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Cervical Myelopathy Secondary to Hunter Syndrome in an Adult

Matthieu Vinchon, Anne Cotten, Jacques Clarisse, Rozenn Chiki, and Jean-Louis Christiaens

Summary: We present a case of type II mucopolysaccharidosis in which the diagnosis was delayed until the onset of cervical myelopathy in adulthood. Radiographic features were characteristic, with striking dural thickening shown on CT and MR imaging.

Index terms: Spinal cord, myelopathy; Metabolic disorders

Mucopolysaccharidoses can cause cervical myelopathy because of spinal stenosis (1). This is usually diagnosed in children, and paraparesis is an unusual clinical presentation (2). We present a case of mucopolysaccharidosis in an adult in whom the presenting feature was cervical myelopathy.

Case Report

A 44-year-old man was referred by his family physician, who had treated him for slowly progressive spastic tetraparesis for 6 years. He had undergone L4-5 discectomy 5 years before and corrective surgery for left-sided genu valgum 1 year before. Six months before, he had consulted a neurologist because of a rapidly worsening neurologic condition.

The patient had a large head and coarse facies. His hands and feet were short, broad, and webbed. He was heavily built, measuring 173 cm and weighing 120 kg, and had a prominent belly. Neurologic examination showed marked weakness in all extremities, and he was unable to stand. He had increased extensor tone and impaired sensation to vibration in the legs, particularly on the left side. Electrophysiologic examination revealed marked slowing of central motor conduction, most marked in the right arm.

Radiographs of the cervical spine showed developmental abnormalities (Fig 1A). On computed tomography, the spinal canal was narrowed and the spinal cord compressed (Fig 1B). Magnetic resonance (MR) imaging of the cervical spine showed marked compression of the spinal cord and thickened hypointense dura on T1- and T2-weighted sequences (Fig 1C and D).

The diagnosis of Hunter syndrome was confirmed by biopsy of both a peripheral nerve and muscle, on which

ultrastructural studies were performed and measurement of urinary glycosaminoglycans and sulfiduronate sulfatase activity in cultured skin fibroblasts.

The patient underwent a laminectomy at C-2 to C-6 with concomitant monitoring of somatosensory evoked potentials. At surgery, the laminae were thickened with enlarged medullary cavities. The dura mater appeared normal. After the operation, there was no clinical improvement, and the patient was confined to a wheelchair.

Discussion

Hunter syndrome is a rare cause of cervical myelopathy, most commonly diagnosed in infancy and childhood (2). It is a hereditary, X-linked recessive disease. The clinical manifestations result from the accumulation of dermatan sulfate and heparan sulfate in the tissues, related to a deficiency in sulfiduronidase sulfatase. Neurologic symptoms generally appear late in the course of the disease (3). They might be caused by deposition of glucosaminoglycans in the brain, abnormalities of myelination, hydrocephalus, stenosis of the foramen magnum, cervical stenosis, severe spinal deformities, or peripheral nerve entrapment (4). Hunter syndrome carries a poor prognosis because of the associated neurologic problems, dementia, and visceral involvement. A milder form of Hunter syndrome can be diagnosed in adolescence or adulthood. Life expectancy is decreased.

Radiographs in mucopolysaccharidosis show characteristic dysplastic features with blunted spinous processes, wedge-shaped vertebral bodies, and stenosis of the spinal canal. MR imaging is the technique of choice in the evaluation of brain and spinal involvement in mucopolysaccharidosis (5). Although dural thickening can be seen on computed tomography and computed myelography (6), it is particu-

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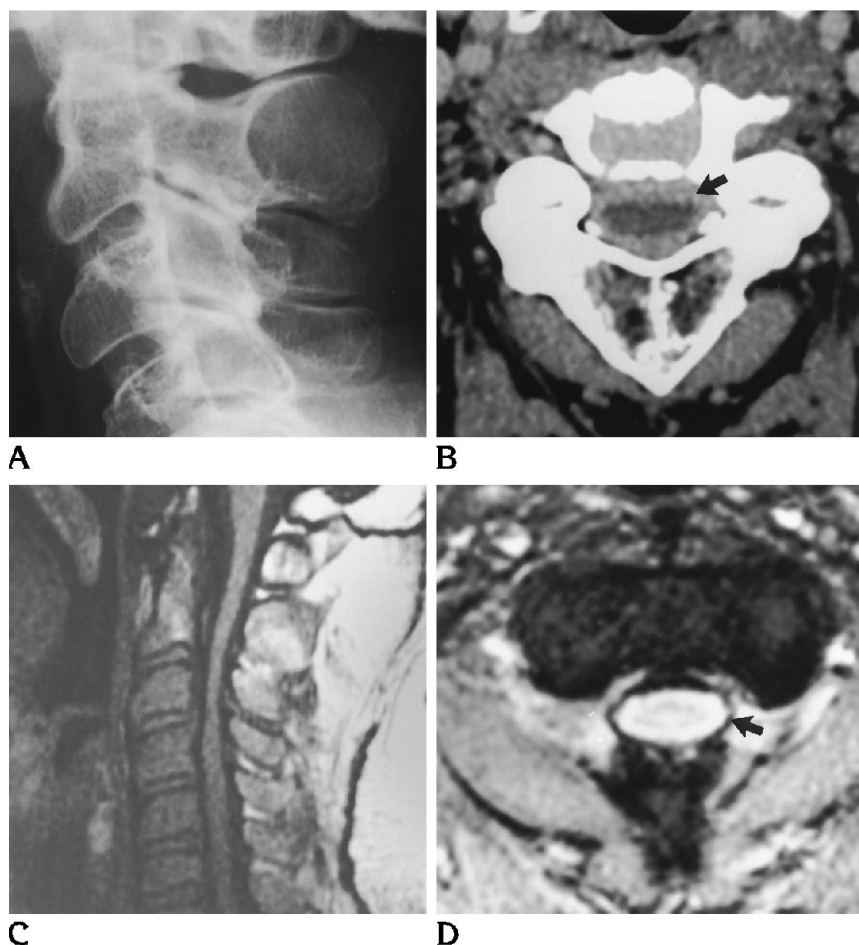


Fig 1. A, Radiograph of the cervical spine, lateral view. Gross dysmorphic features are conspicuous. There is defective development of the anterosuperior part of the vertebral bodies. Spinous processes are large and rounded. The spinal canal is narrow.

B, Axial unenhanced computed tomogram of the cervical spine. The laminae, not clearly demarcated from the spinous process, are markedly thickened. The spinal cord is compressed, surrounded by tissue (arrow). The epidural fat has disappeared.

C, Sagittal T1-weighted MR image (50/20 [repetition time/echo time]). The spinal cord is markedly compressed at the level of the second and third vertebrae.

D, Axial T2-weighted gradient-echo MR image (540/30, 30° flip angle) at the level of C-2. The spinal cord is thin and surrounded by a thickened dura with very low signal (arrow).

larly striking on MR imaging (7). The decreased signal intensity in the thickened dura on T1- and T2-weighted sequences might be related to the deposition of mucopolysaccharides.

Surgery is performed after diagnosis of spinal cord compression. The fate of our patient shows that results are poor when surgery is performed at a late stage of the disease.

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