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A CASE OF FIBROUS DYSPLASIA (JAFFE–LICHTENSTEIN) WITH VERTEBRAL FRACTURE AND COMPRESSION OF THE SPINAL CORD

By

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The concept of fibrous dysplasia was introduced in 1938 by *Lichtenstein.* In a survey, *Jaffe & Lichtenstein* in 1942 described the disease as "a condition affecting one, several or many bones, the graver cases of which may present abnormal pigmentation of skin, premature sexual development, hyperthyreoidism or still other extraskeletal abnormalities".

The lesions may occur in any bone, the monostotic preferably in the ribs and jaws, the polyostotic most commonly in the femur, the tibia, the pelvis and the humerus. Lesions in the vertebral column are, according to the most experienced authors, infrequent (*Jaffe, Lichtenstein, Uehlinger)* and the absence of such lesions has even been regarded as a diagnostic criteria (*Uehlinger*). Reviewing the literature the author has found 29 cases with vertebral lesions out of about 300 cases of fibrous dysplasia so far reported. In most cases the lesions were to be found in the thoracic spine. Symptome from the spinal cord or nerve roots occurred in 8 of the 29 cases, usually with varying degrees of paraplegia. In three of these cases there were compression fractures (*Teng et al., Skanse et al., Jirout, Lewit*). In the remaining five cases the symptoms were caused by expansion of either a vertebral body or of pedicles and articular processes (*Rosendahl-Jensen, Lecocq, Ledoux-Lebard, Jaffe*).

The author has had the opportunity of following a case of polyostotic fibrous dysplasia, an 18 year old boy, who in 1954, at the age of 8, sustained a fracture of the right distal humerus and two years later refracture. A cystic expansion of the bone in the region of the fracture

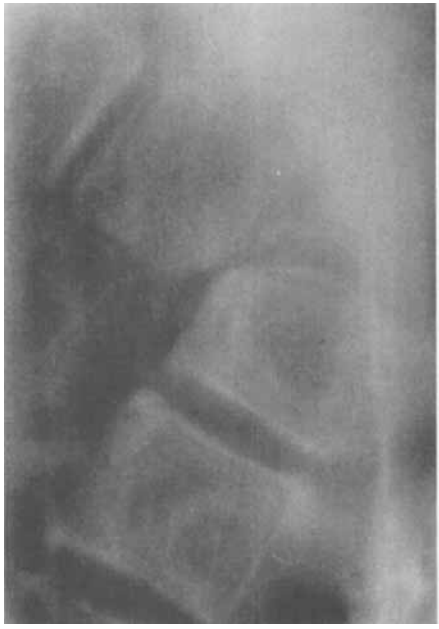


Fig. 1. *Fig. 2.*

Fig. 1. Right angle gibbus at the level of the D6 vertebra of which only a small, wedge-shaped fragment remains. In the posterior parts of the vertebral bodies of D7 and D8 solitary, well demarcated rounded radiolucenses are seen (tomography).

Fig. 2. Gas myelography (cisternal puncture). Partial block at the level of the compressed vertebra (tomography).

gave rise to a radiologic examination of the entire skeleton revealing changes in the skull, several ribs, the right scapula and both hands. At this time there were no signs of vertebral lesions. During the following years he was free of symptoms, but in 1961 increasing pain in the thoracic spine and a gibbus developed. In June, 1962, within a few days, weakness appeared in the lower extremities, there was sensory loss over the lower part of the trunk and both legs and urinary retention.

On admission in October, 1962, examination disclosed a normally developed 16 year old boy with a severe upper thoracic gibbus. There was paraplegia with diminished sensation which varied for different modalities up to the D6 segment.

Radiologic examinations: Figs. 1–3. In addition to those in the vertebrae and the skull there were changes in several ribs bilaterally, in

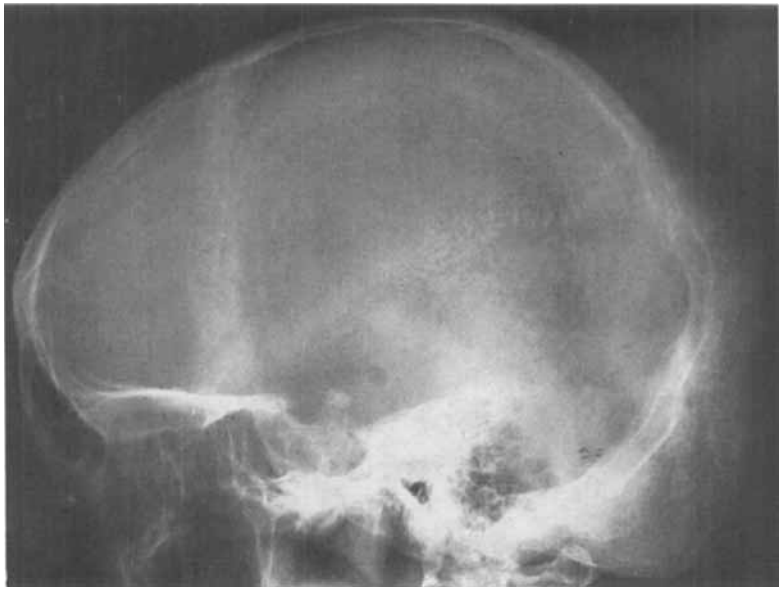


Fig. 3.
Sclerotic thickening of the external table of the right occipital and the sphenoid bone.

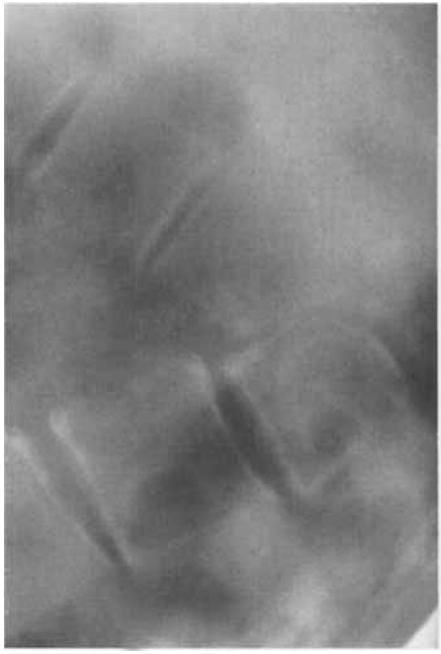
the manubrium, in the right scapula and humerus and in most of the metacarpals and phalanges of both hands.

The blood-chemistry was normal.

In spite of unchanging radiographic status, all signs of spinal cord compression entirely disappeared on conservative treatment consisting of bedrest and a plaster cast. To prevent recurrence a posterior spinal fusion (Albee-Hibbs) with a cortical graft was made in February, 1963. Microscopic examination of bone tissue removed at the operation confirmed the diagnosis of fibrous dysplasia.

At a radiologic examination one year after the operation, the patient was still free of symptoms, and the bone graft was shown to be completely resorbed. The angulation of the spine had increased (Fig. 4). With the exception of those in the humerus (Fig. 5) the other skeletal lesions were unchanged. A small exploratory incision over the easily palpable humeral expansion revealed a very thin cortex covering a huge cyst filled with yellowish fluid.

When last seen, in May, 1964, the patient was still free of symptoms and the radiologic appearance of the lesions remained unchanged.



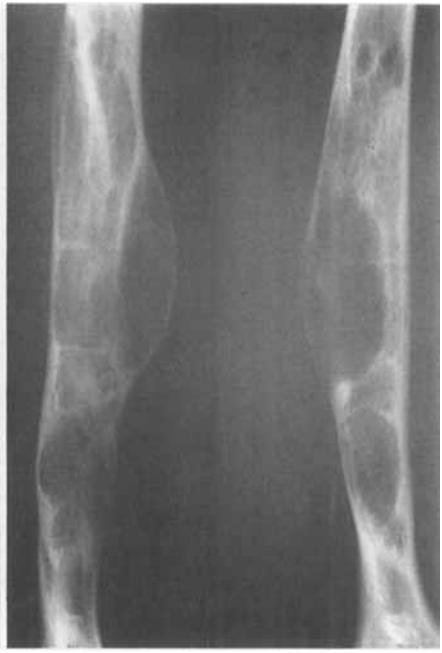


Fig. 4. *Fig. 5.*

Fig. 4. Increased angulation of the spine with the D5 vertebra resting on anterior surface of D7.

Fig. 5. Loculated cyst-like exspansion with extreme thinning of the cortex in the ventro-lateral part of right distal humerus.

DISCUSSION

The isolated bone lesions in fibrous dysplasia are, with the exception of those occurring in the skull, radiographically not characteristic. Cyst-like lesions in the diaphyses of the long bones or in flat bones should give rise to an examination of the entire skeleton, including the skull. In this case the diagnosis was based on the presence of multiple cyst-like expansive bone lesions, sparing the epiphyses, having a mainly unilateral extension in an otherwise normal skeleton of a growing individual and coexisting with a sclerotic thickening of certain skull bones. New manifestations of the disease may, as illustrated by the present case, appear until the closure of the epiphyses. The early diagnosis and treatment of lesions which could conceivable give rise to complications such as fractures or pressure on adjacent tissues are of prime importance. In this severely complicated case surgery seems

to have added nothing to the beneficial effect obtained by conservative treatment.

SUMMARY

A case of fibrous dysplasia with paraplegia caused by lesions in the thoracic spine and successfully treated with immobilization and bone grafting is presented. The localization of the disease to the vertebral column is infrequent, but a review of the literature suggests more common occurrence than previously stated. The radiographic appearance of the lesions is discussed and the importance of early diagnosis and observation is stressed.

RESUME

Il est rapporté un cas de dysplasie fibreuse avec paraplégie causée par des lésions dans la colonne thoracique et traité avec succès par l'immobilisation et une greffe osseuse. La localisation de la maladie dans la colonne vertébrale n'est pas fréquente, mais après avoir passé la littérature en revue, l'auteur estime qu'il convient d'envisager que le pourcentage de ces cas est supérieur à celui que l'on avait prévu juscu'ici. Il est discuté de l'appearance radiographique des lésions et l'importance d'un diagnostic et d'une obsérvation précoces sont soulignés.

ZUSAMMENFASSUNG

Ein Fall von fibröser Dysplasie mit Paraplegie, hervorgerufen durch Ergriffensein der Brustwirbelsäule und erfolgreich mittels Ruhigstellung und Knochentransplantation behandelt, wird vorgestellt. Die Lokalisation der Erkrankung in der Wirbelsäule ist selten, aber der Verfasser meint, nach Durchsicht der Literatur, dass sie doch häufiger vorkommt als früher festgestellt wurde. Das Erscheinen der Erkrankung im Röntgenbilde wird besprochen und die Wichtigkeit der frühzeitigen Diagnose und Beobachtung wird hervorgehoben.

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