COMPRESSION OF SPINAL CORD BY OSTEITIS DEFORMANS (PAGET’S DISEASE), GIANT-CELL TUMOR AND POLYOSTOTIC FIBROUS DYSPLASIA (ALBRIGHT’S SYNDROME) OF VERTEBRAE

A REPORT OF FOUR CASES

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Spinal cord compression by a lesion of the vertebra is most often the result of traumatic injury, metastatic carcinoma, infiltrating sarcoma, tuberculous spondylitis, pyogenic osteomyelitis, osteoarthritis, infiltrating leukemia or multiple myeloma. Primary osteogenic sarcoma and benign neoplasms of the spine rarely cause compression of the spinal cord. Although osteitis deformans (Paget’s disease) is not an uncommon disease and frequently involves the spine, it rarely causes compression of the spinal cord. Polyostotic fibrous dysplasia belongs to the rare category of bone disease. The vertebrae are scarcely ever affected, and involvement of the spinal cord has never before been recorded in this condition to our knowledge.

OSTEITIS DEFORMANS

Among 274,397 admissions to the Mount Sinai Hospital in a period of 17 years from October 1933 to October 1950, 131 were for osteitis deformans, an incidence of approximately 1 to every 2,000 admissions. Of these patients, only 1 had paraplegia as a result of spinal cord compression by the hypertrophic vertebrae. Following is a brief report of this case:


Fifteen months before admission the patient began to have difficulty in walking, associated with numbness of both lower extremities. For the last 3 weeks he had been unable to walk unless assisted and could not stand without support.

Examination. There was marked weakness in both lower limbs, especially the left. The ankle jerk was absent on the left and markedly exaggerated on the right. Other deep tendon reflexes of the lower limbs were all exaggerated but equal on both sides. Signs of Babinski, Chaddock, Oppenheim and Rossolimo were elicited on both sides. Hypalgesia was demonstrated from below D4. Vibration sense was absent from below the D4 spinous process. Deep and position sensation were lost from below the ankles. Abdominal reflexes were all present and equal on both sides. There was a kyphosis of the upper dorsal spine.

Laboratory findings: Hb. 15 gm., WBC 6,550 with 61 per cent neutrocytes. E.S.R. 15 mm./hr., fasting sugar 140 mg. per cent, urea nitrogen 17 mg. per cent,
serum albumin 3.4, globulin 3.0, acid phosphatase 8 KAU, alkaline phosphatase 40 KAU, cholesterol 480 mg. per cent, phosphorus 3.1 mg. per cent, and calcium 10.5 mg. per cent.

Roentgenograms of spine and pelvis revealed loss of normal homogeneous cortical density of the bones of the pelvis and almost all of the vertebral bodies. The cortex was thickened and the trabeculae were markedly coarsened by a wavy thick bony meshwork with radiolucent interstices. Small cystlike areas were scattered between the heavy trabeculae. There was marked flattening of the D3 vertebra with a similar coarsened trabecular pattern and biconcavity of its upper and lower surfaces (Fig. 1). The skull showed several small areas of decreased density as well as a generalized sclerosis. Diagnosis: Generalized osteitis deformans (Paget's disease).

Course. On the 11th hospital day, myelography was done. Before the instillation of pantopaque, manometric test disclosed a complete block of the CSF flow. The fluid was xanthochromic. The myelogram revealed a complete block to the cephalad flow of the pantopaque at the level of the D3 vertebra (Fig. 2). There was an irregular rounded capping to the proximal end of the column, suggesting an extradural lesion.

Operation. On the next day laminectomy was

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Fig. 1. Case 1. Paget's disease. Posteroanterior view of dorsal spine showing coarsened trabecular pattern of the upper segments.

Fig. 2. Case 1. Paget's disease. Myelogram showing obstruction at level of D3.
performed with the removal of D1, D2 and D3 laminal arches, which were soft, vascular, and about 1½ cm. in thickness. The spinal cord did not pulsate until the thickened laminal arch of D1 had been completely removed. Epidural probing with a small-calibre rubber catheter encountered no obstruction in the neighboring segments of the spinal canal. Intradural exploration disclosed a thickened arachnoid membrane and fine fibrous adhesions.

Postoperative Course. The right leg was completely paralyzed. The power of the left lower leg was further reduced. There was complete anesthesia from below the level of D3. However, the strength of the left lower limb slowly improved. On the 37th postoperative day, he could raise his left leg and sensation on the right side of the body and leg recovered completely.

One week later, the power of the left leg was about normal. He could raise his right leg off the bed. However, sensation remained absent on the left from below D4. Lumbar puncture revealed a CSF pressure of 80. The fluid was clear and contained 72 mg. per cent total protein. Manometric test showed that there was no obstruction.

On discharge, 85 days after operation, the patient was walking with crutches. Pathological Diagnosis. Osteitis deformans (Fig. 3).

Fig. 3. Case 1. Paget’s disease. Section of bone removed from lamina. There are dense trabeculae among vacuolated tissue and dilated vascular sinuses. Hematoxylin-eosin stain, X100.

Comment. The paraplegia in Case 1 was caused by the hypertrophic laminal arches of D1, D2 and D3. The laminectomy benefited the patient. However, recovery of function has been slow, but progressive.

Osteitis deformans is a chronic disease and does not in itself endanger life. The patients may live for many years, dying either from its complications or other causes. They may be unaware of having the disease for many years and usually do not enter a hospital unless a complication develops.
Paget's original observation that the most frequent sites of osteitis deformans are in the skull or tibia has been disproved. Careful anatomic study by Schmorl in 190 autopsies proved that osteitis deformans occurs most frequently in the sacrum, spine, and femur. Since Sir James Paget first described the disease in 1876, numerous cases of involvement of the central nervous system, such as platybasia, convexobasia, optic atrophy, deafness and trigeminal neuralgia as a result of destruction and distortion of the base of the skull, have been recorded. Nevertheless, cord compression is rare and it was not until 1923 that the first real evidence of the production of spinal cord symptoms by Paget's disease was brought forward by Wyllie, who reported 2 cases. In 1939, Schwarz and Reback reported 9 cases of paraplegia due to osteitis deformans of the spine. In 1940, Turner added another series of 7 cases to the medical literature. From 1923 up to the present, a total of 37 cases have been recorded (Table 1).

Only 3 of these 37 patients were females. The oldest was 77 and the youngest was 43 years of age. The longest duration of Paget's disease before the onset of neurological symptoms was 20 years. In our case the compression of the spinal cord was the first symptom of the illness.

Osteitis deformans involves the lumbar spine more frequently than the dorsal. In 69 autopsied cases in which the spine was involved, Schmorl found 36 in the lumbar, 23 in the dorsal and 10 in the cervical spine. Spinal cord compression occurred most frequently when the thoracic region was involved. As shown in Table 1, in 31 of these 37 collected cases, the compression of the cord was at the level of the dorsal vertebrae. In only 3 instances was the lesion localized in the cervical segments, and in the remaining 3 the level was not mentioned. However, 2 of these patients had paraplegia and 1 had paraparesis; presumably the dorsal spine was the site of compression.

As far as treatment is concerned, there is no choice other than the removal of the pressure by a laminectomy. In 20 of the 37 reported cases laminectomy was performed. Fourteen patients improved, 3 died from postoperative complications, 2 did not improve, and 1 was able to walk for 3 years and then again became paraplegic. Of the patients who were not operated upon, none improved.

GIANT-CELL TUMOR

In 17 years, 2 cases of giant-cell tumor of the spine with compression of the cord were encountered at the Mount Sinai Hospital. One has been reported previously, and is therefore not included in the present series. The patient was a 22-year-old woman. Spastic paraplegia developed 8 days before admission. Laminectomy of D2 to D4, with the removal of the tumor which involved the lamina of D2, restored her motor power. She made a complete recovery.

The other case is reported below (Case 2). In this instance the tumor of the vertebra involved the cauda equina.
### TABLE 1
Cases of Paget's disease with spinal cord compression reported from 1923 to 1951.

<table>
<thead>
<tr>
<th>No.—Author</th>
<th>Date</th>
<th>Sex</th>
<th>Age</th>
<th>X-ray Findings &amp; Neurological Signs</th>
<th>Treatment &amp; Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>3. Ardin-Delteil et al.</td>
<td>1923</td>
<td>M</td>
<td>55</td>
<td>Spastic parapleg, in flex. Hypal. below pubis.</td>
<td>No specific treatment</td>
</tr>
<tr>
<td>9. Garcin et al.</td>
<td>1937</td>
<td>M</td>
<td>61</td>
<td>Brown-Séquard, level C3–4</td>
<td>No treatment</td>
</tr>
</tbody>
</table>
### TABLE 1—Cont.

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<tr>
<th>No.—Author</th>
<th>Date</th>
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While the patient was at work pushing a hamper, 2 1/2 years before admission, a sharp pain suddenly developed in the middle and lower part of the back. Two weeks later the pain became worse, and he was unable to work. A few months later the pain was much less than before; however, he was still incapacitated. Five weeks before admission, the pain became more severe. One week later his left leg felt heavy and weak, and he could not stand on it. Two weeks before admission the weakness involved the opposite leg.

Examination. There was a marked limitation of motion of the spine in any direction. The muscles of the back were spastic. Tenderness was elicited over the spinous process of L3. His gait was slow, awkward and unsteady. There was weakness in both legs, especially the left. Both ankle jerks were absent. Hypalgesia was found below L1 on both sides. There was muscular atrophy of the lower lumbar group, the calves and the thighs.

Laboratory findings were noncontributory. X-ray of the lumbar spine disclosed an expanding lesion involving the left side of L3 laminal arch. There was evidence of bone destruction associated with irregular patterned new bone formation.
*Course.* On the day after admission a lumbar puncture was done. The CSF was clear, containing 3 lymphocytes. Pandy test was 2+. Manometric study showed that there was no block.

*Operation.* On the 5th hospital day, laminectomy of L2 and L3 was performed by Dr. Ira Cohen. The tumor was found to involve the laminar arch, the left pedicle and part of the vertebral body of L2. It was reddish in color and very vascular. It extended into the epidural space and encroached upon the cauda equina. Some newly formed bone tissue was seen inside the tumor.

*Course.* Two weeks after operation, radiotherapy was initiated. Four weeks later the patient was discharged. All neurological signs were normal. He was last seen on April 5, 1949 and remained asymptomatic.

*Pathological Diagnosis.* Giant-cell tumor of the spine.

In the second case of giant-cell tumor included in this report (Case 3), the patient was treated by one of us at the Beth Israel Hospital.


One year before admission, the patient noted weakness in both arms and legs associated with pain in the back of the neck radiating down to the arms. Six weeks before admission the pain became worse and 4 weeks later she was unable to stand. A feeling of girdle-like constriction had developed around the upper chest.

*Examination.* There was tenderness over the spinous process of C6 and C7. The neck was held stiff with limitation of motion. There was weakness of all four limbs with atrophy of the intrinsic muscles of both hands. Deep tendon reflexes of all extremities were markedly exaggerated. Abdominal reflexes were absent. A zone of hyperesthesia was elicited at the level of C6. Below this zone there was hypalgesia. Vibration sense was impaired in both legs.

Laboratory findings: CSF pressure was 180 mm., with partial block on manometric test. The fluid contained 5 WBC and 128.6 mg. per cent total protein.

X-ray of cervical spine disclosed a compressed fracture-dislocation of C6. Biopsy of the spine was reported as benign giant-cell tumor (Fig. 4).

*Operation.* On the 8th hospital day, laminectomy of C5, C6 and C7 was done.

*Course.* Six days after operation a cast was applied and x-ray treatment was initiated. She made a slow but satisfactory improvement in regaining the function of her limbs. On discharge she was walking about, using a walking aid.

*Comment.* Giant-cell tumor is relatively uncommon, probably half as frequent as primary malignant bone tumor. Its occurrence in the spine is comparatively rare. Kolodny,\textsuperscript{16} in an analysis of the material of the Bone Registry in 1927, found that giant-cell tumor occurred in the spine in 8 per cent of the cases. It affects mainly the younger group of patients, in the second and the third decades of life.

Of 124 patients with giant-cell tumor seen at the Mayo Clinic\textsuperscript{19} from 1916 to 1940, in only 2 was the tumor located in the vertebra and none had signs of spinal cord compression. Coley\textsuperscript{3} listed 509 cases of giant-cell tumor in his book *Neoplasms of Bone*; only 3 were found in the spine.

It is now generally agreed that while the majority of giant-cell tumors are benign, some are either malignant from the onset or undergo malignant
changes. Of the 124 cases reported from the Mayo Clinic, in 101 the lesions were benign.

Since this tumor is susceptible to x-ray treatment, it is usually treated before symptoms of spinal cord compression arise. Only a few cases of giant-cell tumor involving the spinal cord or cauda equina have been reported. Lewis, in reviewing the literature to 1924, found 17 cases of primary giant-cell tumor of the vertebra. In 11 of these patients there were signs of pressure on the spinal cord or cauda equina. In 1930, Santos collected 3 cases of giant-cell tumor of the spine, and reported 1 of his own with cauda equina compression. Brock and Bogart, in 1945 collected 91 cases, and added 1 with spinal cord compression at the cervical segment. In 1947 Verbiest recorded another case of cord compression. Therefore, we have been able to collect 17 cases from the literature including the 1 reported by Kaplan in 1949 and our own 2 cases.

Laminectomy for decompression, followed by radiotherapy, gave satisfactory results in 6 cases recently reported.

POLYOSTOTIC FIBROUS DYSPLASIA (ALBRIGHT'S DISEASE)

A perusal of the literature failed to disclose a case of spinal cord compression due to ostotic fibrous dysplasia. The following is a case of polyostotic fibrous dysplasia involving the mid-thoracic vertebrae, associated with paraplegia.

Case 4. B.L., female, aged 37. Polyostotic fibrous dysplasia involving D7 and D8
with spinal cord compression. Laminectomy of D5, 6, 7. Complete recovery. Recurrence of paraplegia 15 months later. Re-exploration from D4 to D7. Slow recovery.

In 1929 the patient noted a small tender tumor on the right side of the posterior chest wall, and a similar one on the left side, anteriorly situated. Later a section of one rib was removed and a diagnosis of fibrous dysplasia was made.

In December 1942, while walking, she suddenly had weakness and numbness in both legs, accompanied by severe pain in the posterior mid-thoracic region. The numbness was more marked in the right foot, and the weakness was progressing to the time of admission 2 weeks later.

_Examination._ There was a large bulging tender mass over the 7th and 8th ribs on the right side. Tenderness was found over the spinous processes from D4 to D9 and a kyphos at D6 and D7. The gait was slow and unsteady, and there was weakness of both lower extremities. There was no definite sensory change except the loss of vibration sensation in both feet. Deep tendon reflexes of the lower limbs were all exaggerated.

_Laboratory findings_ were noncontributory.

_Roentgenograms_ revealed a marked deformity of the posterior portion of the right 7th and 8th ribs. There was great expansion of the shafts of the bones and marked thinning of the cortices, with areas of translucence suggesting irregular cyst formation. There was an irregular wavy coarse trabeculation within the expanded bone, and a kidney-shaped mass with calcific margins extended into the soft tissue of the lateral thoracic wall. There were similar changes in the left 7th rib posteriorly, and in the bodies and arches of the 7th and 8th dorsal vertebrae. There was anterior and lateral compression of these vertebral bodies, and narrowing of the 6th and 7th interspaces. The expanded portion of the posterior inferior margin of the 7th dorsal body encroached considerably into the spinal canal (Fig. 5).
Lumbar puncture yielded CSF containing 96 mg. per cent total protein. Complete block was demonstrated on manometric test.

**Course.** Two weeks after admission, the pain in her back suddenly became worse, and the weakness of both legs increased. Babinski sign and ankle clonus were elicited on both sides. A zone of hypalgesia was found at the level of D6 and D7. Deep sensation was absent in both lower limbs from below the iliac crests.

**1st Operation.** On the 17th hospital day, a laminectomy of D5, 6, 7 vertebrae was performed by Dr. Ira Cohen. Fibrous tissue was seen within the laminal arches of D5 and D6. An irregular-shaped extradural tumor, $\frac{3}{4} \times 1\frac{1}{2}$ inches in size, was removed from D5 and D6 portion of the spinal canal.

**Course.** Two weeks after operation, the abnormal findings had disappeared and the patient was wearing a brace and walking with a normal gait.

Fifteen months after discharge, paraplegia suddenly developed and she was re-admitted.

**2nd Operation.** The spinal canal from D4 to D7 was re-explored and an extradural tumor was removed from D4.

**Course.** The patient made a slowly progressive recovery and, 6 weeks later, she again was able to walk with support. She was seen 1 year later at the follow-up clinic, walking without difficulty. In 1949 she was admitted to the medical service on three occasions because of hypertensive cardiac failure. However, repeated neurological examinations revealed no abnormalities.

**Pathological Diagnosis.** Polyostotic fibrous dysplasia (Fig. 6).

**Comment.** The bone involvement in Case 4 was bilateral, multiple, extensive and included the vertebrae. The diseased tissue protruded into the

![Image](image_url)
spinal canal from the affected vertebrae and encroached upon the spinal cord, which had been already embarrassed by the angulation of the spine caused by collapse of the vertebrae. Laminectomy and removal of the epidural portion of the dysplastic tissue rapidly and completely restored the motor power of both lower extremities. A similar but slower result was obtained by the second laminectomy which was done because of recurrence of symptoms.

Unlike osteitis deformans, fibrous dysplasia appears to be a congenital disorder of the bone-forming mesenchyme. It usually affects children or adolescents, and females more than males. It is a rare disease, characterized by bone changes, cutaneous pigmentation and sexual precocity. It was first described by Albright and his associates. The disease has a tendency to involve bones of one side of the body. When more than one bone is involved, it is called "polyostotic fibrous dysplasia." If the change is limited to only one bone it is known as "monostotic fibrous dysplasia."

Ostotic fibrous dysplasia involves usually the diaphysis or metaphysis of the long bones and rarely occurs in the epiphysis. The bones most frequently involved are the femur, tibia, humerus and radius. Similar to osteitis deformans, it causes bending deformities. The affected area of the bone is cystic in appearance and contains giant-cells, and abundant fibrous and cartilaginous tissues.

In 1942 Lichtenstein and Jaffe reviewed 90 cases of fibrous dysplasia. In only 1 patient were the vertebrae involved. Schlumberger studied cases of fibrous dysplasia involving a single bone, and in only 1 case the cervical spine was affected. However, in neither was there evidence of spinal cord compression.

**SUMMARY**

1. One case of Paget's disease of the vertebrae with spinal cord compression at D1 and D2 is presented. Laminectomy benefited the patient.

2. Two cases of giant-cell tumor of the vertebra are reported, one with involvement of the cauda equina, and the other with cord compression. Laminectomy resulted in complete recovery of both patients.

3. One case of polyostotic fibrous dysplasia involving the dorsal spine with paraplegia is presented. The patient recovered after laminectomy.

4. The related literature of the aforementioned 3 conditions has been reviewed.

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