INTRATHORACIC MENINGOCELE, SPINAL DEFORMITY, AND MULTIPLE NEUROFIBROMATOSIS

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Multiple neurofibromatosis, originally described by von Recklinghausen (1882) as a disease of the cutaneous nerves, is now recognised to have many pathological manifestations. Its presenting symptoms are protean and often bizarre. In the past twenty years an enormous literature has accumulated which deals with various aspects of the disease, new manifestations and recently discovered associated pathological changes. Much of this information is relevant to orthopaedic surgery in that it concerns the skeletal changes seen in patients with multiple neurofibromatosis.

In an extensive review Preiser and Davenport (1918) stated that von Recklinghausen's disease occurs in about one in two hundred of the general population, and affects both sexes equally in all races and at all ages. Approximately one case in ten shows skeletal changes, and in one half of these the spine is involved. There is a remarkable association of this involvement of the spine with the very uncommon condition of lateral intrathoracic meningocele. In a review of the literature to the end of 1949, reports of only five such cases were found. Two further cases are reported here.

CASE REPORTS

Case 1—A woman aged twenty-three years, first seen in 1948 at St Mary's Hospital, London. She complained of steadily increasing pain between the shoulder-blades during eight years. Eighteen months previously she had been examined radiographically at another hospital, and told that she was suffering from an old crush fracture of a dorsal vertebra. Twelve months' physiotherapy gave no relief. The pain was localised to a small area between the shoulder blades and occasionally radiated round the left side of her chest. It was aggravated by forced movements of the spine, particularly on twisting to the left. She had also noticed that she was becoming round shouldered, but her general health had remained excellent. On clinical examination several large and irregular café-au-lait patches were seen on her trunk and buttocks. She had a slight upper dorsal kyphosis but no obvious scoliosis. There was no limitation of spinal movement. Examination of the central nervous system revealed no abnormality apart from defective posterior column conduction as shown by loss of vibration sensibility in the legs and loss of postural sensibility in both great toes. No other physical abnormality was detected clinically. Radiographic examination of the spine (Figs. 1 and 2) showed disappearance of the pedicles of the fifth, sixth and seventh thoracic vertebrae on the left side, and spreading of the vertebral ends of the left sixth, seventh and eighth ribs, with erosion of their related margins. There was well-marked scalloping of the posterior aspect of the fifth, sixth, seventh and eighth thoracic vertebral bodies, but the intervening disc spaces were relatively unaffected. There was wedging of the affected vertebrae, giving rise to a rounded kypho-scoliosis. A radiograph of the chest (Fig. 3) showed a soft-tissue tumour in relation to the 5–6 intercostal space. Lumbar puncture—The cerebrospinal fluid pressure was 120 millimetres; no block was demonstrated. Chemical analysis of the fluid was normal; protein was 20 milligrams per cent.

In spite of the normal cerebrospinal fluid protein content, a diagnosis of "dumb-bell" neurofibroma was made with some confidence on the basis of the radiographic appearances and the pigmented skin lesions typical of von Recklinghausen's disease.

Operation (Mr A. Dickson Wright)—After costo-transversectomy (left fifth and sixth ribs) a soft-tissue tumour was immediately displayed (Fig. 4). It lay outside the pleura and emerged through a defect in the region of the pedicles of the sixth and seventh thoracic vertebrae. The tumour was
Case 1—Radiographs of the thoracic spine showing, in the antero-posterior view (Fig. 1), marked spreading of the sixth, seventh and eighth ribs, the margins of which are eroded. Note in the lateral view (Fig. 2) the disappearance of the pedicles of the fifth, sixth and seventh thoracic vertebrae, the scalloping of the posterior margins of four vertebral bodies (but not the intervening disc spaces), and the anterior wedging of the affected vertebrae.

Case 1—Chest showing soft tissue opacity in relation to the fifth-sixth intercostal space.
gently pulsatile and collapsible on pressure; its wall resembled dura mater in colour and texture. After incision of the sac, the spinal cord was seen lying in its normal position in no way embarrassed or distorted by the dural herniation. The meningocele was readily excised and the resultant dural opening closed by silk sutures. The patient made an uninterrupted recovery from the operation and was discharged wearing a plaster jacket. Eighteen months later she was free from pain. She is to be readmitted for operative fusion of the mechanically unsound segment of her dorsal spine.

**Case 2**—An alert little girl, first attending the Royal National Orthopaedic Hospital in 1949 at the age of eight and a half years, because of her inability to walk and an increasing curvature of the spine. At three years old she had attended a clinic where she was noted as having a mid-dorsal scoliosis and multiple neurofibromatosis. Her walking was said to be normal at that time, and she had been given a spinal support. Four years later the scoliosis became worse after a fall, and her left leg became spastic. The spasticity improved after head traction in a plaster bed. She continued to wear a spinal support on her discharge from hospital but in the months before the latest examination both legs became so spastic that she was unable to get about.

**Examination**—The skin of the trunk and thighs was covered with lesions typical of neurofibromatosis. There was an acutely angulated kypho-scoliosis convex to the right, the apex at the sixth thoracic vertebra, with compensatory curves above and below (Fig.5). Examination of the central nervous system showed a severe spastic paraplegia and impairment of cutaneous sensibility to light touch and pin-prick below the dermatomes corresponding to the level of T.12. Radiographs of the spine (Figs. 6 and 7) showed gross lateral displacement of the seventh and eighth thoracic vertebrae, which formed the apex of an acute kypho-scoliosis. The 7-8 intercostal space was increased in width and the related margins of the ribs were eroded. The posterior surfaces of the vertebral bodies at the level of the deformity showed well-marked scalloping. The appearances suggested localised bone resorption from the pressure of a tumour mass. **Lumbar puncture**—Cerebrospinal fluid pressure, 275 millimetres. Free rise and fall on jugular compression. Cerebrospinal fluid chemical analysis, normal; protein, 20 milligrams per cent. **Myelography** showed an obstruction at the level of the eleventh thoracic vertebra, but the examination was technically
Case 2—Photograph of the child. Note the gross kyphosis, the areas of pigmentation, and the soft tissue swelling over the left lower ribs.

Case 2—Thoracic spine showing severe kypho-scoliosis. Note in the anteroposterior view the gross lateral displacement of the seventh and eighth thoracic vertebrae, and the increased width of the seventh-eighth intercostal space.

Case 2—The lateral view shows well marked scalloping of the posterior margins of four vertebral bodies.
unsatisfactory and no other useful information was obtained. (Post-operative myelography demonstrated the meningocele more clearly and showed that the dilatation of the subarachnoid space extended over several segments corresponding to the erosion of the vertebral bodies.) Operation was advised for relief of the paraplegia which was presumed to be due to an incomplete obstruction caused by the distorted vertebral column. 

Operation (Mr H. J. Seddon)—Antero-lateral exposure of the spinal cord in the concavity of the scoliosis showed it to be stretched over a sharp bony mass. The cord was feebly pulsatile across the site of obstruction and this pulsation improved when the offending ledge of bone was removed. Exploration at the convexity of the scoliosis revealed a sessile intrathoracic and extra-pleural meningocele, approximately 3 centimetres long by 2-5 centimetres broad, which was readily exposed by removing the medial ends of the three related ribs (Fig. 8). No attempt was made to remove the sac because the spinal cord was apparently in no way embarrassed by it. A mass of tissue at the site of an intercostal nerve was removed for examination and proved to be a plexiform neroma. Post-operative progress was uneventful. There was an initial improvement of the paraplegia as manifested by reduction in the spasticity of the legs, but this early promise was not fulfilled and she remained with severe paralysis three months later. 

Comment—This case is of interest not only as an example of lateral intrathoracic meningocele, it also demonstrates the compression paraplegia which not uncommonly complicates gross kyphoscoliosis in von Recklinghausen's disease.

**DISCUSSION**

Reports of eight cases of lateral intrathoracic meningocele have been found in the literature. Two more are reported here, making a total of ten in all, of which at least seven occurred in association with multiple neurofibromatosis. Such an association is unlikely to be fortuitous and may help to shed some light on the more common occurrence of spinal deformity in this disease. The deformity has been variously attributed to the action of periosteal neurofibromata (Brooks 1924) or an ill-defined osteomalacia (Gould 1918), but the evidence is inconclusive. Cross (1949) was of the opinion that the meningocele is due to a diseased dura and not primarily to a defect in the bone. The drawing of Ameuille's (1940) specimen, which is reproduced in Figure 9, lends support to this view. The gross dilatation and sacculation of the spinal dura in his case of lateral meningocele is very striking.
My attention has been drawn by a personal communication (Bull 1950) to a case of a woman, aged thirty-seven years, who exhibited the cutaneous lesions of neurofibromatosis and who died of a cerebral tumour (spongioblastoma multiforme). The radiographs of her lumbar spine (Figs. 10 and 11) showed the same scalloping of the posterior surfaces of several vertebral bodies that is seen in cases of lateral meningocele; and the transverse processes were eroded. Post-mortem examination of the spine showed that the vertebral bodies from the twelfth thoracic to the third lumbar (inclusive) were grossly deformed and apparently hollowed out from behind by the distended theca which bulged through the large intervertebral foraminae.

One is tempted to put forward the hypothesis that in von Recklinghausen's disease the spinal deformity may be due to the prolonged aneurysmal action of an undisclosed meningocele or of an intrathecal dilatation of the dura mater. Further study is needed to determine how commonly the spinal dura is affected in cases of multiple neurofibromatosis with deformity of the spine.

At present we can go no further than to say that a lateral intrathoracic extrapleural meningocele associated with spinal deformity in von Recklinghausen's disease constitutes a definite clinical entity whose specific features have been evident in the case reports so
regularly that we might now reasonably expect to be able to make a correct pre-operative diagnosis.

SUMMARY

Two cases of lateral meningocele and spinal deformity in von Recklinghausen's disease are reported. Of the ten known cases of lateral meningocele, seven have occurred in patients with neurofibromatosis. The relation of spinal deformity and neurofibromatosis is briefly discussed.

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REFERENCES


BULL, JAMES (1950): Personal communication.


