Proceedings S. Z. P. G. M. I. Vol. 2 (1988), pp. 68–75

Total Mandibulectomy and Immediate Reconstruction A Case of Large Ossifying Fibroma and Osteogenesis Imperfecta

ATIQUR RAHMAN ARAIN AND IQBAL M. SHEIKH
Deptt: of Dental Oral & Maxillofacial Surgery.

A rare case of large ossifying fibroma, mandible, occuring in a patient also affected by osteogenesis imperfecta was seen in the department of oral and maxillofacial surgery of Shaikh Zayed Hospital, Lahore. This was treated by total mandibulectomy and immediate reconstruction by rib grafting. Ossifying fibroma and fibrous dysplasia of bone are both members of the group of fibroosseous lesions and may be difficult to distinguish apart. Pathological fractures of long bones with poor and delayed remodelling of the callus, results in crippling deformities in patients affected by osteogenesis imperfecta.

CASE REPORT:

Patient N.N., an 11 years old girl, presented to the dental department with a huge swelling of mandible, causing facial deformity, inability to masticate, and a severe social handicap. She also had deformity of lower limbs making her an invalid confined to a wheel-chair.

HISTORY OF PRESENTING COMPLAINTS:

At two years of age, she had a minor fall and sustained a fracture of her left leg. The fracture healed in normal time. Since then she has had six fractures of her right and left legs. The fractures healed with malunion every time, resulting in a severe deformity and inability to walk. At three years of age a swelling in the lower jaw was noticed. The swelling was hard and painless and gradually increasing in size. She consulted many doctors but no satisfactory treatment was offered, until it was declared inoperable.

PAST MEDICAL HISTORY:

Was essentially not contributary. Her limb deformity was atributed to osteogenesis imperfecta.

FAMILY HISTORY

She has two brothers and three sisters. No member of her family is affected with a similar condition.

SOCIAL HISTORY

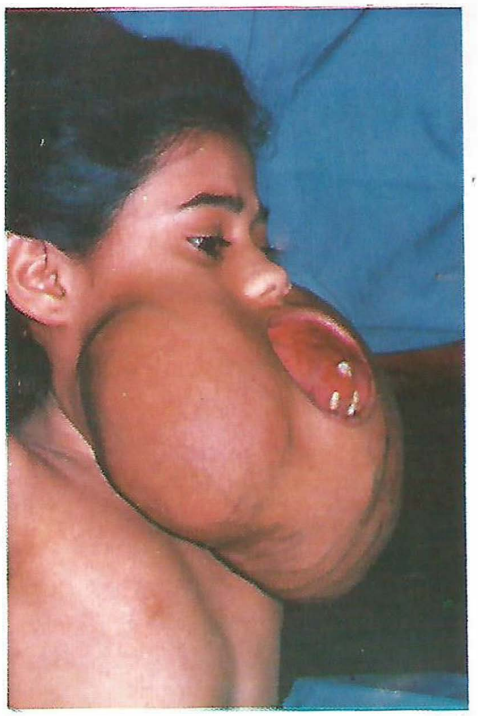
She is unable to attend school and is being educated at home she has read the Holy Quran.

PHYSICAL EXAMINATION

She was a small girl sitting in a baby pram, with deformed legs and a large deformed lower jaw. She was co-operative and intelligent. There was no obvious respiratory distress. There were no pigmented areas on her body. Her legs were severely deformed. Her lower jaw was greatly expanded. The swelling was more on the left side, and was hanging over her chest. The oral commissure was streched into a ring and she could not close her mouth. There was gross malalignment of her teeth, which were scattered all over the swelling. The tongue was pushed right into the back of her throat. The maxillary alveolus and her palate were also deformed due to the pressure from the swelling. There was lack of vertical growth of her maxilla which was flattened in the cranial direction. (Fig. 1)

The skin overlying the swelling was of normal colour, but greatly thinned due to stretching, with blood vessels showing through. There was no ulceration. (Fig. 2.)

The swelling was hard and non tender. No fluctuant areas could be found. Some movement of the temporoman-



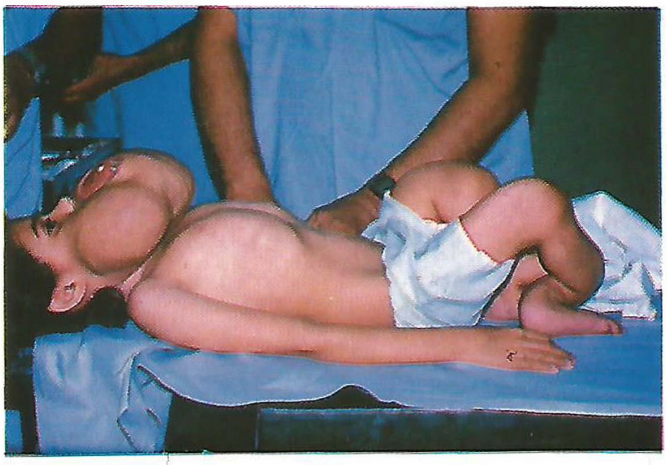


Fig: 1. The patient and the tumour as seen from the side

dibular joints could be demonstrated.
The sclera and teeth were of normal colour.
There were no signs of precocious puberty.

LABORATORY INVESTIGATIONS:

Her blood, urine, renal profile and liver profile examinations were essentially normal including the serum calcium and phosphate, except serum alkaline phosphatase which was 1460 U/L (Normal range 90–300U/L).

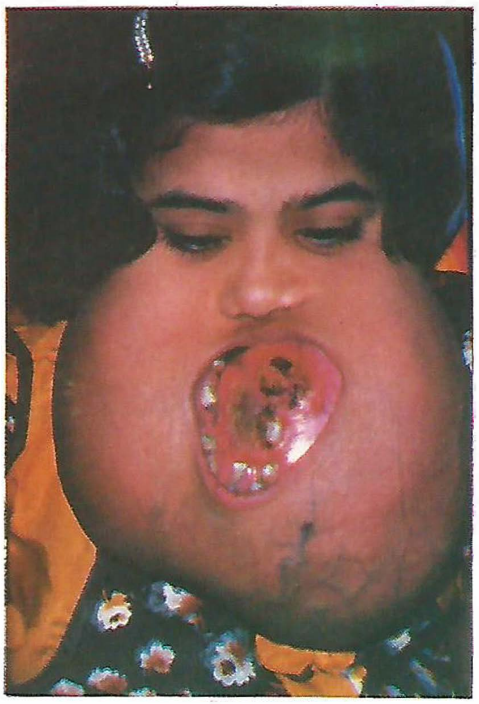


Fig: 2. The patient as seen from the front showing thinned skin and dilated blood vessels.

RADIOLOGICAL EXAMINATION:

1. Roentogenograms of mandible showed a large radio-opaque mass occupying the whole mandible, the normal bone architecture having been replaced by areas of irregular bone. The teeth had been grossly displaced. The radio opacity was greater in some areas than others. (Fig. 3)
DIAGNOSIS OF OSSIFYING FIBROMA of mandible was suggested.

On basis of the clinical picture a provisional diagnosis of ossifying Fibroma or Fibrous Dysplasia of bone was made, in the case of her mandibular swelling. (Table 1)

TREATMENT OBJECTIVES:-

Treatment objectives were considered under the following headings:-

1. IMMEDIATE
   1. Removal of the tumour.
   2. Restoration of cosmetics.
      Disposal of redundant soft tissues.
   3. Restoration of functions.

      (a) Provide anchorage to tongue.
      (b) Provide support to floor of mouth.
      (c) Provide firm base for construction of dental prosthesis in future.

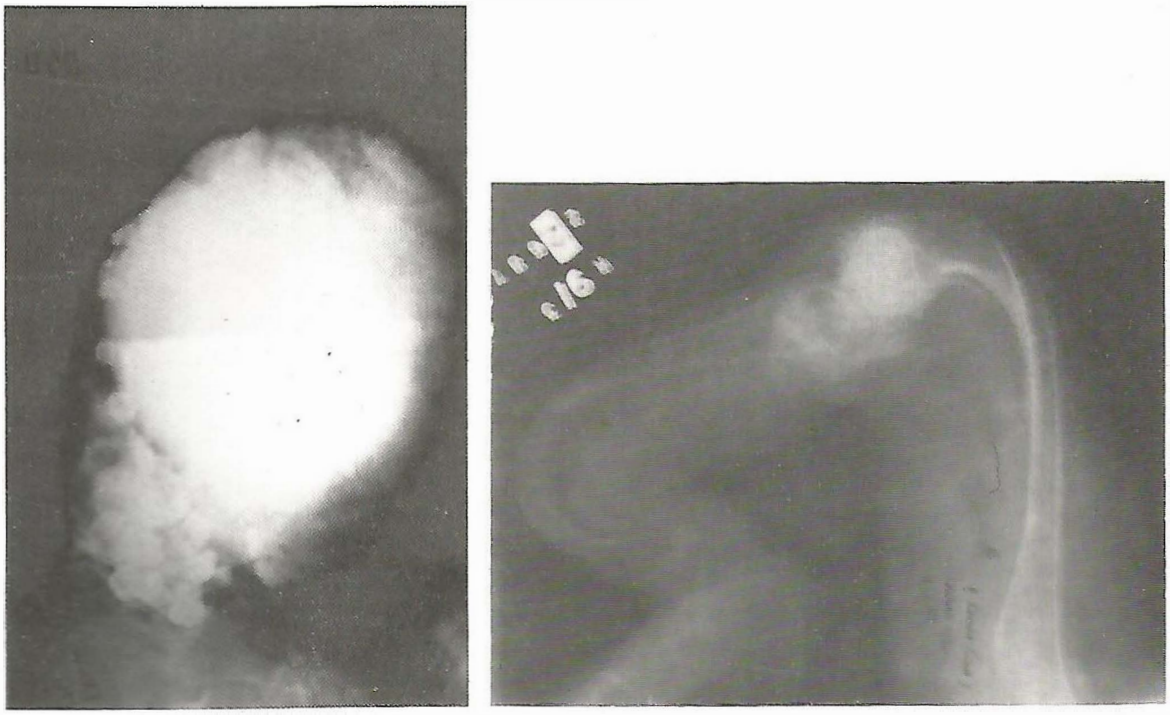


Fig: 3. Radiological appearance of the tumour and the patient's lower limbs.

TABLE – 1

DIFFERENTIAL DIAGNOSIS OF CENTRAL JAW SWELLING

Benign Neoplasms	Malignant Neoplasms	Non Neoplastic	Inflammatory Swellings	Cystic Lesions
1. Ossifying Fibroma also called fibro-osteoma	1. Ameloblastoma	1. Fibrous Dysplasia (a) Monostotic (b) Polyostotic	1. Central giant cell Reparative granuloma	1. Inflammatory & developmental odontogentic cysts.
2. Osteoma	2. Osteo-Sarcoma	(i) Jaffes' type (ii) Albright		2. Fissural cysts.
3. Chondroma	3. Chondro-Sarcoma	Syndrome		
4. Cementoma	4. Fibro-Sarcoma	2. Cherubism		
5. Fibromyxoma				
6. Neuro Fibroma		3. Pagets disease of bone.		

2. LATE
   1. Modification of scars and correction of residual deformities.
   2. Creation of buccolabial sulcus by
      (a) Building up bone
      (b) Buccolabial sulcoplasty.
   3. Construction of dental prosthesis.
   4. Realignment of maxillary teeth..
3. ORTHOPAEDIC
   Correction of deformity of lower limbs.

TREATMENT PLAN

To achieve the immediate objectives, the following treatment plan was envisaged.

1. Excision of the tumourous mandible.
2. Immediate reconstruction, using right and left seventh rib, taking cartilage to form the ascending part and bone to form the horizontal body of mandible.

She was admitted to hospital on 12-07-1987.

Her vital signs recorded were all normal, so was her chest X-Ray and E.C.G. Four units of blood were cross matched and kept ready.

OPERATION

General Anaesthesia was administered through a nasotracheal tube and the throat was packed.

An incision was made from left ear lobe to right ear lobe going around the swelling. Subperiosteal dissection was carried out and the mandible was exposed. Genial tubercles were found to be elongated in the form of a distinct spur. This bony spur was chiseled free from the Mandible. Intra oral incision was made along the duccal and lingual sulcus, sacrificing part of the oral mucosal covering of the swelling. Subperiosteal dissection was continued until the whole mandible was free except the heads of condyles. Swelling did not seem to extend to the compact bone of necks of condyles. Necks of condyles were osteotomized, saving both heads of condyles (Fig 4-5).

Two six inch pieces of 7th rib inlcuding cartilage and bone were obtained bilaterally. The bony ends of ribs were split longitudinallyto facilitate bending. The bony ends were wired together after appropriate bending and trimming. (Fig. 6)

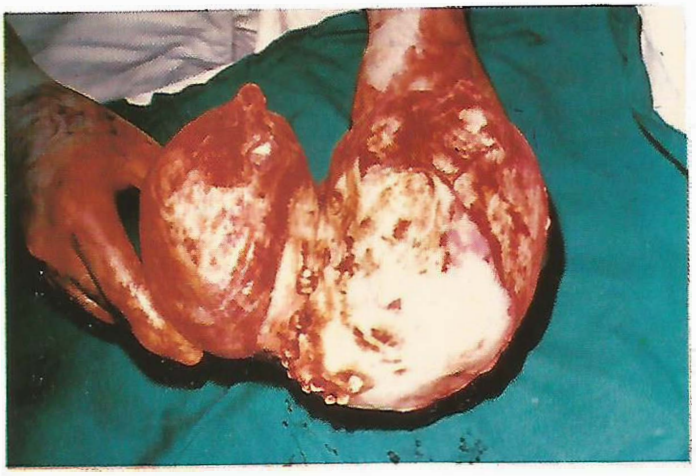


Fig : 4. Resected Tumour.

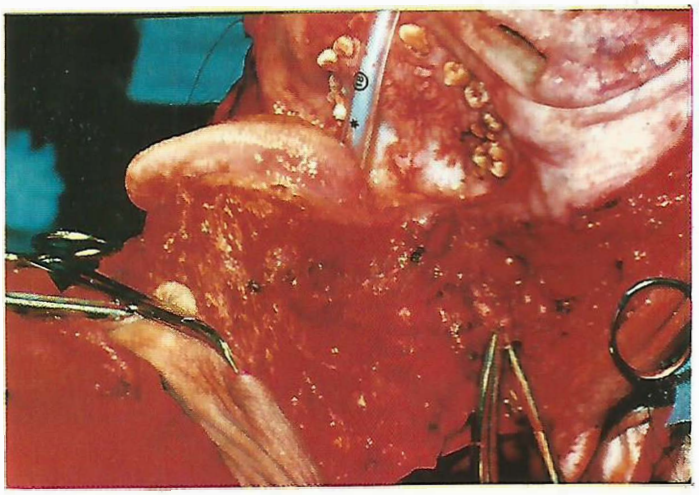


Fig: 5. The tumour bed after removal of the specimen

Fig: 6. Osteochondral rib graft bent and joined in mid-line.

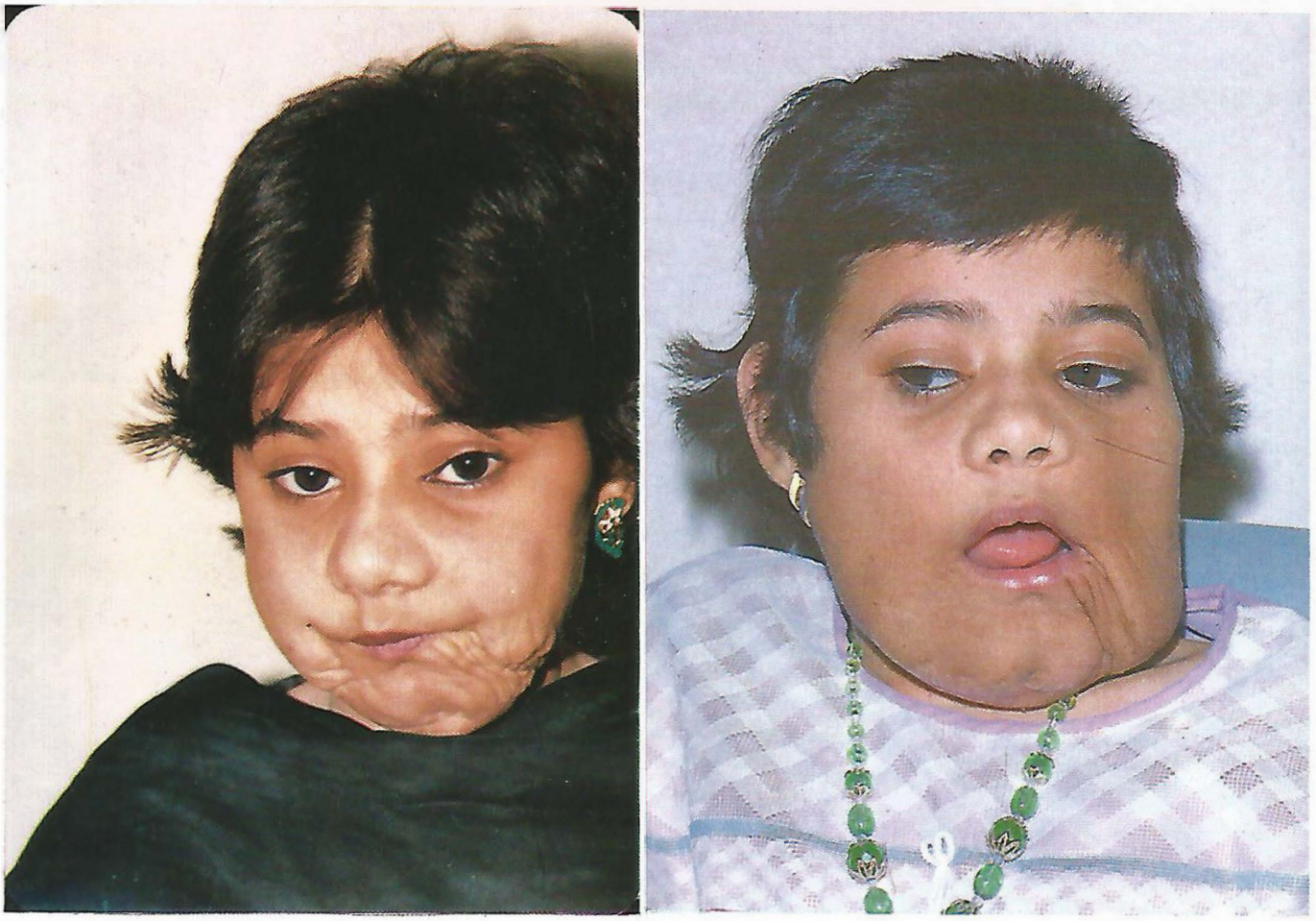


Fig: 7. Patient seen three months and one year post-operatively.

The chondral ends were wired to the condyle stumps and the genial tubercles were wired to the graft in the midline. Tissues on the oral side of graft were sutured in two layers. Redundant skin was dissected away from the underlying muscles and discarded. Masseters and medial pterygoids were identified and sutured over the bone graft. Rest of the wound was closed in layers, with vacuum drainage. Four units of blood were transfused during the operation.

Post operatively, the patient was transferred to I.C.U., where she made an uneventful recovery.

Feeding, was started through a nasogastric tube from the next day. After 3 days she was encouraged to take liquids orally.

All sutures from skin were removed on the 7th post operative day and the patient was discharged to go home. She was being kept on a regular weekly check ups.

(Fig 7)

HISTOPATHOLOGY

GROSS DESCRIPTION:- Specimen received fixed in formalin consisted of an irregular bilobed hard mass measuring 20 x 18 x 12cm. Weighing 3 kg/331 gms. Specimen bissected with saw and knife. Cut surface is grey white. There is a rim of 1.5 cm thickness, cancellous bone towards the periphery, centre is grey white firm with hard dark grey punctuations. Four representative section were taken in four blocks.

MICROSCOPIC DESCRIPTION

The histologic examination reveals a section of neoplastic tissue which is comprised of mature fibrous tissue and multiple foci of calcification and bone formation. Fibrous tissue and osteoid tissue is benign.

DIAGNOSIS

Ossifying Fibroma (Mandible)

Table I shows the various commoner causes of central jaw swellings. On the basis of clinical, radiological and histological picture, the diagnosis of Fibrous dysplasia of bone or ossifying fibroma of bone have to be considered. An overview of these entities is given below:-

FIBROUS DYSPLASIA OF BONE':-

(1) Von Recklinghausen[1] (1891)
- Described osteitis fibroma as an entity.

(2) Mandle[2] (1926)
- Reported first case of parathroidectomy resulting in exclusion of lesion due to hyperaparathyroidism.

(3) Albright, Bulter,[3] (1937)
Hampton & Smith
- Described five cases of what is now known as Albright's syndrome consisting of : Fibro-osseous lesions in several or many bones. Areas of light yellow or yellowish brown pigmentation. Sexual precocity particularly in girls. Precocious skeletal maturation in some cases. serum Ca & PO4 in normal range. Serum alkaline phosphatase could be raised.

(4) Lichtenstein[4] (1938)
- Showed that bone lesions could occur in the absence of extra skeletal features.

(5) Lichtenstein & Jaffe[5] (1942)
- Further showed that only one bone might be affected. At the same time, they coined the term fibrous dysplasia of bone occuring in a polyostotic or monostotic form.

DISTRIBUTION & INCIDENCE, GENERAL.
- Any bone of the body may be affected in various combinations.
- There may or may not be skin pigmentation.
- Only4–5% of cases have extra skeletal involvement.
- Female to male ratio is 2 : 1.
- 10% of monostotic lesions occur in skull bones.[8 9];
- 12% of skull lesions occur in jaw bones.[6 9])

JAW LESIONS
- Mostly solitary.
- Maxilla more commonly involved than mandible.
- Less frequently multiple jaw lesions.
- Least common are jaw lesions associated with trunk or limb bone lesions.

CLINICAL FEATURES.
- Most cases are seen in childhood.
- A few cases of solitary lesions are discovered in later life sometimes incidentally.
- Deformity - rounded or fusiform swelling.
- Bone pains.
- Invalidism.
- Pathological fractures.
- Nasal obstruction.
- Proptosis.
- Exopthalmos.
- Disturbance of eruption and malalignment of teeth.
- Pigmentation. (Cafe-au-lait spots).
- Endocrine disturbances may occur.
- Radiological appearance.

The affected bones are enlarged. There is usually a change in the bone pattern giving an orange peel, stippled. ground glass or fingerprint appearance. There may be areas of radiolucency and radiopacity. Radioluccent, areas may be large giving cystic appearance. In jaws, the teeth may be moved bodily and their eruption may be retarded.

PATHOLOGY

1. Yellowish or greyish white colour.
2. Gritty sensation to the knife on cutting.
3. Cysts, although rare in jaw lesions may be present[10]
4. Fibrous tissue replaces normal bone.
5. Trabeculae consisting of immature bone in varying proportion.
6. Fibrous tissue may be
   (a) Highly cellular-spindle cells arranged in whorled manner.
   (b) Thick interlacing strands of collagen.
7. Lameller hone may be present. [11,13]
8. Micro cyst formation due to focal degeneration of fibrous tissue.[13]
9. Presence of foam cells related to areas of degeneration & haemorrhage.[9]
10. Occurence of cartilage in polyostotic lesions.
11. Jaw lesions are more heavily ossified and trabeculac are thicker and blunter.[9]

ETIOLOGY THEORIES.

The following causes have been considered at various times.

1. Liver damage.
2. Glandular Dysfunction.
3. Infection.
4. Neurofibromatosis.
5. Lipoid Granulomatosis.
6. Trauma.
7. Maldevelopment (Current view).

BEHAVIOUR

- Benign
- Non neoplastic
- Self limiting
- Sometimes aggressive.

TREATMENT

- Not radiosensitive.
- Conservative surgical treatment.

OSSIFYING FIBROMA

Clinical features are very similar to those of solitary lesions of fibrous dysplasia i.e.,

1. All ages & sexes may be involved.
2. Localized hard swelling of jaw.
3. Painless & nontender.
4. Slow growing.
5. Mandible more commonly involved than maxilla.
6. More than one lesion may be present.
7. Usually a rounded well circumscribed swelling.

RADIOLOGICALLY

- Well defined radiolucent area, with a thin osteosclerotic rim, containing small irregular radiopaque areas or well circumscribed dense radiopaque area

PATHOLOGICALLY

- Usually well circumscribed.
- Fibrous content similar to fibrous dysplasia.
- Sometimes foci of spherical calcified material resembling dental cementum are scattered through-out the lesion.

DISTINCTION BETWEEN FIBROUS DYSPLASIA & OSSIFYING FIBROMA IS CLINICAL RATHER THAN HISTOPATHOLOGICAL:

OSSIFYING FIBROMA	FIBROUS DYSPLASIA
1. Boundary - Distinct.	1. Boundary Diffuse.
2. Thin bony shell of normal bone is usual and is more localized	2. May be extensive.
3. Behaviour is more like a neoplasm.	3. Behaviour is not like a true neoplasm but can grow to large dimensions over a long period.
4. Alkaline phosphatase not raised.	4. Alkaline phosphatase is raised.

The other condition which the patient is suffering from is osteogenesis imperfecta.

The salient features of this condtition are given below:-

OSTEOGENESIS IMPERFECTA

There is generalized osteoporosis of bone resulting in slender fragile bones. There is a disturbance of mesenchymal tissues particularly the calcified tissues.

Two forms are recognised

(a) Congenita - Lethal - Autosomal recessive.
(b) Tarda - Autosomal Dominant mostly milder form
- Autosomal recessive form exists.

CLINICAL FEATURES.

1. Long bones have thin narrow poorly formed cortices.
2. Spontaneous fractures —— resulting in short bowed deformed bones, specially in the lower limbs.
3. Hyperplastic callus may resemble a neoplasm.
4. Biconcave "Cod Fish" vertebral bodies.
5. Skull thin bulging particularly over the ears.
6. Thin translucent skin.
7. Sclera appears blue.
8. Dentine is Opalacent due to Dentinogenesis imperfecta.
9. Deafness due to otosclerosis.
10. Laxity of ligaments and joints.
11. Tendency for capillary bleeding.
12. Normal alkaline phosphatase.

PATHOLOGY

- Small amount of matrix, normally mineralized.
- Inability for reticulin to mature into collagen
- Reduction in the amount of type 1 (bone) collegen.

Disordered bone remodelling leads to osteoporosis.
Disordered callus remodelling leads to bending of fractured bones.

COMMENTS

Fibrous dysplasia and ossifying fibroma are similar in many

respects. Both have bone formation in a fibrous stroma, are slow growing and can achieve a large size. In this case, there was no clear demarcation of the lesion and it was bilateral. These favour a diagnosis of fibrous dysplasia. On the the other hand the bone formation in the lesion was mostly compact lamellar bone, which is common in ossifying fibroma. Lamellar bone that has been found in fibrous dysplasia has been in much older patients (Waldronetal) Both conditions are benign and the treatment is conservative. This patient went from doctor to doctor who refused to treat her in anticipation of the difficulties of reconstructions of total mandible, particularly in view of her other condition i.e. Osteogenesis Imperfecta. In this condition fractures heal in normal time. Deformity occurs during the remodelling phase which is excessively prolonged. But this is not a contra-indication to essential surgery. The situation of a young girl with grossly deformed legs and markedly enlarged mandible presented a pathetic situation demanding radical approach. Her new mandible grafted from her own ribs is working well. The danger of resorption of bone graft has to be kept in mind and any future plans of augmentation of body of mandible will have to depend upon the fate of her first bone graft. In case the graft does resorb, resort to some form of alloplastic implant will have to be made.

REFERENCES

1. Von Recklinghausen, F. Die fibrose oder deformiende Osteite. Festschrift Rudolf Virchow zu seinem 71 Gebrustage. *Berlin.* 1891.
2. Mandle, F. Therapeutscher Versuch bei einem Falle von Ostitis fibrosa generalisata mittels Exstirpation eines Epithelkorperchentumors. *Zentralb.f. Chir.*, 1926; 53,260.
3. Albright, F., Bulter, A.M., Hampton, A.O., and Smith, P. Syndrome Characterized by osteitis fibrosa dissemina areas of pigmentation and endocrine dysfunction, with precocious puberty in females, Report of five cases. *New Engl. J.Med.*, 1937; 216,727.
4. Lichtenstein, L. Polyostotic fibrous dysplasia Arch. Surg., 1938; 36,874.
5. Lichtenstein, L., and Jaffe, H.L. fibrous dysplasia of bone. A condition affecting one, several or many bones, the graver cases of which may present abnormal pigmentation of skin, premature sexual dvelopment, hyperthyroidism or still other extraskeletal abnormalities. *Arch. Path.*, 1941; 33, 777.
6. Schlumberger, H.G. fibrous dysplasia (ossifying fibroma) of the maxilla and mandible. *Amer. J. Orthodont.* (Oral Surg. Sect.) 1946; 32,579.
7. Jaffe, H.L. fibrous dysplasia of bone. A disease entity and specifically not an expression of neurofibromatosis, *J.Mt. Sinai Hosp.*, 1945; 12, 364.
8. Windholz, F. Cranial manifestations of fibrous dysplasia of bone. Their relation to toleontiasis ossea and to simple bone cysts of the vault. *Amer. J. Roentgenol.*, 1947; 58, 51.
9. Lucas:

9. Lucas: Pathology of tumours of the oral tissues (Churchil Livingstone, London, New York). 1984;
10. Obwegeser, H.L., Freihofer, H.P.M., and Horejs, J. Variations of fibrous dysplasia in the jaws. *J. max.-fac. Surg.*, 1973; I, 161.
11. Waldron, C.A., and Giansanti, J.S. Benign Fibroosseous lesions of the jaws: a clinical-radiologichistologic review of sixty-five cases. *Oral surg.*, 1973; 35, 190., 340.
12. Dahlgren, S.E., Lind, P.O., Lindbon, A., and Martensson, G. Fibrous dysplasia of jaw bones. A clinical, roentgenorgraphic and histopathologic study. *Acta Otolaryngol,* 1969; 68, 257.
13. El Deeb, M., Waite, E.E. and Jaspers, M.T. Fibrous dysplasia of the jaws. Report of five cases. *Oral Surg.*, 1979; 47, 312.