Polyostotic Fibrous Dysplasia and Spinal Cord Compression

Case Report

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Fibrous dysplasia of bone is a disturbance in postnatal maintenance of intramedullary bone, in which normal bone that has undergone physiologic lysis is replaced by abnormal proliferation of fibrous tissue. When the disease is associated with skin pigmentation, polyostotic lesions, and endocrine abnormalities, especially precocious puberty in females, it is called Albright's syndrome. It is thought to be caused by a neuroendocrinologic dysfunction, but the pathologic findings in the few autopsies performed have been inconclusive, and the etiology of this disorder therefore remains a challenge to the pathologist and the investigator in the field of genetics.

Polyostotic fibrous dysplasia is usually manifested in childhood and occasionally in infancy. All bones in the skeleton can become affected, especially the long bones. The disease usually begins in the metaphysis, affecting cortical and cancellous bone; the lesions expand from within, and the peristeme is reduced to a thin lamina. The weakness of the cortex is one of the reasons for the multiple fractures which are often the first evidence of the disorder and which may cause severe deformities.

The macroscopic appearance of the lesions is that of normal bone replaced in portions by irregular masses of pale tissue. True macroscopic cysts are not common. The cystic appearance in the x-rays is due to replacement of bone by fibrous tissue. Microscopically the basic process is that of direct fibrous metaplasia. Spicules of bone are embedded in a matrix of collagenous tissue; islands of cartilage are present.

The x-rays show areas of rarefaction resembling bone cysts, which vary in size and shape. The lesions may occur in flat and in long bones. The borders of the lesions may be sharply defined, or fade in continuity with normal bone without demarcation. Areas of increased density may be present, especially in the skull. Thinning of the cortex is another significant radiologic finding.

This report concerns a case of polyostotic fibrous dysplasia that involved the dorsal spine and caused compression of the spinal cord, which responded favorably to decompressive laminectomy.

Case Report

This 56-year-old woman was first examined at the Cleveland Clinic on March 8, 1966, because of "trouble" with her legs. Nine months previously an uncomfortable feeling had developed in the right side of the abdomen, which had spread down both legs into the feet as "numbness." This discomfort was mild at first and was not related to gastrointestinal function. Within 4 months the patient noted that her legs had become weak and "heavy", that she had difficulty in walking, and had frequent leg spasms. In October, 1965, she was hospitalized and underwent gastrointestinal studies, the results of which were normal. The symptoms increased in severity, and the patient became constipated but retained bladder function. There was increasing numbness and weakness of both legs until she was unable to walk.

Past history showed that the patient had fallen and fractured the left hip when she was 6 years old and again when she was 12. Subsequently, the left leg became deformed and shortened. The right arm was fractured 12 times and the left arm three times. When she was 26 years old, the right supraorbital ridge was fractured, and a progressive swelling developed in this region. She had been told that she had fragile bones, but she had never consulted a specialist. She had not had precocious puberty.

Examination. The patient was a dwarfed, greatly deformed woman with a prominent right supraorbital mass (Fig. 1). The vital
signs were normal. There were prominences of the skull. The right eyeball was depressed, and consensual eye motion was disturbed. The chest cage revealed lumbar kyphosis and severe thoracic scoliosis. The heart was normal. The abdomen was compressed between the thoracic cage and the pelvis. There was restriction of motion in both elbow joints, the left being most severely affected. The left hip was extremely deformed by shortening of more than 5 inches. There were numerous large bony masses over the left humerus, and there was restricted motion of the left shoulder. There was decreased sensation to pinprick below the sixth thoracic vertebral level. Plantar reflexes were extensor bilaterally. Vibration sense was absent; proprioception was absent in the legs. Light touch was impaired. Deep tendon reflexes of the ankles and knees were brisk.

X-ray examination showed multiple bony lesions compatible with polyostotic fibrous dysplasia (Fig. 2). Laboratory studies revealed a normal leukocyte count and hematocrit value. The initial alkaline phosphatase value was 130 King-Armstrong units. The initial serum calcium content was 11.8 mg/100 ml, and the serum phosphorus content was 3.2 mg/100 ml. Urinalysis was normal. The blood sugar content was 78 mg/100 ml in the fasting state. The cerebrospinal fluid was xanthochromic, with 1,000 erythrocytes and no leukocytes. The spinal fluid protein content was 297 mg/100 ml. The spinal fluid serologic test was negative. Culture of the spinal fluid was sterile. Pantopaque myelography showed a partial block at the fourth thoracic vertebra (Fig. 3).

Operation. On March 11, 1966, spinal cord decompression was accomplished by means of laminectomy from the third to the sixth thoracic vertebrae. A large mass of soft gritty tissue that filled the epidural space at these levels was removed. Frozen section showed the presence of fibrous dysplasia (Fig. 4) which was verified in the permanent histologic sections.

Postoperative Course. The patient had an uneventful recovery and on the eighth postoperative day was able to move the legs and demonstrate good strength. Perception of pinprick and touch, although depressed, was improved. The reflexes remained hyperactive, and bilateral extensor plantar reflexes were present. Coarse involuntary movements of the legs persisted. Physical therapy was begun on the tenth postoperative day. On March 22, determinations showed a serum calcium content of 11 mg/100 ml and a serum phosphorus content of 3.3 mg/100 ml; acid phosphatase was 14 units; and the serum alkaline phosphatase content decreased to 71 King-Armstrong units. Urinary...
content of hydroxyproline was 129.7 mg/24 hr. The patient was discharged from the hospital the twelfth postoperative day.

When seen 18 months after operation, the patient was walking and able to take care of her elderly father.

**Discussion**

Fibrous dysplasia affecting the spine is rather rare. Furst and Shapiro\(^6\) found the spinal canal obliterated by masses of fibrous tissue. They reported one case of involvement of the lumbar spine. Albright and Reifenstein\(^3\) reported two cases in 1948. Dockerty and associates\(^4\) in a series of six cases found two instances of involvement of the dorsal spine. Warrick\(^15\) reported one case in which the fifth lumbar vertebra was affected. Schlumberger\(^12\) reviewed 67 cases of monostotic fibrous dysplasia in soldiers and found one case in which the fourth cervical vertebra was affected.

Epstein\(^2\) for several years followed the progress of a patient who had monostotic fibrous dysplasia involving the cervical spine without significant neurological deficit. Van Horn, *et al.*,\(^14\) reported 57 cases, seven of which had vertebral involvement.

Spinal cord compression caused by polyostotic fibrous dysplasia affecting the vertebra is even more rare. Jaffe\(^8\) reported one case of a 37-year-old woman who had signs of cord compression at the dorsal level; she was operated on twice and made substantial recovery. This same case was also reported by Teng, *et al.*\(^13\)

Rosencrantz\(^11\) found seven cases of spinal cord compression in the literature up to 1965, and reported the case of an 18-year-old boy with paraplegia due to fibrous dysplasia affecting the dorsal spine.

**Summary**

We have reported the case of a 56-year-old woman with polyostotic fibrous dysplasia affecting the thoracic spine and causing compression of the spinal cord. Decompressive laminectomy and excision of a fibrous epidural mass were followed by neurological improvement.
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References