CONGENITAL SPINAL EXTRADURAL CYST (LATERAL MENINGOCELE) SIMULATING ACUTE TRANSVERSE MYELITIS

REPORT OF A CASE

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Congenital spinal extradural cysts are rare and may be the cause of acute paraplegia. In their clinical features they closely resemble acute transverse myelitis. Immediate decompression of the spinal cord and removal of the cyst may lead to restoration of normal function. Myelography differentiates these two conditions by showing a cyst in communication with the spinal canal. This investigation must be mandatory.

Congenital spinal extradural cysts are herniations of the arachnoid mater through an aperture in the dura mater, producing a cyst containing cerebrospinal fluid in communication with the spinal canal. More commonly completely extradural, the cyst may however have a significant portion which is an intradural extra-arachnoid extension. The syndrome of congenital spinal extradural cyst was first described by Elsberg, Dyke and Brewer (1934). Adolescents are usually affected and the condition is more common in males. The patient presents with progressive paraplegia, the motor component being more obvious than its sensory counterpart. In a review of sixty-one cases (Gortvai 1963), these cysts were found most commonly in the lower thoracic spine, less commonly in the upper thoracic and lumbar regions. The characteristic lesions of the axial skeleton in these individuals include a kyphos, widening of the interpedicular distance, erosion of a pedicle and excavation of the contiguous margins and posterior surfaces of the involved vertebrae.

The so-called lateral meningocele (Shapiro 1968) is a lesion of similar features and origin but usually presents as an asymptomatic posterior mediastinal mass. It only rarely occurs in the lumbar region but has been reported in association with neurofibromatosis (Sammons and Thomas 1959). It frequently coexists with scoliosis, projecting to the side of the convexity of the curve. Both the lateral meningocele and the extradural cyst are characterised by myelographic evidence of flow of contrast medium from the spinal canal into the cyst. We report a patient presenting with acute paraplegia similar to that of transverse myelitis, subsequently shown to be due to a congenital spinal extradural cyst.

CASE REPORT

In October 1964 a girl aged twelve was found sitting on the kitchen floor by her mother who had been out shopping for the previous three hours. She said that her legs felt limp and that she could not move them. She had vomited once. There was no history of injury and she was in good previous general health with no relevant past medical history. She was admitted to hospital immediately and found to have complete paralysis of the lower limbs with a sensory level fluctuating.

Fig. 1

Supine anteroposterior radiograph of the lower thoracic and lumbar spine in 1971. There is a structural thoracolumbar scoliosis and congenital failures of fusion of the posterior elements (arrowed).

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THE JOURNAL OF BONE AND JOINT SURGERY
between the eighth and tenth thoracic levels. She had urinary retention but there were no other relevant physical findings. Radiographs of the spine were reported as showing only slight narrowing of the disc space between the seventh and eighth thoracic vertebrae. All haematological and biochemical investigations were normal. The cerebrospinal fluid was clear and colourless, with a pressure of 260 millimetres of water, a free rise and fall being observed. There was no abnormality of cellular content, protein or sugar. A provisional diagnosis of transverse myelitis was made. The paraplegia remained complete but she was rehabilitated and learned to walk with calipers and crutches, expressing her bladder three-hourly. Ten months later she was discharged home. In October 1971, seven years after the onset, a cholecystogram showed a non-functioning gall bladder, and an anteroposterior radiograph of the lower thoracic and lumbar spines showed a structural scoliosis extending from the ninth thoracic to the third lumbar vertebra, with widening of the interpedicular distances at the L2–3 level and widening of the intervertebral space (Fig. 1). There were extensive posterior defects throughout the lumbar spine, the most obvious being a defect in the pars interarticularis and a spina bifida occulta at the L2 level, and a fusion of the posterior elements of the fourth and fifth lumbar vertebrae.

She was admitted to the Nuffield Orthopaedic Centre in August 1976 with a four-week history of pain on movement of the upper lumbar spine, associated with occipital headaches. There were no other symptoms and she was in good general health. On examination the right thoracolumbar scoliosis was noted and pain could be reproduced on extreme flexion of the lumbar spine. The neurological state had remained static with a complete loss of motor and sensory function below the T8 level. Anteroposterior and lateral radiographs of the lumbar spine (Figs. 2 and 3) showed that extensive excavation of the contiguous margins of the second and third lumbar vertebral bodies had occurred, with a large area of new bone posterolaterally. A lateral radiograph taken with the patient sitting (Fig. 4), clearly showed the instability at this level with the presence of a kyphos and the spine collapsing in flexion. Haematological and biochemical investigations were normal except for a raised erythrocyte sedimentation rate of 43 millimetres in the first hour. No antibodies were detected to Staphylococcus aureus or Brucella. In view of the possibility of infection at this level a needle was introduced into the L2–3 interspace and 20 millilitres of clear fluid obtained. A small quantity of radio-opaque dye was then injected, and a subsequent radiograph of the spine revealed that the dye had filled a cyst (Fig. 5). A further radiograph taken two days later, as a preliminary film before excretion urography (Fig. 6), showed that the dye injected had now filled a large cyst, the greater part of which was lateral to the convexity of the scoliosis (Fig. 7). Furthermore, the dye had entered the spinal canal. Myelography was therefore performed: the anteroposterior view showed that the contrast material injected into the spinal canal had begun to refill the cyst and, in addition, that the cyst had an intradural extension as far proximal as the tenth thoracic vertebral body (Fig. 8); the lateral myelogram showed a complete block (Fig. 9). The fluid aspirated from the cyst grew neither pyogenic nor acid-fast organisms.

The diagnosis of congenital spinal extradural cyst (lateral meningocele) was made. The local pain was considered to be readily explicable on the basis of an unstable kyphos at the L2–3 level due to loss of anterior bone substance and posterior element insufficiency. Posterior spinal fusion with Harrington instrumentation was performed and the large quantity of posterolateral bone was used as an autogenous graft. The cyst was clearly seen and its neck ligated, but it was not resected. Now, two years after the operation, she has a solid straight lumbar spine with no instability and no pain.

**DISCUSSION**

Acute transverse myelitis is one of the forms of acute disseminated encephalomyelitis affecting the spinal cord only. It is a disease of childhood, adolescence and early adult life, and the mid-thoracic region is usually affected. It may be initiated by vaccination or non-specific respiratory infection. It is considered to be a non-specific hypersensitivity phenomenon and can be produced in experimental animals by injecting sterile extracts of nervous tissue, when it is known as experimental allergic encephalomyelitis (Matthews and Miller 1972). The condition is characterised by an acute, often catastrophic, onset that may frequently show remarkable recovery over a period of weeks with ultimate minimal disablement. A more unfavourable prognosis exists when there is a less acute onset and the
cord is damaged below the lesion; this is termed subacute necrotic myelitis. There are frequently no changes in the cerebrospinal fluid other than a raised gammaglobulin content. Acute transverse myelitis therefore has many features in common with the syndrome of congenital spinal extradural cyst (Elsberg et al. 1934). Both conditions tend to affect the younger age group, an acute onset is common, complete paralysis may be produced, and there are frequently no changes in the cerebrospinal fluid.

The characteristic radiographic appearances of the axial skeleton in patients with congenital spinal extradural cysts are important in differentiating this condition from acute transverse myelitis. The widening of the interpedicular distance, excavation of the contiguous surfaces of the vertebral bodies involved, erosion of the pedicles, and the production of a kyphos are typical. The lateral meningocele is a similar herniation of the arachnoid mater through an aperture in the dura mater and is usually associated with neurofibromatosis and scoliosis (Sammons and Thomas 1959). These congenital cysts are produced by a persistence of part of the neureneric canal which connects the ectodermal neural plate to the anterior endoderm from which the alimentary system develops. This canal normally atrophies and disappears. If the neureneric canal remains widely patent, complete rachischisis occurs, whereas persistence of parts of the canal result in such lesions as diastematomyelia, spina bifida occulta, or adhesions between the spinal cord and the intestinal tract (Bremer 1944; Neuhauser, Harris and Berrett 1958). These adhesions may produce glial or alimentary diverticulae resulting in anterior meningoceles, spinal enteric cysts or congenital spinal extradural cysts. Myelography may demonstrate partial or complete obstruction in the spinal canal, but if there is a free flow of dye between the spinal canal and the cyst the diagnosis is incontrovertibly established (Shapiro 1968).

In our case the neck of the cyst was tied at operation but the meningocele sac was not excised and, the patient having been completely paraplegic for a period of fourteen years, no attempt was made to decompress the spinal canal.
REFERENCES