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Peridural lipomatosis

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Fifteen cases of peridural lipomatosis are presented. All patients were treated surgically in the years 1965 to 1974 and reexamined in 1975. In 5 cases the condition improved, in 4 cases it was unchanged and in 6 patients it deteriorated. It is emphasized that the question of surgical therapy must be decided individually.

Since the description of its typical neurological and radiological symptoms [5, 10] peridural lipomatosis has been an entity well-known to neurologists and neurosurgeons. The clinical picture comprises gait disturbances, paresis of the lower limbs, bladder dysfunction and other neurological and orthopaedic signs. The diagnosis is based upon electromyographic and X-ray studies including air myelography. By the latter method the peridural space is determined indirectly on the basis of the distance between the subarachnoidal space and the spine. The distance is normally 3—4 mm (Fig. 1). If the widening of the peridural space and narrowing of the subarachnoidal space is associated with radiological or other signs of pressure and progressive neurological symptoms, surgery is indicated.

In the years 1965 to 1974, we observed 15 children. Their age ranged from 2 to 7 years. Their symptoms are shown in Table I; characteristic were the gait disturbances, spastic paresis and hypotonic paresis of the lower limbs, long tract signs, foot deformities, sensory disturbances and primary bowel or bladder disturbances. EMG and X-ray studies including air myelography were carried out in all patients. Particularly in spastic cases the EMG

Table I

Presenting signs and symptoms in 15 patients with peridural lipomatosis

Main symptoms	Number
Gait disturbance	8
Spastic paresis of lower limbs	4
Atonic or hypotonic paresis of lower limbs	5
Long tract signs	5
Foot deformity	4
Growth asymmetry of leg	3
Primary bowel or bladder dysfunction	2
Sensory disturbance	1

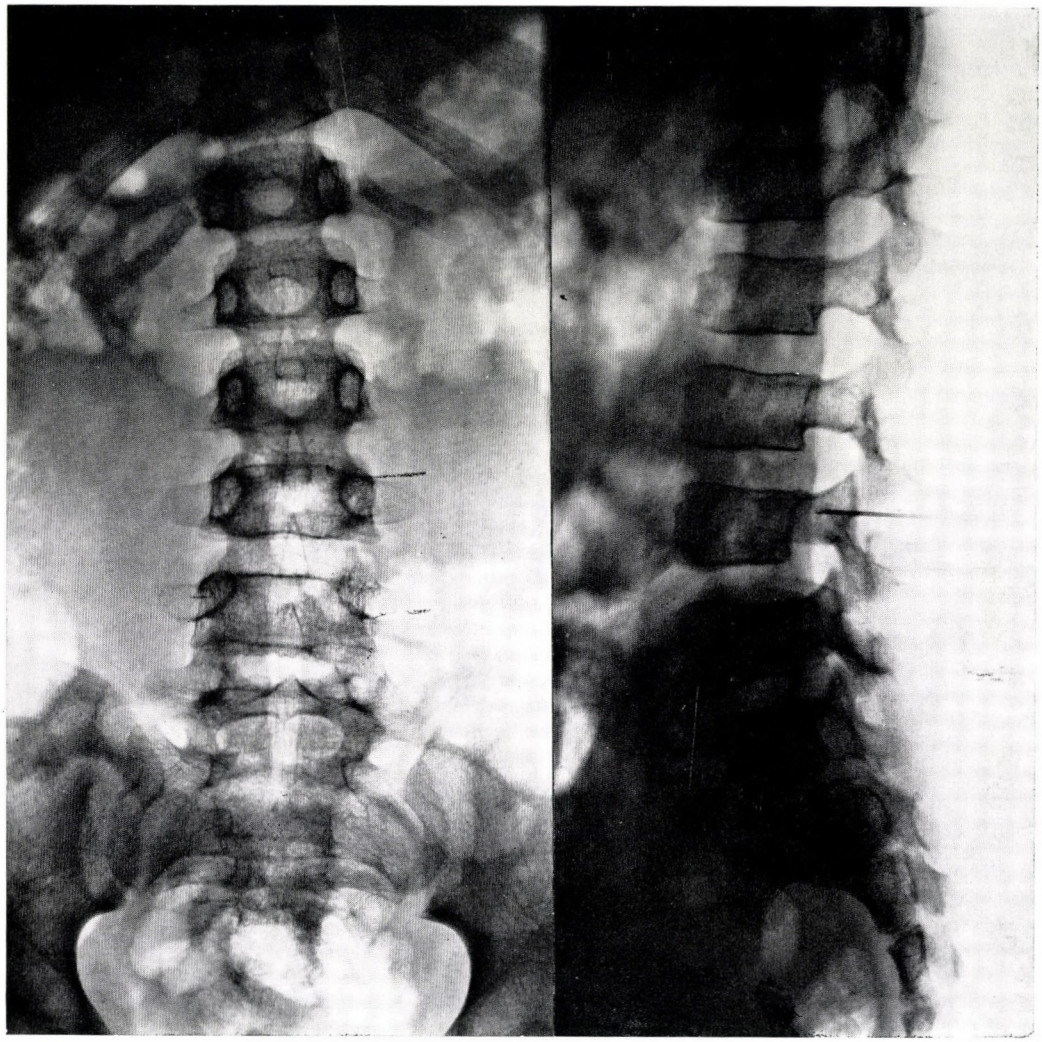
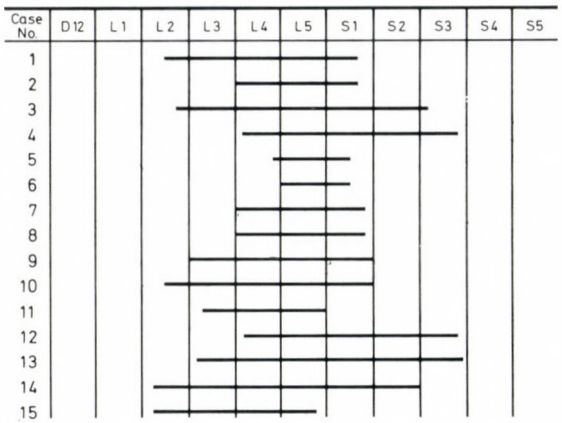


FIG. 8-year-old boy with peridural lipomatosis L4/L5. The distance between spine and subarachnoidal space is 8 mm.

revealed a proximal damage of the peripheral neurone in addition to supraspinal disturbances. Typical X-ray signs were observed in every case, including a narrowing of the spinal canal, excavation of the dorsal part of the spine, widening of the interpedicular space, deformation of the pedicle, anomalies of the spines and laminar arches, etc. These signs were often combined with dysraphic malformations: spina bifida in 7 cases, hydrospina in 4 cases, scoliosis connected with postural anomalies in different regions of the vertebral column in 8 cases, etc.

TABLE II

Level of peridural lipomatosis in the lumbosacral region in 15 cases



The localization of peridural lipomatosis in these patients is shown in Table II; it is seen that the favoured site was the lumbosacral region.

Treatment was surgical in all the 15 cases. The operation consisted in decompressive laminectomy with biopsy and total or subtotal removal of fatty tissue, particularly in the dorsal and lateral parts of the peridural space. In the ventral part the surgical procedure was often limited, because the nerve roots were enlarged and connected with the fatty tissue.

The characteristic histological finding was mature fatty tissue partly mixed with fibrous elements. Signs of malignancy were absent.

When examining the patients in 1975, the results were as follows.

In 5 cases an improvement was found with complete or incomplete regression, particularly of the gait disturbance.

In 4 cases the neurological condition persisted in an unchanged form, showing no improvement.

TABLE III

Peridural lipomatosis associated with neuromuscular systemic diseases

Disease	Number
Spinal muscle atrophy	
Werdnig-Hoffmann type	2
Kugelberg-Welander type	1
Congenital muscle dystrophy	1
Neurogenic muscle atrophy, unclassified	1
Neurofibromatosis Recklinghausen	1

In 6 patients a deterioration of the neurological symptoms occurred. In these cases the condition was connected with systemic diseases, as shown in Table III. Muscle atrophy in the upper limbs developed after the operation in 2 cases.

Comment

In accordance with a number of authors [1, 4, 6, 7, 9, 11, 12] it may be said that the exact pathogenesis of spinal lipomas and of peridural lipomatosis is unclear. The high incidence of associated congenital anomalies, the age at onset and the preferentially lumbosacral location suggest a developmental origin from embryonic mesenchymal cells or residues of embryonic ectodermal cells involved in the formation of the spinal cord [2, 3]. The cause of the dysraphic syndromes in the lower back could be a metabolic error before the fifth somite stage [3].

The clinical features in our 15 cases were not specific. The same symptoms may occur in various other diseases, for instance different forms of restriction of cranial migration of the spinal cord [9], tumours in the lumbosacral region [1, 8, 11, 12], systemic diseases beginning with neurological symptoms in the legs [13], etc. In most cases the combination of clinical neurological and radiological findings will allow to decide for a surgical approach. It is nevertheless important that the widening of the peridural space revealed by air myelography is not always a sign of mechanical pressure, as shown by those of our cases in which the lipomatosis was associated with some systemic disease.

In accordance with Matthias and Lausberg [9] it should be emphasized that the question whether or not an operation is indicated in childhood, must be considered individually in each case.

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