Spinal cord compression by extramedullary haematopoiesis in myelosclerosis

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Spinal cord compression by extradural deposits is not uncommon in myeloma (Clarke, 1956; McKissock, Bloom, and Chynn, 1961), leukaemia (Wilhyde, Jane, and Mullan, 1963), and Hodgkin's disease (Bhagwati and McKissock, 1961). Cord compression by the malignant lymphomas is discussed by Love, Miller, and Kernohan (1954) and by Hutchinson, Leonard, Maudsley, and Yates (1958).

Close, Taira, and Cleveland (1958) reported a case of myelosclerosis in which an extradural mass of haematopoietic tissue compressed the spinal cord. They were unable to find any reports of similar cases in a review of the literature over the previous 40 years and we have not found any recorded subsequently. We report another patient with this rare complication of myelosclerosis, in whom surgical treatment led to gratifying clinical improvement.

CASE REPORT

The patient was a 58-year-old housewife whose past history was uneventful except that in 1939 tuberculous neck glands were excised. In 1962 she had an epithelioma of the left wrist removed. At that time the spleen was noted to be enlarged and a blood count showed a haemoglobin of 15.8 g./100 ml. and a white cell count of 16,100/c.mm. with 3% of myelocytes. Serum proteins, serum bilirubin, erythrocyte sedimentation rate, and erythrocyte fragility were normal; the direct Coombs test and Paul-Bunnell test were negative.

In March 1963 examination of the sternal marrow showed no abnormality. Soon afterwards the patient fell on her back and had some difficulty in walking, but this improved during the next three weeks, although she could still not walk upstairs normally. Over the ensuing three months her legs became insidiously stiffer, and numbness spread from the feet to her groins and buttocks and by June 1963 she could only totter. She had no urinary symptoms. She was referred to the Cumberland Infirmary, where a paraparesis with impaired sensation below the second lumbar dermatome was found. Radiographs showed bone change in the bodies of the third, eleventh, and twelfth dorsal vertebrae and in the lower ends of the

femora, suggestive of myelosclerosis or leukaemic infiltration (Fig. 1). Spinal puncture yielded xanthochromic fluid under 90 mm. pressure with no rise on jugular venous compression and a protein content of 235 mg./100 ml. She was transferred to the Department of Neurology of the Royal Victoria Infirmary.

On examination she had a plethoric face and a small nodular goitre of the left lobe of the thyroid (present for 20 years), but no lymphadenopathy. The spleen was firm and filled most of the left upper abdominal quadrant, but the liver was not palpable.

The patient was able to stand and take a few steps with support. The cranial nerves and upper limbs were normal.

FIG. 1. Subperiosteal new bone formation and rather coarse trabecular pattern of the femur.

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In the lower limbs tone was flaccid and power extremely poor peripherally, although she retained some hip flexion. Appreciation of pain, touch, and temperature was diminished below the first lumbar dermatome bilaterally. There was loss of vibration sense below the iliac crests, and positional sense was impaired in the feet. The abdominal reflexes were absent and the knee and ankle jerks were depressed. There was no plantar response.

INVESTIGATIONS

HAEMATOLOGY The blood, bone marrow, and splenic pulp were examined by Dr. W. Walker who found the haemoglobin to be 17.2 g./100 ml. and cell volume 55% giving a mean corpuscular haemoglobin concentration of 31.5%. The reticulocyte count was 4%, platelet count 221,100/c.mm., and white cell count 15,300/c. mm. (polymorphs 75%, lymphocytes 16%, monocytes 5%, eosinophils 2%, normoblasts 2%). The erythrocyte sedimentation rate was 1 mm. in the first hour (Westergren). The red cells showed anisocytosis, some poikilocytes, and polychromasia. The white cells were normal but there was a shift to the left in the granulocyte series to the myelocyte level. Platelets were plentiful with some giant forms. A buffy coat film showed more obvious shift to the left and a few myeloblasts were seen.

Sternal marrow was obtained easily but was not very cellular and fibrous tissue was slightly excessive. Erythropoiesis was normoblastic and there was a little shift to the left in the myeloid series. The megakaryocytes were normal. The splenic pulp showed myeloid metaplasia and the films could not be distinguished from the bone marrow films. Most leucocytes were alkaline phosphatase positive.

Dr. Walker thought that this was a myeloproliferative disorder displaying the features of myelosclerosis and that the findings were against a diagnosis of leukaemia.

RADIOLOGY A chest radiograph was normal. Myelography (A.A.) revealed a complete block to the flow of myodil at the lower border of the twelfth dorsal vertebra where there was abnormal bone change consisting of vertical striation and possibly some subperiosteal new bone formation. The bone changes demonstrated in the previous films of the spine and femora were consistent with myelosclerosis or possibly leukaemia. They suggested that the differential diagnosis of the compressing lesion lay between a leukaemic deposit, metastasis, or perhaps extra-osseous haematopoietic tissue (Fig. 2).

OPERATION Myelography precipitated acute retention of urine. The patient was transferred to the Newcastle General Hospital, where a laminectomy centred on the twelfth dorsal vertebra was performed (L.P.L.). The laminae were very vascular; the spinal cord was not pulsating and was pushed posteriorly by an extradural mass of highly vascular tissue lying anteriorly to the cord and extending round on both sides, particularly the left. The situation of the tumour made its exact origin difficult to determine but as much of it as possible was removed.

HISTOLOGY Examination by Dr. B. E. Tomlinson showed that the material removed consisted of vascular fibrous

FIG. 2. Complete block to the Myodil column, the nature of the obstruction suggesting an extradural lesion. Coarse vertical striation of vertebral bodies above the level of the block.

tissue and a nodule of material identical with the bone specimen from the lamina. The section of lamina showed that highly cellular marrow extended through the bone rim and it seemed likely that the extradural mass was coming from the vertebra. The entire marrow was ex-

FIG. 3. Section of tumour showing megakaryocyte hyperplasia and some bone. Haematoxylin and eosin $\times 160$.

tremely cellular and replaced bony trabeculae. It consisted largely of myelopoietic tissue with numerous eosinophils and eosinophil myelocytes as well as large bizarre giant cells which were almost certainly megakaryocytes. Some areas of normoblastic proliferation were present. There was no evidence of secondary carcinoma and a few clumps of undifferentiated cells in the marrow were interpreted as blast cells. The appearances were therefore of intense marrow hyperplasia, chiefly involving white cells and causing bone destruction. The most likely diagnosis was considered to be myelogenous leukaemia (Fig. 3).

SUBSEQUENT PROGRESS

The patient was discharged to the Cumberland Infirmary, Carlisle, after a course of radiotherapy (1,000 rads.) directed at D5-D12 vertebrae. When examined after four months of regular post-operative physiotherapy she could walk a mile unaided, though her legs were a little stiff, and she could do all her own housework. There were no urinary or sensory symptoms. The cranial nerves and upper limbs were normal. There was slight hypertonus of the lower limbs; power was excellent with only slight weakness of inversion of the feet and dorsiflexion of the right foot. There was minimal depression of pain and touch sensation below the second lumbar dermatome bilaterally but the second and third lumbar dermatomes on the left were anaesthetic. Vibration sense was diminished

below mid-calf on the right but positional sense was unimpaired. The plantar responses were extensor; the abdominal reflexes and the left knee jerk were absent and the other deep reflexes were exaggerated. At this time the haemoglobin was 15.1 g./100 ml. and white cell count 12,000/c.mm. with a normal differential count and a platelet count of 230,000/c.mm.

DISCUSSION

Two points of interest arise from this case: which is the correct haematological diagnosis and what is the anatomical origin of the tumour? Dameshek (1951) and Hutt, Pinniger, and Wetherley-Mein (1953) have claimed that polycythaemia, myelosclerosis, and chronic myeloid leukaemia are all parts of a spectrum of myeloproliferative disorders, but this is disputed by Leonard, Israëls, and Wilkinson (1957). The histology of the tumour in our patient indicated myelogenous leukaemia. The splenomegaly would suggest chronic myeloid leukaemia or myelosclerosis but the blood picture and polycythaemia, bone marrow, splenic puncture, and the positive leucocyte alkaline phosphatase (Mitus, Bergna, Mednicoff, and Dameshek, 1958) favour a diagnosis of myelosclerosis. Taking the haematology, radiology, and progress of the patient into account, we feel that all the findings, including the histology, are explicable as myelosclerosis with extramedullary haematopoiesis.

The tumour was at the level of the vertebral changes which were typical of myelosclerosis (Moseley, 1961, 1963) and were not present in the case of Close *et al.* (1958). There was no evidence of vertebral fracture allowing marrow to extrude nor any evidence of paravertebral haematopoietic tissue (Knoblich, 1960) which might have grown through the intervertebral foramina. Whether the bone marrow burst out of the vertebra or the tissue originated in the extradural space (Close *et al.*, 1958) is not certain. Nests of lymphocytes are present in the extradural space (Bhagwati and McKissock, 1961) and it is probable that pluripotential reticulo-endothelial cells lie in this space and may occasionally proliferate to form masses of differing types, including haematopoietic tissue.

SUMMARY

A case of myelosclerosis with spinal cord compression due to extramedullary haematopoiesis is reported and this very rare occurrence is discussed. We wish to thank Dr. T. McL. Galloway and Dr. Henry Miller for permission to publish this case, Dr. B. E. Tomlinson for the histological report, and Dr. W. Walker for the haematological studies.

ADDENDUM

One year after operation the patient remains symptom free, with haemoglobin 13.3 g./100 ml. and white blood count 10,700/c.mm., with a normal differential count.

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