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**Compressive meningeal hypertrophy in
mucopolysaccharidosis.**

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Compressive Meningeal Hypertrophy in Mucopolysaccharidosis

Compression of the spinal cord in patients with mucopolysaccharidosis is often caused by an osseous deformity at the thoracolumbar or the craniocervical region. The purpose of this paper is to draw attention to hypertrophy of the meninges as a possible cause of cervical cord compression in these patients.

Case Report

A 19-year-old girl suffering from mucopolysaccharidosis type VI was under periodic observation at McMaster University Medical Center. Her neurologic problems began at age 16 with back pain that was ascribed to an existing deformity of the lumbar vertebrae (Fig. 1). Her symptoms improved after physiotherapy.

Fig. 1.—Lateral radiograph of lumbar spine illustrating abnormal ossification of ring epiphyses, hypoplasia of first lumbar vertebra, and kyphosis at dorsolumbar junction.



Six months later, the patient complained of pains and parasthesiae in both hands. A diagnosis of bilateral carpal tunnel syndrome was made and she improved significantly after surgical decompression. However, she returned a few months later with severe exacerbation of her symptoms and on examination she had increased reflexes in all four extremities and bilateral upgoing toes.

CT myelogram showed uniform extradural compression around the spinal cord from C1 to C3 (Fig. 2). At surgery, this was seen to be caused by thickening of the dura mater. The dura was very thick, but of abnormal consistency; it could not hold the sutures well. After decompression of the spinal cord, the patient gradually improved.

Discussion

Several neurologic complications result from mucopolysaccharidosis (MPS).

1. Mental retardation: this is most severe in Hurler syndrome (MPS IH) and Sanfilippo syndrome (MPS III); it is mild in Hunter syndrome (MPS II), and it is absent in Scheie syndrome (MPS IS), Morquio's syndrome (MPS IV), and Maroteaux-Lamy syndrome (MPS VI).

2. Hydrocephalus: communicating hydrocephalus may develop in the severe forms of mucopolysaccharidosis that are associated with mental retardation. The development of hydrocephalus is due to thickening of the leptomeninges, which may interfere with the absorption of CSF at the arachnoid granulations or may cause partial obstruction of the CSF at the exit foramina of the fourth ventricle.

3. Compression of the spinal cord at the craniocervical junction: this is caused by atlantoaxial subluxation secondary to hypoplasia of the odontoid process. This complication is more frequent in children with Morquio's syndrome.

4. Spinal cord compression at the dorsolumbar area: this is caused by the gibbus deformity.

5. Spinal cord compression in the cervical region: a complication that is caused by thickening of the leptomeninges. This is an exceedingly rare cause, but a few cases have been reported in association with Maroteaux-Lamy syndrome [1-4].

Apart from mucopolysaccharidosis, other conditions that

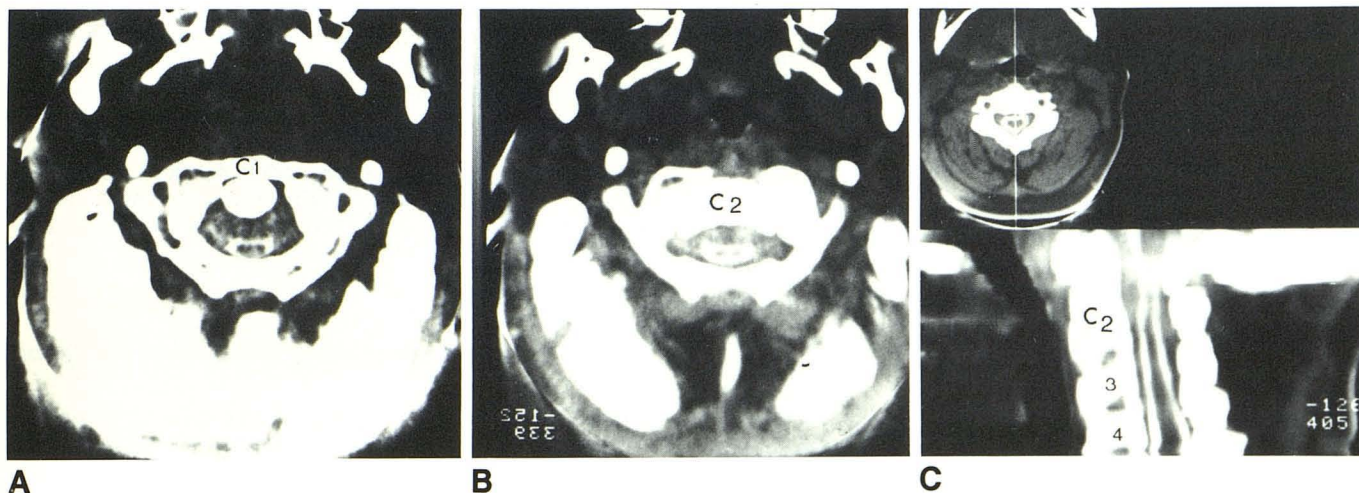


Fig. 2.—CT myelogram of cervical region illustrating severe atrophy of spinal cord, which is maximal at level of atlas and axis, and a wide gap between opacified arachnoid sac and spinal canal, caused by thickening of dura and epidural tissues.

might cause thickening of the leptomeninges to the extent of causing cord compression are: adhesive arachnoiditis from an infectious or inflammatory condition, syphilitic pachymeningitis hypertrophica, and pachymeningeal carcinomatosis [5]. Myelography in all these conditions will demonstrate concentric narrowing of the subarachnoid space with increased distance between the contrast column and the spinal bony canal [6].

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