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A LATE NEUROLOGIC COMPLICATION OF SCOLIOSIS SURGERY IN CONNECTION WITH SYRINGOMYELIA

ANDERS NORDWALL & CARSTEN WIKKELSØ

Department of Orthopaedic Surgery, and Department of Neurology, Sahlgren Hospital, Göteborg, Sweden

A case of scoliosis in connection with syringomyelia is described. Theories are proposed to explain the progression of the neurological symptoms after surgical correction and fusion of the deformity. Special points are emphasized that will aid in the recognition of syringomyelia in scoliosis patients.

i) Abnormal neurology, in particular a dissociated disturbance of pain and temperature in the upper extremity.
ii) Abnormal localization of a scoliosis curve.
iii) Rapid progression of the scoliosis.
iv) Bony anomalies of the upper cervical spine.
v) Increased diameter of the cervical spinal canal.

Key words: complications; scoliosis; surgery; syringomyelia

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Scoliosis has long been recognized in connection with syringomyelia, which is always mentioned as one of the neurogenic causes of scoliosis. There is an abundant literature on syringomyelia and its etiology and treatment but very few reports on the treatment of scoliosis in association with this disease have been published (Huebert & MacKinnon 1969, Hall et al. 1976). In the present work, neurologic complications after surgical treatment for scoliosis in a patient with syringomyelia are reported. Theories are proposed to explain the progression of syringomyelia in connection with the operative treatment of scoliosis, and special points are emphasized for the recognition of syringomyelia in patients with scoliosis.

CASE REPORT

The patient is a 15-year-old boy, normally delivered and with normal development. At the age of five, he refused for a period of 1 week to stand on his left leg and was thought to have meningitis. There were no known sequelea from this, and he was otherwise reported to be healthy with normal physical activity. At the age of 13, a right convex structural scoliosis was noted and considered to be idiopathic. The curve measured 37 degrees (Cobb); it corrected well in a Milwaukee brace to 28 degrees. The apex of the curve was at T5. In spite of adequate brace treatment the curve progressed, and after 2 years it was 55 degrees, extending from T1 to T10.

It was then decided to operate. When the boy was admitted to hospital 4 months later, the deformity had rapidly increased further and now measured 80 degrees in a standing A-P X-ray. His skeletal age was 14 years, his iliac apophyses had not yet appeared. Routine neurological examination was normal, with normal tendon reflexes in the upper and lower extremities, and normal sensibility and muscle tone and power.

At surgery the curve was corrected with a Harrington distraction rod inserted from T2 to L1, and fused. Nothing abnormal was experienced at the operation. However, because of unusual

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fragility of the laminae, a force of only 200 N was used for correction, while ordinarily 400 N is used (Nachemson & Elfström 1969). Postoperatively the curve measured 50 degrees.

Immediately after the operation the patient was well, with full sensibility and muscle function, voiding normally. Ten days later he complained of difficulty in voiding. Examination revealed a diminished sensibility for pain and touch at the T10 level and a positive Babinsky sign on the left side, but no paresis.

Six days later, however, the symptoms had progressed to spastic paraparesis and the patient was hardly able to lift his legs. There was diminished sensibility bilaterally from T10 distally. In addition, there was loss of the right biceps reflex and weakness of the triceps muscle of the right arm. Gas myelography showed a complete obstruction at the T10 level upwards, and obstruction at the cervical level from C5 and downwards, due to enlargement of the cervical medulla. The cervical spine had an increased sagittal diameter on X-ray. The diagnosis of syringomyelia was suspected.

The patient was again operated on 17 days after the scoliosis operation with decompressive laminectomy of T8 and T9. The dura was distended and the medulla was found to be bulging from several thin walled cysts containing a clear fluid, 2 cm^3 of which was aspirated. The Harrington rod was in the original position and the remaining distraction force amounted to only approximately 50 N. The rod was left in place to stabilize the fusion.

Postoperatively, the patient was ambulated in a Milwaukee brace. First, crutches were used because of paraparesis. There was a steady regression of symptoms and increase of muscle power. Six months after the scoliosis operation, the patient had normal muscle power in his arms; the legs still showed atrophy and slight weakness although muscle function tests all gave a normal (5) rating on a 0 to 5 scale. He walked without support, also on stairs, and had normal bladder and fecal control. The correction of the scoliosis remained unchanged at 50 degrees.

DISCUSSION

The deformity in this patient was first classified as an idiopathic curve, yet three traits were unusual for an idiopathic scoliosis. One was the high location of the curve in the spine, which occurs only in approximately 2 per cent of idiopathic cases (Nordwall 1973). Another was the sudden onset of a fast progression of the deformity, 25 degrees in 4 months. Such a fast progression is uncommon in idiopathic scoliosis. A third and most important unusual trait was the widened diameter of the cervical spinal canal, visible on plain radiograms. These signs would all make a neurological diagnosis probable, and are compatible with syringomyelia.

Various neurological diseases are often complicated by scoliosis and syringomyelia is considered to be one such cause of deformity. In different reports the incidence of scoliosis in syringomyelia has been from 20 to 80 per cent (Perret 1963, McIlroy & Richardson 1965, MacRae & Standen 1966), the incidence being higher and the scoliosis more severe if the neurological disease starts before the end of the growth period. The spinal deformity may appear before the onset of neurological symptoms (Hertel et al. 1973).

The only work describing operative treatment of scoliosis in connection with syringomyelia is that of Huebert & MacKinnon (1969). These authors report two cases treated surgically for scoliosis, one of which became paraplegic after the operation. As these authors point out there is obviously an increased risk in corrective operative treatment of these patients, and it is very important to recognize and differentiate patients with syringomyelia from those with idiopathic scoliosis before the operation.

Clinically, the first symptoms of syringomyelia are usually a dissociated disturbance of pain and temperature sensibility in one upper extremity. There is usually a slow progression, although a fast progression has been reported as well (McIlroy & Richardson 1965). The first symptoms can be explained by the location of cysts centrally in the cord. As the cavities multiply and grow, other parts of the medulla will be compressed or destroyed, producing muscle weakness, atrophy and spasticity.

Most commonly the cervical spine is involved. Bony anomalies are found in the upper part of the spine in approximately 25 per cent of these patients (MacRae & Standen

1966, Logue 1971) and often syringomyelia is associated with defects such as Arnold-Chiari deformity, basilar impression, spina bifida, myelomeningocele and Klippel-Feil syndrome. Also, patients with syringomelia developing at a young age have been shown to have an increased diameter of the cervical spine compared with normal (Wells et al. 1959, Hertel 1973).

Huebert & MacKinnon (1969) described a theory for the etiology of scoliosis in syringomyelia. They believe the cysts compromise the medial nuclei groups of cells which innervate the muscles of the trunk. However, the development of scoliosis does not depend solely on engagement of the motor neurons. In experimental work in animals, Liszka (1961) showed that division of the posterior nerve roots produced an even greater scoliosis than if the anterior roots alone were divided. This would fit well with the other sensory disturbances that these patients demonstrate in the early course of the disease. The location and extent of the cysts will probably determine whether scoliosis is an early sign or not.

The connection between the operative correction of the scoliosis and the occurrence of neurological symptoms must be considered. Williams (1969) proposed the theory that increase of intracranial pressure may cause enlargement of the cysts due to a valve-like mechanism in anomalies around the foramen magnum. This may occur during the operation and early postoperative period as there certainly will be transitory venous pressure changes within the spine and skull from coughing, apnea at induction of anesthesia, intubation and extubation, positive pressure breathing, lifting and positioning of the patient, etc.

Circulatory changes cannot be excluded as a pathogenic mechanism for the progression of neurological deficits in connection with the surgery. Domisse (1974) has shown that cord levels T4 to T10 are most sensitive to circulatory changes. The syringomyelia cysts may interfere with the circulation here, and this may be further compromised by the stretching of the spine in corrective surgery. If this is suspected the correction device should be immediately removed (MacEwen et al. 1975). In the present case, however, the gradual onset of symptoms over weeks, and the picture of compression of the pyramidal tracts did not imply a circulatory change and hence the Harrington rod was left in place to stabilize the fusion.

Consequently, in the presence of normal neurology, the following signs will point to the diagnosis of syringomyelia in scoliosis patients:

(1) Abnormal curve
(2) Very rapid progression
(3) Bony anomalies of the upper cervical or occipital region (X-ray of whole spine). Increased sagittal diameter or interpeduncular distance in the cervical spine on X-ray.

If syringomyelia is suspected in a patient who is to be surgically corrected, an extended neurological examination should include pain, temperature and touch sensibility tests of upper and lower extremities. Special roentgenograms of the upper cervical spine should be taken. A gas myelography should be carried out.

At surgery the correction should be moderate, the Harrington rod mainly used as a stabilizing device. The patient should be extremely carefully handled in lifting, turning and during the anesthesia.

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Correspondence to: Anders Nordwall, M.D., Dept. of Orthopaedic Surgery, Sahlgren Hospital, S-413 45 Göteborg, Sweden.