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Scoliosis associated with syringomyelia presenting in children

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Abstract. The clinical presentations and radiological features of scoliosis accompanying syringomyelia were analyzed in 14 cases of syringomyelia associated with a Chiari malformation in children. Scoliosis was the initial symptom in 11 out of 16 patients (64%) with syringomyelia and present in 14 (88%) at the initial examination. The scoliosis associated with syringomyelia was characterized by a higher incidence of a single curve (6 cases, 43%) and convexity to the left (7 cases, 50%) than seen in idiopathic scoliosis. The syrinx was shifted to the convex side of scoliosis on the axial section at the middle or lower thoracic level in patients with a single curve, and at the cervical or upper thoracic level in patients with a double curve. The authors think that the scoliosis develops in children as a result of damage done to the anterior horn, which innervates the muscles of the trunk, by an asymmetrically expanded syrinx.

Key words: Chiari malformation – Children – Magnetic resonance imaging – Scoliosis – Syringomyelia

It is known that syringomyelia is often associated with scoliosis [1–9] and that the incidence of scoliosis is especially high in children [2, 3, 5]. However, there have been only a few detailed reports about the scoliosis associated with syringomyelia [2, 6, 9]. The purpose of this article is to describe the clinical presentations and radiological features of the scoliosis associated with syringomyelia. The pathogenesis of the scoliosis is also discussed.

Patients, materials and methods

This series included 16 pediatric patients (8 male and 8 female, all younger than 15 years of age) with syringomyelia associated with a Chiari malformation who were treated surgically. None of the patients had spinal dysraphism. The age at onset ranged from 3 years to 15 years with a mean of 10 years. The average duration of symptoms preoperatively was 6 years (range 6 months to 40 years). Syringomyelia was diagnosed by computed tomographic (CT) myelography and/or magnetic resonance imaging (MRI). CT myelography was performed 6–24 h after myelography using a Somatom II scanner (Siemens). After April 1985, 14 patients were studied presurgically and postsurgically by MRI, using a Toshiba 0.15-T MR imager, a GE 1.5-T Signa MR imager, or a Siemens 1.5-T Magnetom MR imager, using surface coil. All patients were studied with T1-weighted spin-echo images in both sagittal and axial planes. Sixteen patients underwent 19 operative procedures for syringomyelia. A syringosubarachnoid shunt was performed in 15 patients, foramen magnum decompression in one, foramen magnum decompression with syringosubarachnoid shunt in one, ventriculoperitoneal shunt in one, and terminal syringostomy in one.

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Case reports

Case 1

Thoracic scoliosis was discovered in a 12-year-old girl during a medical examination at school, 1 year before admission to hospital. After that, she noticed numbness in the chest on the left side. A neurological examination showed diminished pin prick, temperature, and light touch sensibility from T4 to L3 on the left side. There was left-convex thoracic scoliosis in a single curve (30° by Cobb measurement; Fig. 1a). An MRI scan (Fig. 1b) demonstrated a Chiari I malformation and syringomyelia from C2 to T10. A syringosubarachnoid shunt was placed in the posterior midline at the C6–C7 level. The patient's postoperative course was uneventful, and her sensory disturbance improved. A postoperative MRI scan demonstrated a marked decrease in the size of the syrinx. Three years and 3 months after the operation, the radiograph of the spine showed that the curve decreased to 22°.

Case 2

A 8-year-old boy was found to have thoracic scoliosis 4 years before his admission to hospital. After that, he noticed weakness of his right upper extremity and gait disturbance. A neurological examination revealed weakness of the right upper and lower extremities. A reduction in pin prick, temperature and light touch sensibility below T6 on the left side was found. There was hyperreflexia in the

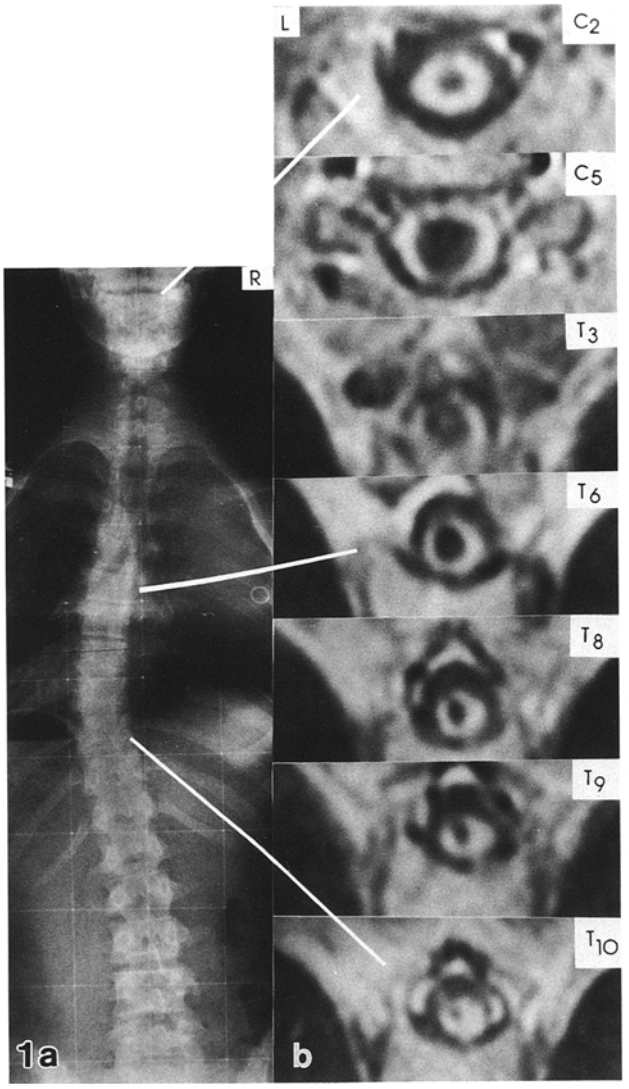

Fig. 1 a, b. Case 1. **a** Anteroposterior radiograph of the spine showing a left-convex thoracic scoliosis with a Cobb angle of 30°. The type of scoliotic curve was single. **b** Preoperative MRI scan (Toshiba 0.15-T; SE 500/40) demonstrating syringomyelia from C2 to T10. At the C2 and C5 levels, the syrinx was centrally located. At the T3, T6, T8, T9, and T10 levels, the syrinx was eccentrically placed to the left

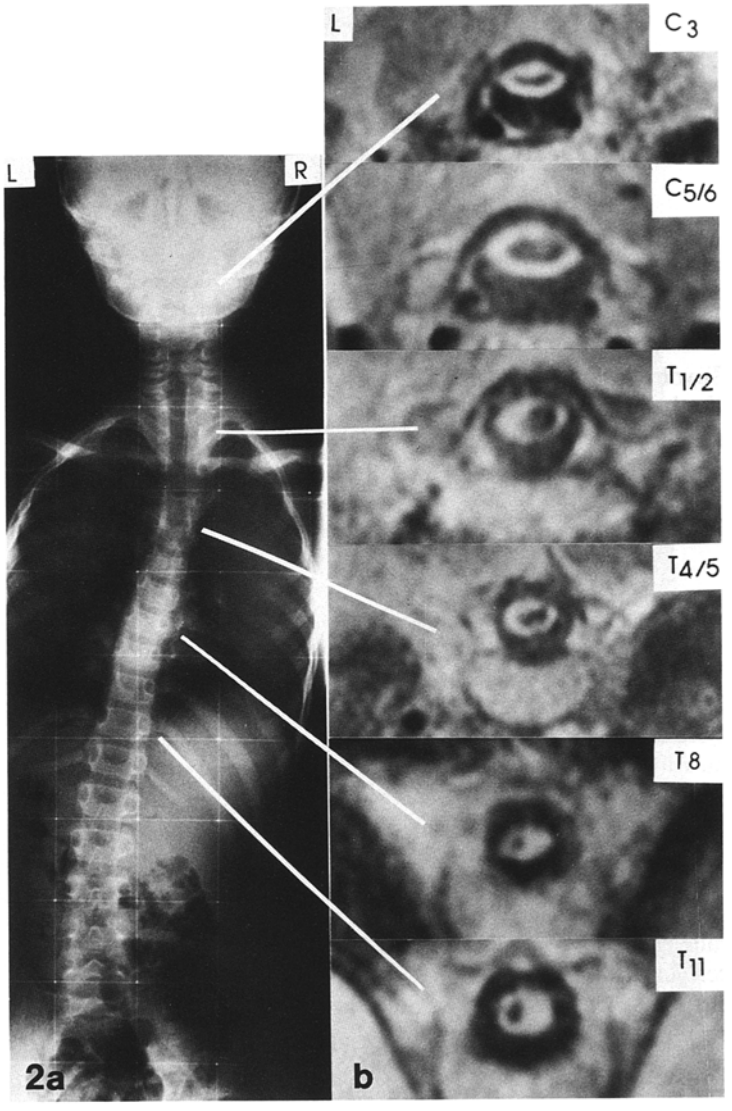

Fig. 2 a, b. Case 2. **a** Anteroposterior radiograph of the spine revealed thoracic scoliosis with a Cobb angle of 20°. The type of scoliotic curve was double. The major scoliotic curve was convex to the right and occurred at the upper thoracic level. **b** Preoperative MRI scan (Toshiba 0.15-T; SE 500/30) showing a syringomyelia from C2 to the conus medullaris. At the C3, C5–6, T1–2, and T4–5 levels the syrinx was found in the central portion, with extension into the right posterolateral portion. At the T6 and T11 levels, the syrinx was eccentrically located on the left

right leg and hyporeflexia in the left leg. The boy had thoracic scoliosis with a double curve (Fig. 2a) and pes cavus. The scoliotic curve measured 20°. An MRI scan (Fig. 2b) showed a Chiari I malformation and syringomyelia from C2 to the conus medullaris. A syringosubarachnoid shunt was placed along the dorsal root entry zone on the right side at C7/T1. Postoperatively, the weakness and the sensory disturbance improved. A postoperative MRI scan showed a reduction in the size of the syrinx.

Results

Incidence and clinical presentation of scoliosis associated with syringomyelia

The scoliosis was the initial symptom in 11 out of 16 patients (64%) with syringomyelia and present in 14 (88%) on admission. Three of the other 5 patients had pain or numbness, one patient had muscle weakness of the upper and lower extremities, and one had a foot deformity (pes cavus).

Table 1 shows the characteristics of scoliosis associated with syringomyelia. There was a single curve in 6 patients (43%) and a double curve in 8 (57%). In the patients with a single curve, the major curve of scoliosis occurred at the middle or lower thoracic level. In the patients with a double curve, the major curve occurred at the upper thoracic level and the minor curve at the thoraco-lumbar level was compensatory. The scoliosis was convex to the right in 7 out of 14 patients (50%) and to the left in 7. Three out of 14 patients had marked scoliosis

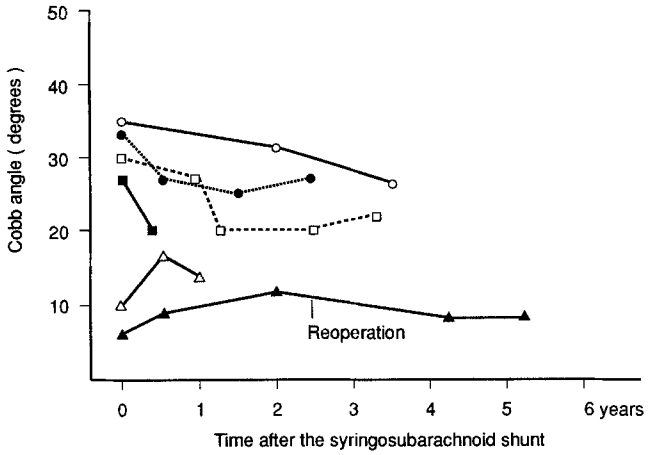

Fig. 3. Prognosis of scoliosis after operation for syringomyelia: data from six patients

Table 1. Characteristics of scoliosis associated with syringomyelia

Type of scoliotic curve:	
single curve	6 cases
double curve	8 cases
Right or left convex curve:	
right convex	7 cases
left convex	7 cases
Magnitude of scoliotic curve:	
marked scoliosis (>50° by Cobb measurement)	3 cases
moderate scoliosis (25°–50°)	7 cases
mild scoliosis (<25°)	4 cases

with a Cobb angle of over 50°. Moderate scoliosis (25°–50° by Cobb measurement) was found in 7 patients and mild scoliosis (0°–25° by Cobb measurement) in 4.

Relationship between location of the syrinx and scoliosis

The syrinx was visualized from the upper cervical to the lower thoracic level or the conus medullaris. The localization of the syrinx on the axial section varied with the level of the spinal cord, even in the same patient. In all 14 patients, the syrinx was eccentrically located in the cervical or thoracic level and shifted to the scoliotic convex side. In all 6 patients with a single curve, the eccentric part of the syrinx was located at middle or lower thoracic level. On the other hand, in all 8 patients with a double curve, the eccentric part was present at the cervical or upper thoracic level.

Prognosis of scoliosis after the operation for syringomyelia (Fig. 3)

The prognosis of scoliosis was analyzed in 6 patients in whom the curve of the scoliosis was measured preoperatively and postoperatively. The average follow-up period after the surgery for the syringomyelia was 2 years and 2 months (range 4 months to 3 years and 6 months). The Cobb angle decreased distinctly in 4 out of 6 patients, in the other 2 the Cobb angle markedly did not increase. These patients were managed in a brace and needed no orthopedic surgery after the operations for syringomyelia.

Discussion

It is known that syringomyelia is often associated with scoliosis [1–9] in addition to the characteristic clinical symptoms (temperature and pain disturbances of the suspended type, muscular atrophy and weakness of the upper limbs, and spastic paraparesis). Scoliosis has been previously reported in 44% of syringomyelia patients by McIlroy [6], followed by Logue [4] (31%), Schlesinger [7] (38%), and Tashiro [8] (50%). The incidence of scoliosis with syringomyelia is especially high in children [2, 3, 5] (87% in patients younger than 16 years of age and 48% in patients older than 16 years of age [5]). In our experience, scoliosis was observed as the initial symptom in 11 patients (64%) younger than 15 years of age. Moreover, at the initial hospital examination, scoliosis was detected in 14 patients (88%). Therefore, the authors want to emphasize that MRI scanning should be performed on children with idiopathic scoliosis of unknown cause and some evidence of a neurological deficit. MRI is a very useful technique for revealing the syrinx noninvasively.

The scoliosis associated with syringomyelia is explained by the unbalanced strength of paravertebral muscles [2]. Detailed examination of the relationship between the location of the syrinx and scoliosis was not possible until the advent of MRI. The axial location of the syrinx varies at different levels of the spine even in the same patient [3] and is often eccentrically located at the cervical or thoracic level [3]. The present study by MRI demonstrated that the syrinx extended along almost the entire thoracic level and was shifted on the axial section to the convex side of the scoliosis. It was therefore assumed that the anterior horn, which innervates the muscles of the trunk, had been damaged by an asymmetrically expanded syrinx [2]. More interestingly, there was a difference between the patients with a single curve and those with a double curve with regard to the location of the syrinx on axial section: the eccentric part of the syrinx was at the middle or lower thoracic level in the former group, whereas it was at the cervical or upper thoracic level in the latter. Thus, the major scoliotic curve occurred in each group at the level where the biased syrinx was present. The minor curve at the thoracolumbar vertebrae in patients with a double curve may be compensatory; when the syrinx is biased at the middle or lower thoracic level, no compensatory mechanism can work, so only a single curve results. In view of these facts, the authors think that the scoliosis associated with syringomyelia is characterized by a higher incidence of single curves and convexity to the left, compared with idiopathic scoliosis, which is typically convex to the right and has a double curve.

There has been only one published report about the prognosis of scoliosis associated with syringomyelia [2].

In our study group, all three patients who had marked scoliosis with a Cobb angle of over 50° underwent spinal fusion for scoliosis after the treatment for syringomyelia. On the other hand, all 11 patients who had mild or moderate scoliosis (less than 50° by Cobb measurement) were managed in a brace with good results and needed no orthopaedic operation. In 4 out of 6 patients in whom the curve of the scoliosis was measured preoperatively and postoperatively, the Cobb angle was distinctly reduced after surgery for syringomyelia, and in 2 the angle was controlled at a few degrees. In view of these facts, early treatment for syringomyelia is thought to be important for the prognosis of mild or moderate scoliosis associated with syringomyelia in children.

References

1. Cahan LD, Bentson JR (1982) Considerations in the diagnosis and treatment of syringomyelia and the Chiari malformation. J Neurosurg 57:24–31
2. Huebert HT, Mackinnon WB (1969) Syringomyelia and scoliosis. J Bone Joint Surg [Br] 51:338–343
3. Isu T, Iwasaki Y, Akino M, Abe H (1990) Hydrosyringomyelia associated with a Chiari malformation in children and adolescents. Neurosurgery 26:591–597
4. Logue V, Edwards MR (1981) Syringomyelia and its surgical treatment. J Neurol Neurosurg Psychiatry 44:273–284
5. MacRae DL, Standen J (1966) Roentgenologic findings in syringomyelia and hydromyelia. AJNR 98:695–703
6. McIlroy WJ, Richardson JC (1965) Syringomyelia. Can Med Assoc J 93:731–734
7. Schlesinger EB, Antunes JL, Michelsen WJ, Louis KM (1981) Hydromyelia. Neurosurgery 9:356–365
8. Tashiro K, Fukazawa T, Moriwaka F, Hamada T, Isu T, Iwasaki Y, Abe H (1987) Syringomyelic syndrome: clinical features in 31 cases confirmed by CT myelography or magnetic resonance imaging. J Neurol 235:26–30
9. Williams B (1979) Orthopaedic features in the presentation of syringomyelia. J Bone Joint Surg [Br] 61:314–324

Geographic editor's comment

The authors stress that the scoliosis associated with a syrinx should not be classified with idiopathic scoliosis, since it belongs to the group of paralytic scolioses. For this reason, MRI should be performed in children with scoliosis of unknown etiology. The majority of children with scoliosis may be found on MRI to have syringomyelia.

In four out of six patients reported on by the authors the scoliotic curve distinctly decreased after shunt surgery; however, the remaining two patients with scoliosis associated with syringomyelia did not improve after shunt surgery. One wonders why two out of six patients did not improve at all.

Satoshi Matsumoto