CASE REPORTS

SPINAL-CORD COMPRESSION SECONDARY TO GAUCHER'S DISEASE

CASE REPORT

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Fig. 1. Normal anteroposterior and lateral views of thoracolumbar spine, April 1961.

Gaucher's disease is a lesion of the reticuloendothelial system. Normal cells are replaced by histiocytes containing the cerebrosidase keratin.2,7 When Gaucher first described this disease in 1882 the marked prevalence of involvement of bones was not appreciated. In 1909 Risel6 first noted the characteristic microscopic lesions of bone in Gaucher's disease. Klercker4 and Junghagen3 did the first complete roentgen-ray studies in this disease. Since then the true incidence of the skeletal lesions has been recognized.10 Indeed, Snapper has stated that the marrow of the bone is always involved.8,9 Certainly after splenic involvement, osteoarticular pathology causes the most important signs and symptoms.2 Though the usual site of pathology of the bone is the femur, the vertebral bodies are involved frequently.1,5 Kerasin-laden histiocytes invade and destroy normal trabecular structure with resultant vertebral collapse and gibbus formation. It would be expected that spinal-cord compression may occur following such collapse. About 300 cases of Gaucher's disease have now been described in the world literature but there does not appear to be

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any report of spinal-cord compression following vertebral collapse. Such a case was seen recently at the Neurological Institute of New York and forms the basis of this report.

**CASE REPORT**

*History.* The patient was a 41-year-old Jewish female in whom a diagnosis of Gaucher's disease was made in 1947 and a splenectomy was carried out. In 1951 pain in the back and hip developed, radiating into the right thigh. On neurological examination there was some weakness of muscles of the right thigh, and the right knee and ankle jerks were depressed slightly. Roentgenograms of the spine and right leg were normal. Symptoms disappeared in 2 months. This pain recurred in December 1953 and roentgenograms then showed irregular translucencies and increased densities of the right pubic bone, hip and sacrum. There was minimal weakness of flexion of the right hip; the remainder of the neurological findings were normal. Again symptoms subsided spontaneously. In 1955 pain recurred. On examination there was limitation of motion of the right hip. There were no
neurological abnormalities. Roentgenograms revealed involvement of the right sacroiliac joint in addition to the findings described previously. The patient was given a course of radiotherapy to the innominate bone and femur. Relief from pain was dramatic but the roentgen-ray appearance of the area was unchanged.

In March 1959 a nontoxic nodular goiter infiltrated with Gaucher's cells was removed. A traumatic fracture of the supracondylar portion of the right femur occurred in December 1959, and of the right tibia and humerus in July 1960.

In April 1961 the patient was admitted to the orthopedic service because of lumbosacral pain of 40 hours' duration.

Examination. There was tenderness of the L5-S1 area. Movement of the legs was limited by pain but there did not appear to be any weakness. Sensory findings were normal and reflexes were active and equal. Plantar responses were flexor in type. Roentgenograms of the spine were normal (Fig. 1).

Course. The pain subsided with rest in bed. She was discharged 4 days later to be followed in the outpatient clinic. Pain was present intermittently until June 28, 1961 when the patient first noted radiation of the pain and numbness down the left leg, and had difficulty in standing. She was seen in the outpatient clinic where marked tenderness was observed on palpation of the lumbosacral area. Motor and sensory findings were normal. There were no deep tendon reflexes in either leg. Roentgenograms showed almost complete collapse of L1 vertebra (Fig. 2), and it was felt wise to admit the patient to the orthopedic service.

Neurological examination on June 30, 1961 disclosed moderate weakness in all groups of muscles in the legs. Sense of position was decreased in the toes and sensation of pain and touch was reduced from the L1 dermatome caudally. Several hours later sensory level was found to start at the T11 dermatome. Weakness in the legs was marked and was greater proximally than distally. Deep tendon reflexes were absent in the legs and Babinski's response was present bilaterally. Myelography demonstrated a complete block to the flow of contrast medium at L1 (Fig. 3).

Operation. A decompressive laminectomy was done immediately. The lamina of T12 was removed first. At the lower end of the exposure the dura mater was compressed because of a sharp angulation in the spinal canal. The lamina of L1 was then removed and the dural sac was seen to be angulated at about 30° with a sharp constriction at T12-L1 level. After the L1 lamina had been removed the dura mater pulsated well and the sharp angulation assumed a gentle curve. A biopsy of the bone was taken. The orthopedic service fused the 12th thoracic to the 3rd lumbar vertebrae to complete the procedure.

Pathological Examination. Microscopic sections of the surgical specimen showed that the marrow spaces were largely replaced by numerous cells which were large, often polygonal or fusiform, and were closely approximated to one another (Fig. 4). With hematoxylin and eosin stain the abundant cytoplasm was pale pink and finely fibrillar. The nuclei of the cells were relatively small, round and uniform and often were located centrally (Fig. 5). With Masson's trichrome stain the cyto-

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**FIG. 4.** Bone with marrow spaces largely replaced by numerous large, often polygonal, or fusiform cells. Hematoxylin and eosin stain, ×105.
plasm of the cells showed fine, reddish granules in fibrils on a hazy, pale gray-blue ground.

Postoperatively, the patient’s neurological status improved markedly. When seen several months later there remained only mild residual weakness in the legs and the patient walked well. At follow-up visit to the clinic in March 1962 there was no essential change in her condition.

DISCUSSION

Lesions of the bone occur frequently, if not always, in Gaucher’s disease. Vertebral-body involvement by Gaucher cells predisposes to bony collapse and gibbus formation. Though neurological complications secondary to such collapse appear to be very rare, spinal-cord compression can occur. This possibility must be kept in mind in the presence of advancing neurological changes.

SUMMARY

A case of Gaucher’s disease involving the thoracolumbar vertebrae is reported. Collapse of T12 with subsequent spinal-cord compression occurred. No previous reports of such an occurrence could be found.

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REFERENCES