FIBROUS DYSPLASIA OF THE MAXILLA WITH A METHOD FOR PREVENTING CONTRACTURES AFTER MAXILLECTOMY

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The benign bony tumours which originate in the maxilla and which are not associated with the development of the teeth are usually ivory osteomata. This is in accord with the well-known rule that benign osseous tumours arising from bones whose development is in membrane are usually ivory osteomata, whereas those arising from bones whose development is in cartilage are, as a rule, cancellous in type. For this reason it must be extremely rare for a cancellous osteoma to be found in a maxilla.

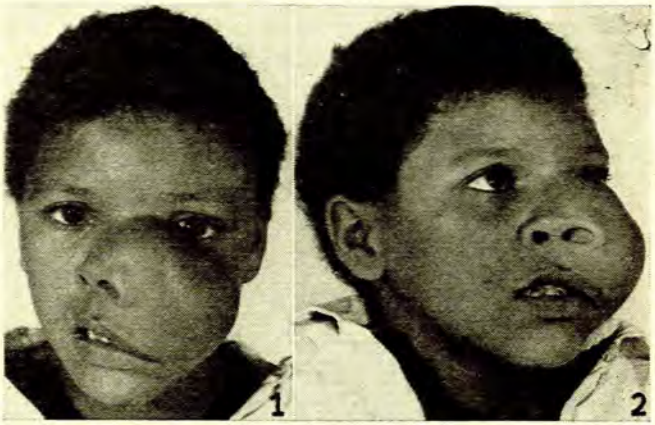
Fibrous dysplasia which affects a bone is, however, a well-known clinical entity. Two forms of this condition are described. The *polyostotic* variety or osteitis fibrosa disseminata is the commoner and has been the subject of a certain amount of controversy. Lichtenstein and Jaffe (1942) consider that this disease is related to some upset of the calcium metabolism and is in some ways allied to parathyroid disease, and Thannhauser (1944) believes that the condition is associated with neurofibromatosis of the multiple or von Recklinghausen type. The other and less common variety is the *monostotic* type which has been extensively studied by Schlumberger (1946). This author reviews 67 cases of monostotic fibrous dysplasia, of which 7 occurred in the maxilla. He considers 'that the condition is not a congenital anomaly and that it may represent a disturbance of the normal reparative processes following any variety of bone injury. In no case was there pigmentation and no other bone ever became involved. For this reason Schlumberger thinks that the condition is not an early stage of the polyostotic variety. He notes that the X-ray appearances are not specific and that the correct diagnosis is seldom made pre-operatively.

The *monostotic* variety is thus accepted by all authorities as a separate disease. The condition is never malignant nor does it ever become so. As a rule it is slow-growing, and it occurs mainly in children and in young adults (Phemister and Grimson, 1937), which is why Schlumberger, who collected his cases from the files of the American Armed Forces, found such a wealth of material at his disposal.

Several cases in the maxilla have been diagnosed as ossifying fibromas by the pathologists, but Lichtenstein (1938) points out 'that the histological picture in this condition in the maxilla is identical with fibrous dysplasia in other bones.' Most authorities agree that complete removal when the mass is small, and partial removal to prevent deformity should it be large, is the correct treatment to adopt. Radiotherapy is given post-operatively to keep the rate of growth down and repeated removals are advised should the condition recur.

CASE REPORT

C. le G., a Coloured female aged 15, had suffered from epilepsy since the age of 6 months. In the middle of 1948, when she was 13 years old, she noticed a swelling under the left eye about the size of a marble. The swelling increased slowly in size, and was painless; about 3 months before admission a rather more rapid growth occurred (Figs. 1 and 2). Some bleeding from the left



Figs. 1 *and* 2. The condition of the patient in January 1949.

nostril, by now pushed well over to the right, and increasing difficulty in seeing out of the left eye, brought the patient to hospital, where on 1 February 1949 a biopsy of the mass was performed. The histological report read as follows:

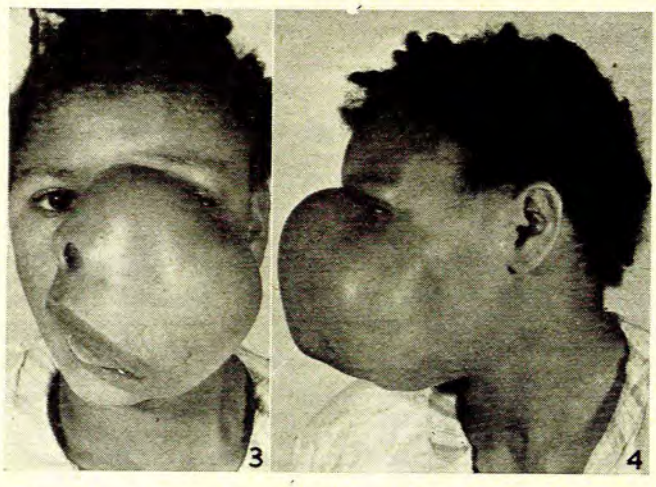
'Specimen is a piece of fibrous-looking tissue 0·8 cm. long. Histologically this shows bone and osteoid tissue embedded in fairly dense fibrous tissue. Bone formation is active as judged by the frequency of osteoblasts, and while some bone absorption is going on, this is much less than one would expect in an osteitis fibrosa cystica. If the lesion is a large one, it may be that this is not representative of the whole, but in the absence of an adequate clinical history one can only report activity getting near to but not reaching the degree seen in bone sarcomas, associated with a fibrous stroma reminiscent but not entirely typical of that seen in osteitis fibrosa cystica.' *Signed* J. L. Thomson.

At this stage the blood chemistry was done and the following figures are available: Serum albumin 4·2 g.%, serum globulin 2·9 g.%, total protein 7·1 g.%; serum calcium 10·6 mg.%; serum alkaline phosphatase 45 units (B); Wassermann reaction negative.

The blood count was as follows: RBCs 5,200,000 per c.mm., haemoglobin 13 g.%; WBC 7,700 per c.mm. (polymorphs 65%, lymphocytes 20%, mononuclears 15%); BSR 14 mm. per hour.

On the assumption that the rapid increase of growth indicated a sarcomatous change, the patient was referred for radiotherapy to Dr. J. M. Grieve, who commented that 'the possibility of cure by X-ray therapy has been ruled out in this patient' and referred the case for surgery.

By August 1949 the mass had grown to the size indicated in the photographs (Figs. 3 and 4) and the vision from the left eye



Figs. 3 *and* 4. Same patient in August 1949.

had been completely obscured by the size of the tumour. The palate was pushed down and the alveolus was covered with large veins which crossed the bony mass; at no place had the mucosa become involved. An X-ray (Fig. 5) showed the large soft-tissue

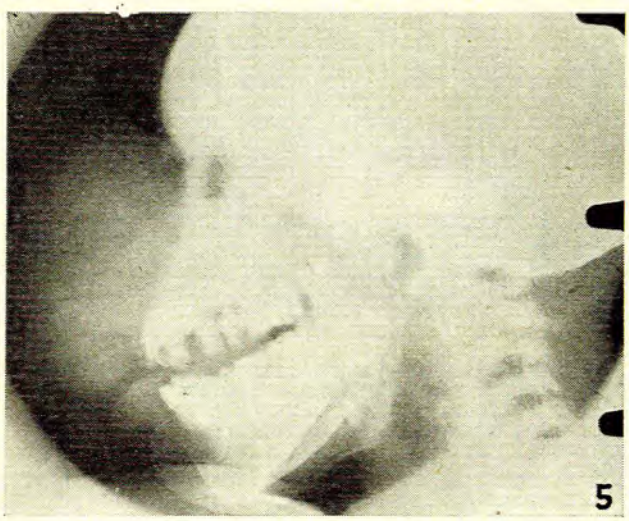


Fig. 5. X-ray, August 1949.

swelling with discrete calcification and destruction of the anterior wall of the maxilla. An osteogenic sarcoma was diagnosed. X-rays of the chest and a careful examination of the remaining bones of the body showed that there were no other bones involved.

On 29 August 1949, under intratracheal anaesthesia, given by Dr. P. Jenkin, the usual incision was made for removal of a maxilla, and the skin of the face was stripped off the bone and reflected laterally. The zygomatic arch, the lateral orbital pillar and the nasal process of the maxilla were individually exposed and divided. The mucosa of the palate was now incised in the mid-line and stripped off the tumour, and the hard palate was divided at its septal attachment with bone cutters. The soft palate was now separated from its attachment to the hard palate. The tumour was then rocked and with a little easing the maxilla and the mass came away. A roll of gauze, previously prepared, was immediately pressed into the cavity but there was comparatively little haemorrhage.

During the cutting of the lateral orbital process and the zygomatic process, it was noticed that the bone was very soft and could be cut with the knife, giving a gritty sensation as this was being done.

In order to fill the large space occupied by the mass, a ball of gauze, soaked in flavine, about the size of a golf ball, was now prepared and a Thiersch graft was cut from the inside of the thigh by my colleague, Mr. D. S. Davies, who kindly assisted at the operation. The skin graft was wrapped around the gauze ball with the epithelial surface inwards. Haemostasis was now secured, and the distorted nasal bones which had been pressed outwards by the tumour were fractured, straightened, and pushed back into position. A soft-paraffin plug was inserted into the left nostril. The wound was sprayed with a mixture of penicillin and thrombin and, after the skin-covered ball was placed into the cavity, the skin and mucosa were sutured over the ball. The face and nose were then moulded to an approximately normal shape.

At the conclusion of the operation, which took 3 hours, there was a fair degree of shock, but after the patient had received 2 pints of blood, her condition was fully restored to normal.

Convalescence was smooth, both temperature and pulse returning to normal by the 5th day. On the 7th day, under general anaesthesia, the mucosa of the hard palate, which had by now practically healed, was widely excised so that a large foramen could be left leading into the skin-lined cavity. The gauze plug

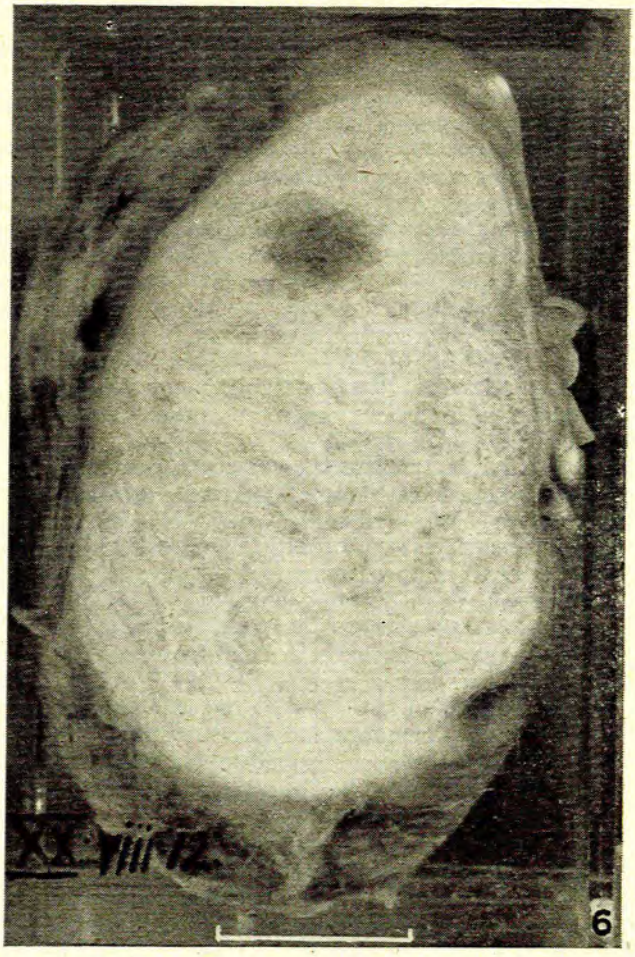


Fig. 6. The tumour removed, showing the whorled cut section. Scale represents 1 inch.

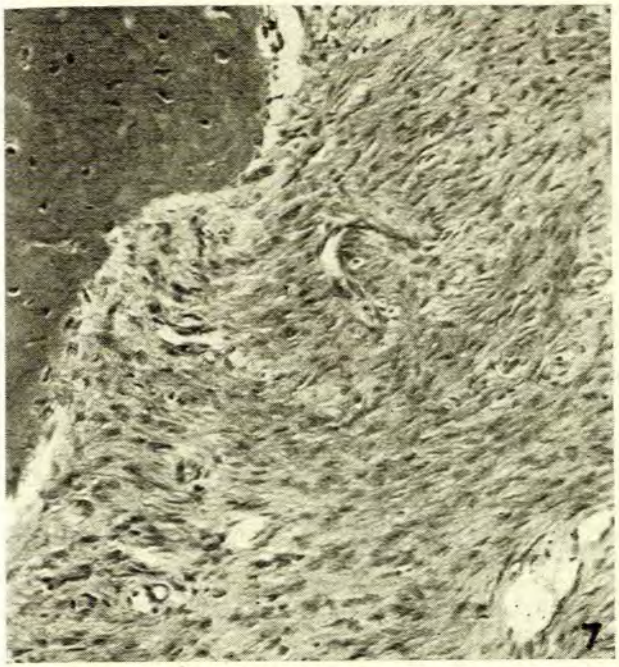


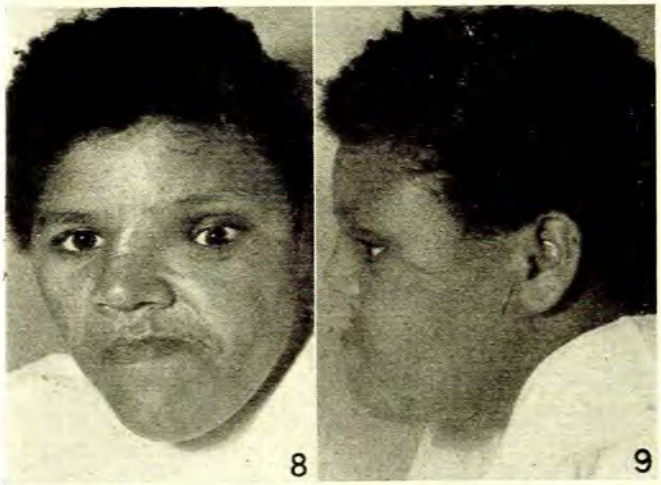
Fig. 7. Microscopic appearance of the tumour (×120).

was removed and it was seen that the epithelium had taken over most of the raw area. Ten days later, healing was complete and a large cavity, easily accommodating the end of a thumb, occupied the position of the antrum. The cavity communicated freely with the mouth.

The pathological report of the tumour was as follows:

'Specimen consists of an ovoid encapsulated mass 12·5×10×8 cm., on the under surface of which are recognizable the left half of maxilla and the corresponding teeth. The cut surface is very dense and fibrous, white with strands of translucent softer tissue. It creaks and grates with the knife but local calcification or bone formation is not recognized. The lines of section through maxilla show bone so decalcified as to be cut readily with a knife. The exterior presents a smooth rounded appearance and removal would appear to have been complete (Fig. 6). Histologically, the tumour consists of fibrous tissue containing lamellae of osteoid tissue, and it is only exceptionally that some calcification is recognized. The connective tissue adjacent to the plates of osteoid tissue is formed of plump spindle-shaped cells, but elsewhere it is of normal type (Fig. 7). In a few areas the osteoid tissue is being absorbed by osteoclasts. A section from the maxilla shows a marked lacunar resorption of bone, with abundant osteoclasts. Much of the bone is decalcified and between the bony lamellae is much fibrous tissue, similar to that of an osteitis fibrosa cystica in amount, but much denser, and the picture is that of *fibrous dysplasia of bone*. The tumour is well differentiated and slow growing; to name it is not easy, but a *cancellous osteoma* is suggested'. *Signed* J. L. Thomson.

The patient had been getting major epileptic fits at intervals and was referred to the Neurological Department for attention. She appeared at their out-patient clinic at irregular intervals and in May 1953 another series of pictures were taken (Figs. 8 and 9).



Figs. 8 *and* 9. Appearance of the patient in May 1953.

DISCUSSION

Removal of the maxilla in an adolescent must of necessity lead to great deformity as the years pass and the rest of the bones of the face develop. In addition, loss of the orbital plate often leads to dropping of the level of the eye, with an increase in the grotesque appearance. It was felt that immediate lining of the raw surfaces of the post-operative cavity with a Thiersch graft would, by reducing contractures, help to prevent much of the deformity that would arise, and that the bulk of the Thiersch graft itself would, to a certain extent, help to keep the eye in its normal position. Later, it was hoped to fit a prosthesis attached to the upper dental plate which would maintain the contour of the face. The retarded mental condition of the patient and the nature of her disease would not allow this to be done, but we were fortunate in that contractures were to a large extent prevented, presumably by the absence of raw surfaces. The large foramen made in the palatal mucosa allowed the skin-lined cavity to drain naturally into the mouth and made it to all intents and purposes self-cleansing; this is an important point in dealing with a mentally retarded patient.

All in all we are satisfied that the method adopted to prevent deformity in this case is one that could, with advantage, be applied on those occasions where large maxillary tumours require to be extirpated. There is almost a complete take in the graft, embedded as it is within the wound, and not only is healing greatly accelerated, but there has been a gratifying reduction in those contractures which had been anticipated.

SUMMARY

1. A case of a large monostotic fibrous dysplasia of the maxilla and its removal is recorded.

2. A method is described which may be of use in preventing deformity after removal of the maxilla.

I should like to thank my colleagues Messrs. G. Sacks and D. S. Davies, of the Departments of Surgery and Plastic Surgery, for their interest and assistance in this case. Mr. G. McManus, of the Department of Surgery, kindly prepared the photographs.

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