PARAPLEGIA IN OSTEOPHYSIS IMPERFECTA
A CASE REPORT

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Paraplegia occurred in an adolescent girl with osteogenesis imperfecta after chiropractic manipulation. The child had been able to walk freely out of doors. Complete motor paralysis with sensory sparing resulted due to anterior compression of the cord by spondyloptotic cervical vertebrae. Reconstructed computerised tomography was very helpful in demonstrating the abnormality. Anterior and then posterior decompression relieved the tethered spinal cord and were supplemented with bone grafting. Early diagnosis and surgical treatment will prevent similar neurological accidents.

Osteogenesis imperfecta is a hereditary disease of the connective tissue. Although progressive spinal deformities and spondylolisthesis have been reported (King and Bobechko 1971), a review of the literature revealed no cases of neurological insult due to cervical spondyloptosis.

CASE REPORT

Paraplegia developed in a 12-year-old girl with osteogenesis imperfecta after a chiropractic manipulation. Previously, despite multiple fractures of the limbs, the child had been able to walk freely out of doors and was independent for everyday activities. Her family had noticed gradual sagging of her chin after a minor fall at school. The child was taken to the chiropractor because of headaches, and pain in the neck and low back that interfered with everyday activities. Two weeks after the chiropractic treatment she experienced deterioration of power in the legs accompanied by clonus at rest, urinary urgency and frequency. Motor paraplegia developed and three weeks later she was admitted to The Hospital for Sick Children, Toronto.

The patient’s history included surgical release of knee contracture during infancy. After a tibial fracture in the previous year she needed crutches for walking. There was no family history of osteogenesis imperfecta. She showed signs of anterior compression of the cord. She had complete motor paralysis below C7 with sensory sparing of the sacral dermatomes and proprioception. Signs of upper motor neuron lesions (clonus, hyper-reflexia, and pathological reflexes) were seen. Intrinsic weakness in both hands, particularly the right, gave evidence of a lesion of the first thoracic nerve root.

Reconstructed computerised tomography using sagittal scans showed severe progressive spondyloptosis at the cervicothoracic junction (Fig. 1) that was not obvious on lateral cervical radiographs. Fractures through the elongated pedicles were also visible (Fig. 2). Complete blockage of cerebrospinal fluid was demonstrated by computerised tomography (Fig. 3) and by myelography (Fig. 4). All respiratory, haematological, and urinalysis investigations gave results within normal limits.

Halo traction was applied, followed by anterior decompression of the spinal cord. A T-shaped skin incision permitted an anterior costotransversectomy approach through the bed of the first and second rib to the spondyloptotic vertebrae. The posterior part of the C7 and T1 vertebral bodies was excised to allow later posterior decompression of the cord. Bone grafting was accomplished using the second rib.

Within three days the child began to move her feet and the clonus had disappeared but motor paralysis and clonus both returned when she was immobilised in a halo-vest one week later. Posterior decompression from C2 to T1 was then carried out. The cord had been tethered by the proximal, spondyloptotic vertebrae and was pulsating after decompression. Autogenous bone was supplemented from the bone bank for the posterior fusion.

After this circumferential decompression the patient experienced a progressive increase in power in her hands and legs. Three weeks later she could perform the straight-leg raising test with both legs to 30 centimetres above bed level for 25 seconds and was soon able to walk.

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DISCUSSION

Spinal deformity in osteogenesis imperfecta is due to laxity of spinal ligaments (allowing spinal curvature) and to brittleness of the bone (leading to vertebral compression fractures) (King and Bobechko 1971). This laxity of spinal ligaments is related to abnormal collagen metabolism. A Type I collagen defect has been demonstrated in patients with autosomal recessive osteogenesis imperfecta and in their parents (Penttinen et al. 1975; Nicholls, Pope and Schloon 1979).

The most common spinal problems in osteogenesis imperfecta, however, are compression fractures that lead to loss of vertebral height, kyphoscoliosis, and elongated pedicles that cause lumbosacral spondyloysis (Newman and Stone 1963). Brainstem compression and hydrocephalus have been reported as complications in adolescence (Frank, Berger and Tew 1982).

Patients with severe vertebral slippage have significant symptoms of local and radicular pains due to involvement of the nerve roots. In such cases the nerve roots should be decompressed as well.

We had to use a combined anterior and posterior approach to achieve adequate decompression of the cord and the roots. The application of the halo did not appear to be a problem despite the patient's osteoporotic, soft skull bones.

Prevention of neurological disasters is always important. However, it is even more essential in osteogenesis imperfecta. Prophylactic surgical stabilisation is indicated since spinal bracing is likely to fail (Yong-Hing and MacEwen 1982). Reconstructed computerised tomographic scans were very helpful in demonstrating the anatomical abnormalities.

REFERENCES


