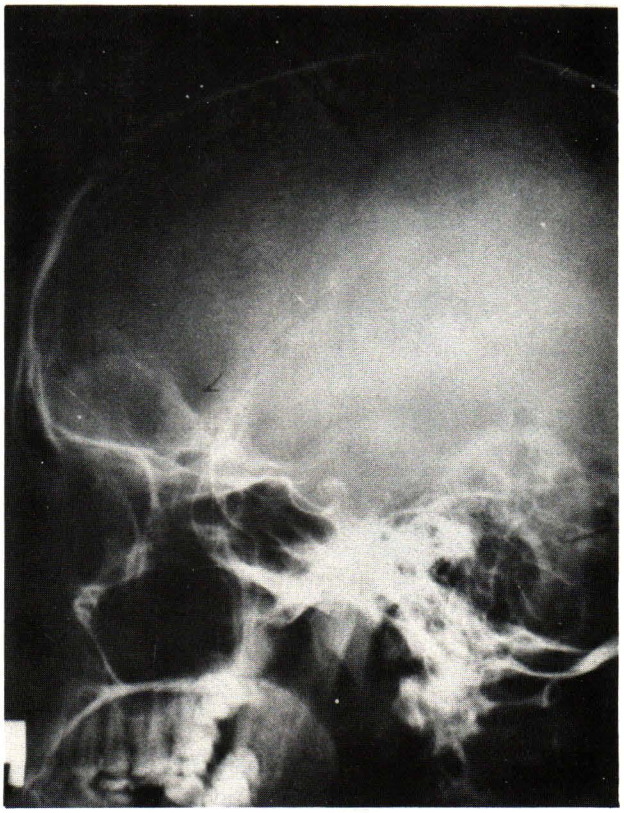

Figure 1 (Case 26). Roentgenograph of skull (lateral view) showing evidence of expansion of bone at anterior cranial fossa. The lesion has ground-glass appearance and well defined outline.

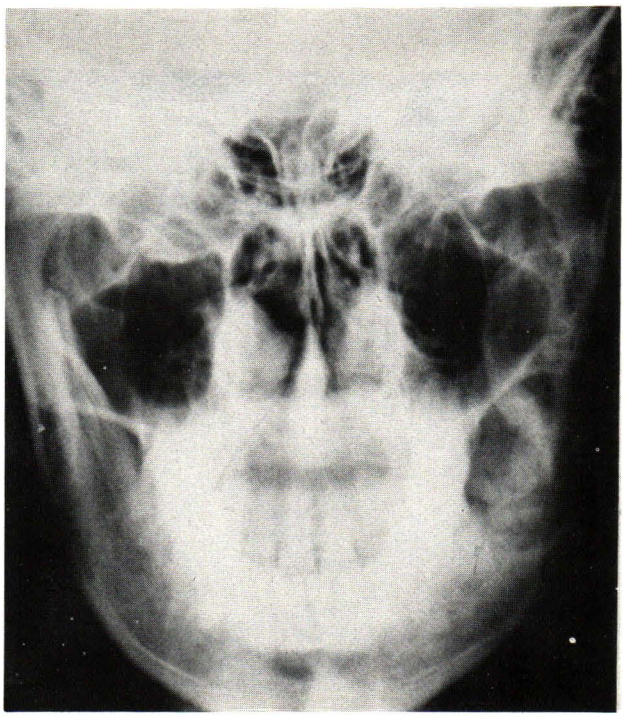

Figure 2 (Case 28). Roentgenograph of fibrous dysplasia of the right mandible (angle). Note well circumscribed, expansile, and radiolucent patterns.

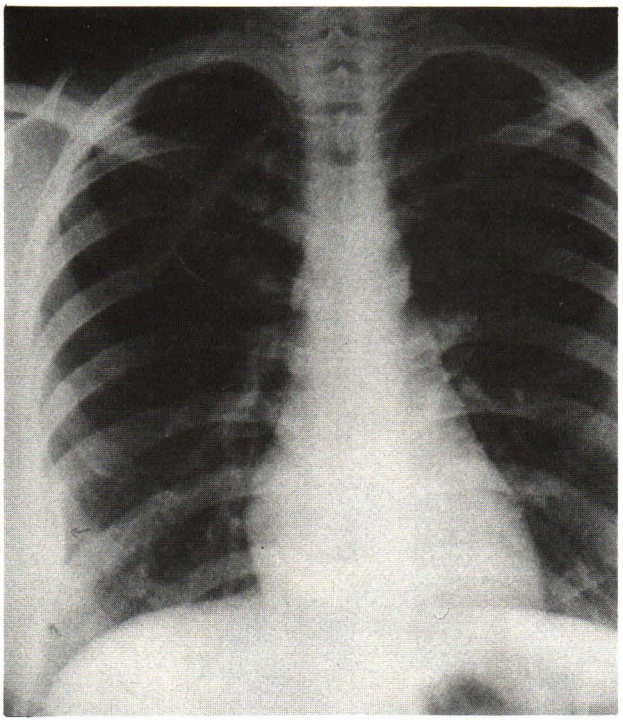

Figure 3 (Case 41). Fibrous dysplasia of rib. Note expansile lesion of the right 9^{th} rib without cortical destruction.

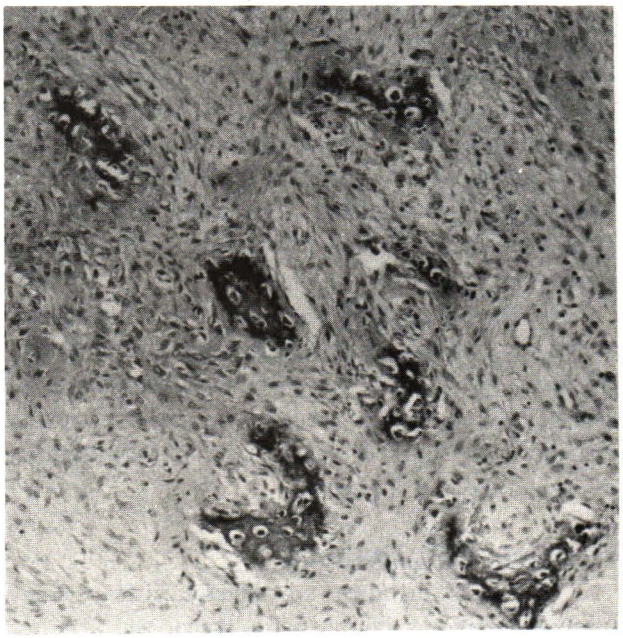

Figure 5 (Case 25). Lesion from tibia. Note poorly formed bony trabeculae without osteoblastic rimming. (H & E, × 100).

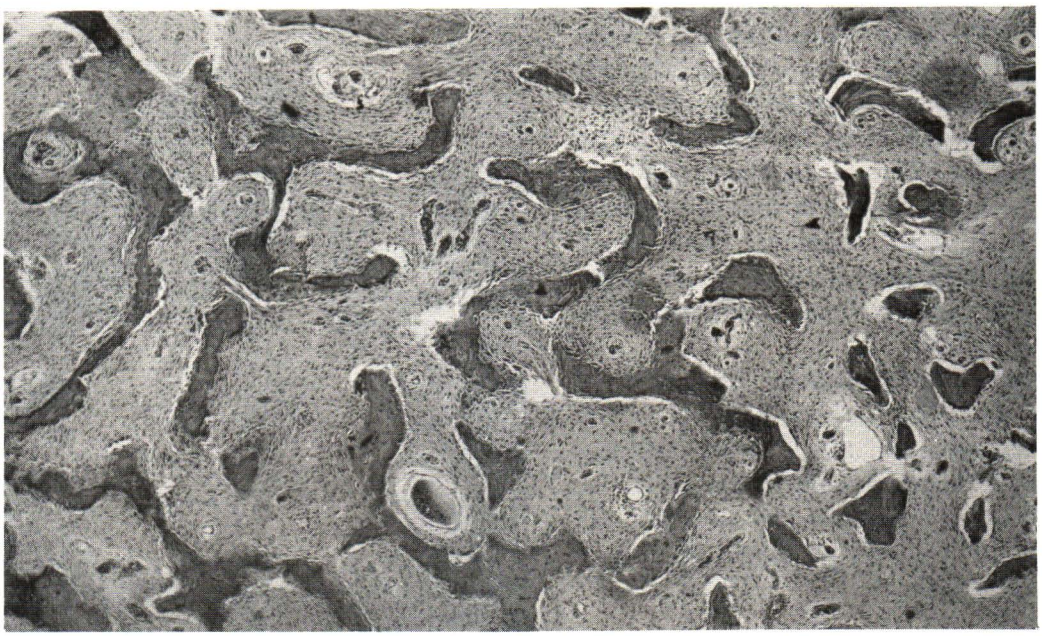

Figure 4 (Case 6). Fibrous dysplasia of bone. Note irregularly distributed bony spicules embedded in fibrous stroma. (H & E, × 40).

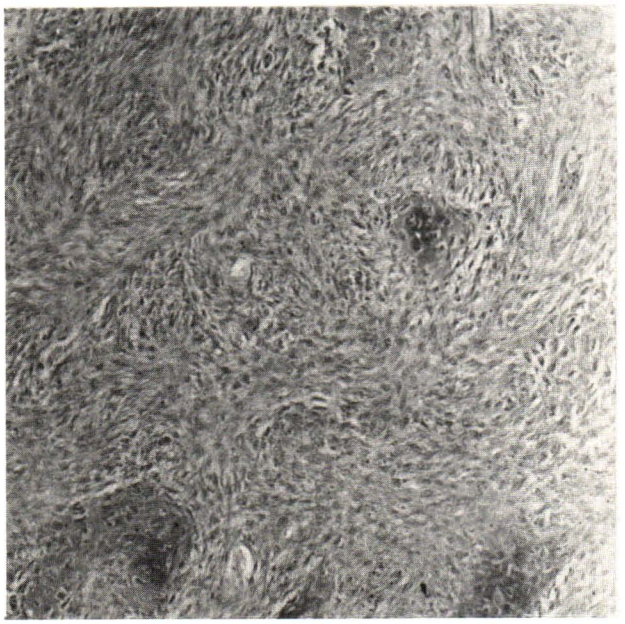

Figure 6 (Case 41). Cellular fibrous stroma showing storiform arrangement. (H & E, × 100).

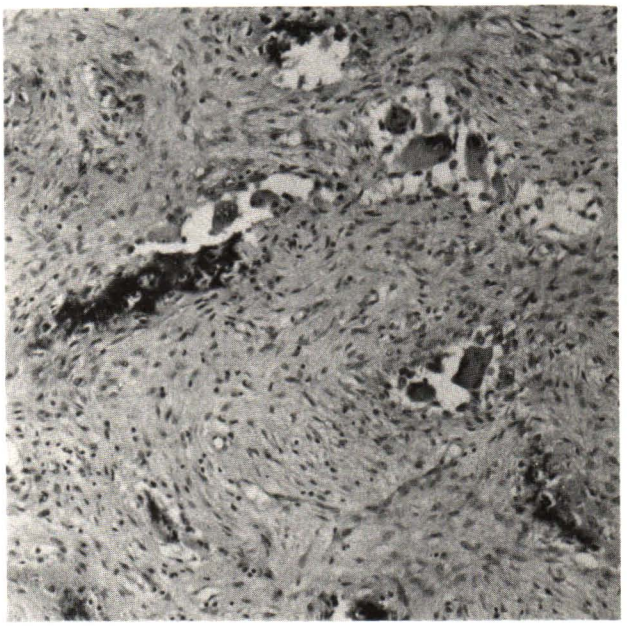

Figure 7 (Case 20). Scattered giant cells in the stroma are exhibited. (H & E, × 100).

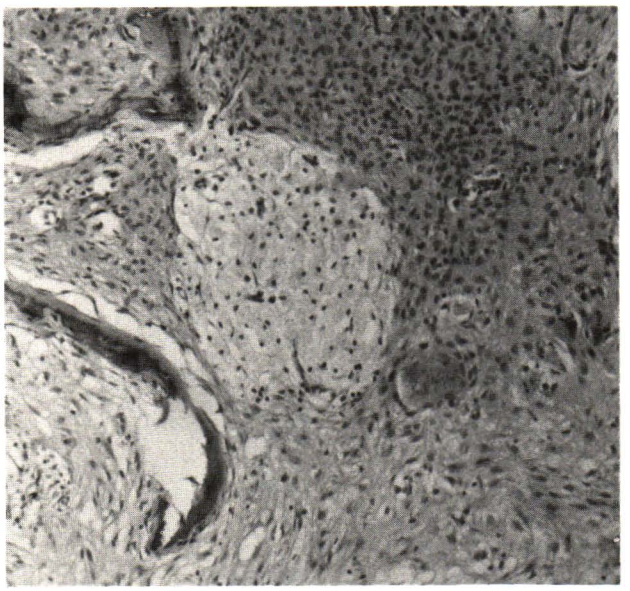

Figure 8 (Case 20). Foamy histiocyutes are occasionally seen in fibrous dysplasia. (H & E, × 100).

Results

Table 1 shows the age and sex of patients, the affected bones, and the manifestations in 44 cases of fibrous dysplasia.

The age of the patients at presentation of the malady ranged from 4 to 42 years with an average age of 21 years. Of the 44 patients, 25 were male and 19 were female. Only 2 patients had polyostotic lesions, both were males, 24 and 40 years old respectively. The younger patient presented with pain in the right leg which made him limping. Radiologic bone survey revealed multiple radiolucent areas in bones on the right side, eg. skull, femur, tibia, and fibula. The older patient experienced chest pain without history of trauma. Roentgenograph showed radiolucent lesions in the 3^{rd}, 6^{th}, 7^{th}, 8^{th} and 9^{th} ribs on the left side. The rest (42 patients) had monostotic lesions.

Thirty-four patients were 15 years old or older at the time of biopsy. Ten patients were under 15 years of age. In childhood period, males were more common (male : female = 7 : 3)

Locations of the disease are presented in Table 2. The most commonly involved region was craniofacial (66%) with maxilla being the most frequent site. For extracraniofacial region, femur, tibia, rib, and hip were affected in descending order.

For initial complaints, considering craniofacial bones, most of the patients came with mass or swelling in the involved region without pain except for cases of frontal bones that also had headache, or blurred vision, or deformity of the eyes. Lesions involved the long bones usually produced mass and pain that occasionally restricted the movement.

Table 1 Fibrous dysplasia showing age, sex, affected bones, and manifestations in 44 patients.

Case No.	Age at Biopsy (yr)	Sex	Bones biopsied	Manifestations*	Remarks
1	Not recorded	M	Tibia (rt)	Limitation of movement (4 mo)	
2	40	F	Maxilla (rt)	Mass (2 yr)	
3	18	F	Mandible (lt)	Mass	Recurrence twice within 3yr.
4	18	M	Maxilla (lt)	Swelling (2 yr)	
5	32	M	2nd rib (lt)	Not recorded	
6	22	M	Maxilla (lt)	Mass (5 yr)	Recurrence within 1 yr
7	24	F	Maxilla (lt)	Mass (many yr)	
8	30	F	Maxilla (lt)	Mass (24 yr)	
9	21	M	Frontal bone (lt)	Headache (1 yr)	
10	13	M	Maxilla (rt)	Mass (10 yr)	
11	22	F	Femur (rt)	Pain (1 mo)	
12	6	M	Maxilla (rt)	Swelling (4 yr)	
13	33	M	Frontal bone (lt)	Headache, dizziness (1 mo)	
14	30	F	Maxilla	Mass (3 yr)	
15	12	F	Femur (lt)	Pain (5 mo)	
16	18	M	Frontal bone (rt)	Pain (3 yr)	
17	14	M	Mastoid	Mass (7 mo)	
18	11	M	Maxilla	Swelling (1 yr)	
19	13	F	Maxilla (lt)	Mass (1 yr)	
20	42	F	Femur (lt)	Pain (1 mo)	
21	17	M	Maxilla (rt)	Mass (2 yr)	
22	39	F	Mandible (lt)	Mass (1 yr)	
23	21	M	5th rib (rt)	Pain (1 yr)	
24	14	F	Tibia (rt)	Mass, pain (2 mo)	
25	13	F	Tibia (rt)	Mass, pain (5 mo)	
26	15	F	Frontal bone (lt)	Face asymmetry (4 mo)	
27	16	F	Maxilla (lt)	Mass (3 yr)	
28	29	F	Mandible (rt)	Purulent discharge in the mouth	Recurrence within 2 mo.
29	13	M	Nasal bone	Mass	
30	28	M	Hip (rt)	Pain (1 yr)	
31	6	M	Maxilla	Dental caries, swelling (2 mo)	
32	24	M	Femur (rt)	Pain, limitation of movement	Polyostotic
33	40	M	6th rib (lt)	Pain	Polyostotic
34	4	M	Tibia	Mass (1 yr)	
35	28	M	Femur (rt)	Pain, fracture (2 mo)	
36	10	M	Frontal bone (rt)	Swelling	Recurrence within 10 yr.
37	22	M	Tibia (rt)	Pain	
38	21	M	Mandible (lt)	Mass (3 mo)	
39	15	F	Ethmoid	Nasal mass (3 yr)	
40	17	M	Orbital roof, temporal bone (rt)	Mass (many years)	
41	24	F	9th rib (rt)	Mass (3 mo)	
42	23	F	Mandible (lt)	Mass (4 mo)	Recurrence within 5 mo.
43	20	M	Vertex of skull	Mass (8 yr)	
44	16	F	Frontal bone (lt)	Mass (7 yr), pain (1 yr)	

* In the brackets are duration of the complaints. yr = year, mo = month.

Table 2 Locations of bones involvement in fibrous dysplasia.

LOCATIONS	NUMBER	PER CENT
Craniofacial bones		
Maxilla	13	29.55
Frontal bone	6	13.64
Mandible	5	11.36
Orbital roof and temporal bone	1	2.27
Vertex of skull	1	2.27
Ethmoid bone	1	2.27
Mastoid bone	1	2.27
Nasal bone	1	2.27
Total	29	65.91
Femur	5	11.36
Tibia	5	11.36
Rib	4	9.09
Hip	1	2.27
Total	44	

Discussion

Fibrous dysplasia is a circumscribed, slowly expanded condition of bone. This condition was first reported by Lichtenstein in 1938.[1] Nevertheless, the disorder had been recognized in association with cutaneous pigmentation and precocious puberty by McCune[2] in 1936 and Albright[3] in 1937. Subsequently, many cases were recognized in single bone as well as in multiple bones. Currently, fibrous dysplasia can be separated into three groups.[4] The first, monostotic fibrous dysplasia, affects only one bone. Most of fibrous dysplasias fall in this category (about 70% of cases).[4,5] The second, polyostotic fibrous dysplasia, involves multiple bones. It comprises about 30% or less, and is common in females. The third, the rare McCune-Albright's syndrome, is characterized by polyostotic fibrous dysplasia, cutaneous pigmentation, and endocrine abnormalities. The endocrine abnormality is usually manifested by precocious puberty and is more common in female.

In our study, most cases were monostotic fibrous dysplasia. We found only two cases that had multiple osteolytic lesions. Biopsy of one lesion in each case revealed fibrous dysplaia of the involved bone. Hence, we assummed that both patients had polyostotic variants. They had neither cutaneous pigmentation nor endocrine abnormalities. Thus, these cases are inconsistent with McCune-Albrith's syndrome.

Age and sex.

The disease occurs at any age but usually manifests in the second decade. In this study, the age at first biopsy ranged from 4 years to 42 years with the average age of 21 years. Most had history of prolonged symptoms before consulting the physicians. Thus, the accurate onset of the disease was difficult to asses. Again, because of prolonged symptoms, the age at time of biopsy was also considerably varied. The disorder was slightly more common in males while Lichtenstein[1] reported female predominance which was probably related to the number of cases of polyostotic variant included in his study. As stated earlier, the latter variant of fibrous dysplasia of bone is more common in female than in male.

Sites and manifestations.

The craniofacial bones are most commonly affected (Table 2). This was similar to the study of Reed[6] but contrasted with others.[5] Maxilla is the most frequently involved bone, accounting for 29.55 per cent of cases. This differed from the study of Ramsey[7] in which the mandible was found to be the most common site of involvement. For long bones, femur, tibia, and rib are the common sites of occurrence.

According to the literature, lesions that involved ethmoid sinus[8,9] and temporal bone[5,10] were uncommon. This is also true in our study.

We found one each in ethmoid sinus and temporal bone. The former, a 15-year-old female, presented with nasal stuffiness for 3 years. This case was reported separately.[11]

The presenting symptoms are usually related to the location of the lesions. The cranifoacial lesions were often silent and slowly expansive. They produced mass effect or swelling of the involved parts. Pain was not the chief complaint of the patients. This observation was similar to some studies.[7,12] Nevertheless, fibrous dysplasia of the frontal bone almost always presented with pain and/or facial deformity. A patient had swelling of the forehead for 7 years and then had pain that necessitated him to see the physician. Nearly all lesions of the long bones had varying degree of pain that sometimes was severe and limited the motion. It was inconsistent with the observation of Grabias who stated that lesions of the long bones caused no pain.[4] Pathological fracture was recorded in one patient. There were only two patients that presented with mass without complaint of pain. One had involvement of the rib for two to three months and another had swelling of the proximal tibia for one year.

Sarcoma arising in fibrous dysplasia has been observed, either simultaneously or subsequently discovered.[13,14] Most were osteogenic sarcoma. Fibrosarcoma, chondrosarcoma, and spindle cell sarcoma were also reported. However, we do not encounter any malignancy coexisting with fibrous dysplasia in our series.

Generally, surgical resection is considered when the lesion causes pain or advancing deformity, or interferes the normal function. Complete removal is often not done because of cosmetic reason. Radiotherapy is not recommended owing to increased risk of malignant transformation.[14,15,16] Recurrence is not uncommon especially when complete resection cannot be achieved and the patients undergo only currettage. The recurrence rate is about 20 to 30 per cent.[5,6,7,12] In our series, we had recurrent lesions in six pateints (13.63%)

Acknowledgement.

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