EOSINOPHIL GRANULOMA OF BONE
IN TWO ADJACENT THORACIC VERTEBRAE

Report of a Case

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For many years it has been considered that eosinophil granuloma of bone, Hand-Schüller-Christian disease and Letterer-Siwe disease are different manifestations of the same condition (Lichtenstein and Jaffe 1940; Otani and Ehrlich 1940; Farber 1941; Jaffe and Lichtenstein 1944; Engelbreth-Holm, Teilum and Christensen 1944; Lightwood 1955). Lichtenstein (1953) grouped these diseases under one heading which he called histiocytosis X.

It is generally accepted that in the young and in persons in whom the disease is widespread (Letterer-Siwe) the prognosis is poor. Older children and young adults appear to have a better prognosis.

The most benign form of the condition is when the lesions are confined to bone, be they single or multiple (eosinophil granuloma). These may, however, spread to involve skin and other soft tissues, and conversely the Letterer-Siwe and Hand-Schüller-Christian forms may occur without bone involvement.

Solitary bone lesions may occur almost anywhere, except perhaps the phalanges of the hands and feet (Hodgson, Kennedy and Camp 1951), the most common sites being the ribs, the pelvis, the femur, the vertebrae, the tibia, the humerus and the scapula. Involvement below the elbow and knee is uncommon. This report describes an unusual vertebral lesion presenting with spinal compression which later was relieved by operation.

CASE REPORT

A bricklayer of thirty-three had complained for a year of pain in the small of the back which extended down the left buttock, the thigh and the leg. Recently the pain had begun to involve the right leg in a similar distribution. For three months he had been aware of numbness of both legs down as far as the knees and had noticed some difficulty in walking. His general health was good. There was no history of back injury, and he had no complaint with regard to his bowels or urinary function.

On examination he was a well built man with a wide based, mildly spastic gait. There was spasticity of his left leg, but the tone in the right leg was normal. There was generalised weakness of both legs involving all muscle groups. Both knee jerks were absent, but the ankle jerks were present. The left plantar response was extensor and the abdominal reflexes were normal. No sensory level could be detected, there was no saddle anaesthesia, and position sense in both legs was normal. No defect of the spine was visible on clinical examination, but on spinal movement there was spasm of the erector spinae muscles on the left side.

Radiographs of the thoraco-lumbar spine showed erosion of the left pedicles of the tenth and eleventh thoracic vertebrae, and erosion of the body of the eleventh thoracic vertebra (Fig. 1). The intervertebral disc space was normal.

Lumbar puncture drew clear fluid, but manometry demonstrated a complete block. The protein content of the fluid was 180 milligrams per cent.

Operation—On the left side the laminae were deficient and once the muscle was retracted, caseous, necrotic looking tissue was seen. A limited hemilaminectomy upwards and downwards was carried out on this side to give adequate exposure and to define normal tissue margins.
FIG. 1
Radiographs showing the lesions in the bodies of the tenth and eleventh thoracic vertebrae.

FIG. 2
Photomicrograph of the excised material showing xanthomatous tissue. (× 200.)

FIG. 3
Photomicrograph of the excised material showing granulomatous tissue. (× 200.)
The abnormal tissue mass was easily defined from the surrounding tissues and was found to be displacing the dura and its contents towards the right side. The mass was easily removed with pituitary forceps and by suction. It was caseous and yellow in appearance but was not adherent to the dura.

**Pathological examination**—This showed large islands composed of foam cells (Fig. 2) with shrunken, often eccentric, nuclei alternating with areas of granulomatous appearance (Fig. 3), where the cells were plump and spindle-shaped and were associated with the formation of collagenous bands. Occasional giant cells were present and there was a sparse infiltration with plasma cells; eosinophils were inconspicuous. Transitional cells whose cytoplasm contained a peripheral ring of lipoid were identified. In frozen sections most of the foam cells contained granules staining orange with Scharlach R, the remainder staining bright red, consistent with cholesterol and neutral fat respectively. The orange granules were doubly refractile, and gave a doubtful Shultz reaction.

A diagnosis of eosinophilic granuloma of bone was made, and in view of this further tests were carried out. A complete skeletal survey did not reveal other areas of bone erosion. The serum cholesterol was 150 milligrams per cent, the plasma proteins were 7.7 grammes per cent with a normal electrophoretic pattern, and in particular there was no increase in the alpha fraction.

The patient had an uneventful convalescence and was discharged a month after admission when all his symptoms had disappeared. He walked normally, the spasticity of the left leg had gone and the extensor plantar response had become flexor. Only the knee jerks remained absent.

**DISCUSSION**

In the case described the lesion was confined to two vertebrae without other bone involvement. Arcomano, Barnett and Wunderlich (1961) described the case of a girl of two and a half who had pain in the small of the back and the legs. There were, however, no abnormal neurological signs. Radiographs showed a destructive lesion of the body of the fourth lumbar vertebra but the child got better without treatment.

In many cases with vertebral involvement previously described, the disease process was classified as the Letterer-Siwe or Hand-Schüller-Christian variations as there was acute or chronic systemic involvement as well (Moe and Hansen 1960, McCulough 1951).

Reports of solitary eosinophilic granuloma of bone include a series of nine cases by Hamilton, Barner, Kennedy and McCort (1946), and a series of forty cases by Avery, McAfee and Guild (1957): nine of these were classified as solitary eosinophilic granuloma of bone. In neither series of solitary bone lesions were the vertebrae involved.

Green and Farber (1942) described ten cases with bone involvement only. In two of these the vertebrae were involved but in both instances many other bones were also affected.

Arcomano et al. (1961) described a girl of three and a half who presented with left facial weakness, weakness of the abdominal muscles and a painful, weak right hip. Radiographs showed multiple bone lesions with involvement of several vertebrae. The case proved to be the Letterer-Siwe variety as there was acute systemic involvement, and she died. We could trace only one other similar case involving vertebrae only and which was treated by operation. MacNab (1955) described a case of a girl of two with a solitary lesion in the second cervical vertebra presenting with stiffness of the neck. There were, however, no abnormal neurological signs. Operation was carried out and four months later the child was better.

Finally, in accordance with most of the reported cases, our patient was uncommonly old to have developed eosinophilic granuloma, the disease most usually affecting infants and young children.
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SUMMARY

A case of eosinophilic granuloma of bone in a man of thirty-three, involving the tenth and eleventh thoracic vertebrae, is described. The presenting symptom was pain and he had spinal compression with spastic paraparesis. Improvement followed operation. The operative findings and the histological features are described. The literature and the unusual features of this case are discussed.

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REFERENCES


Lichtenstein, L. (1953): Histiocytosis X. Archives of Pathology, 56, 84.


