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### Case Report

## A Rare Course of Scoliosis Associated with Chiari Malformation and Syringomyelia

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Spinal deformity is an important clinical manifestation of Chiari I malformation (CM-I) and syringomyelia. Here we report the result of an 8-year follow-up of a 13-year-old girl with severe scoliosis associated with Chiari malformation and a large syringomyelia. The patient presented at our hospital at the age of 13 with a 68° scoliosis. Magnetic resonance imaging showed Chiari malformation and a large syringomyelia. Neurosurgical treatment involved foramen magnum decompression and partial C1 laminectomy, but the scoliosis still progressed. We present the first case report of a rare course of scoliosis in a patient with CM-I and a large syringomyelia.

**Key words:** Chiari I malformation, syringomyelia, scoliosis

Spinal deformities occur in 13% to 26% of patients with Chiari I malformation (CM-I) [1, 2]. The incidence of scoliosis increases to 71% to 85% when patients have both CM-I and syringomyelia [3, 4]. In cases initially believed to be idiopathic scoliosis, magnetic resonance imaging (MRI) has shown the incidence of CM-I at 14% to 23% [5, 6]. The standard treatment of CM-I and syringomyelia is foramen magnum decompression [7, 8], but this procedure does not reduce the syrinx in all cases, and further treatment is needed, such as syringosubarachnoid shunting [9].

Although scoliosis is typically best managed with bracing and observation, it can still be difficult to prevent spinal deformity with orthoses alone in patients with CM-I and syringomyelia because of the neuromuscular nature of this scoliosis curvature. When the deformity becomes severe, most reports recommend surgical management with spinal fusion

[3, 5]. We present the first case report of a rare course of scoliosis in a patient with CM-I and a large syringomyelia.

### Case Report

**Patient history.** The patient's delivery and medical history were normal. Her spinal deformity was diagnosed at another hospital at the age 12. One year later, her scoliosis had progressed and she presented to our hospital at the age of 13 due to severe spinal deformity.

**Physical examination.** On examination, she had no neurological deficits or skin abnormalities. She was 152 cm tall and weighed 40 kg. There was no severe hyperreflexia of her arms or legs and no abnormal abdominal reflex, but she had a severe rib hump and spinal deformity.

**Imaging.** Plain radiographs demonstrated the double thoracic curve. The proximal curve (T3-8)

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was  $68^\circ$  and the distal curve (T9–L1) was  $54^\circ$  (Fig. 1A). The kyphosis angle was  $35^\circ$  (Fig. 1B). In the traction and bending radiographs, the spinal deformity was somewhat rigid: the Cobb angles of the proximal and distal curves were reduced to  $35^\circ$  and  $40^\circ$ , respectively. The patient underwent MRI, which showed CM-I and a large syringomyelia from C2 to T10 (Fig. 2). We took 3D computed tomography reconstruction images for surgical planning in a computer-assisted procedure.

**Surgical intervention.** First we performed foramen magnum decompression and partial C1 laminectomy for the CM-I and syringomyelia according to Isu's method [4]. One year later, we performed a segmental pedicle screw fixation from T2 to L2 using the Stealth station navigation system® (Medtronic, Memphis, TN, USA) and achieved excellent curve correction. The accuracy of the navigation was 0.3mm. The operative time was 5h 45min, and the estimated blood loss was 1,500mL.

Postoperative radiograms demonstrated good correction of the curve in both the coronal and sagittal alignments. The postoperative Cobb angle was  $18^\circ$  in the proximal curve and  $18^\circ$  in the main curve in coronal plain films (Fig. 3). There were no postoperative

complications and no neurological compromise. She had good spinal balance and there were no neurological deficits postoperatively.

However, 1 year after the scoliosis surgery, the syringomyelia enlarged, and the patient experienced



Fig. 2 Preoperative MRI. Preoperative sagittal T1-weighted MRI shows the patient's type 1 Chiari malformation and large syringomyelia.

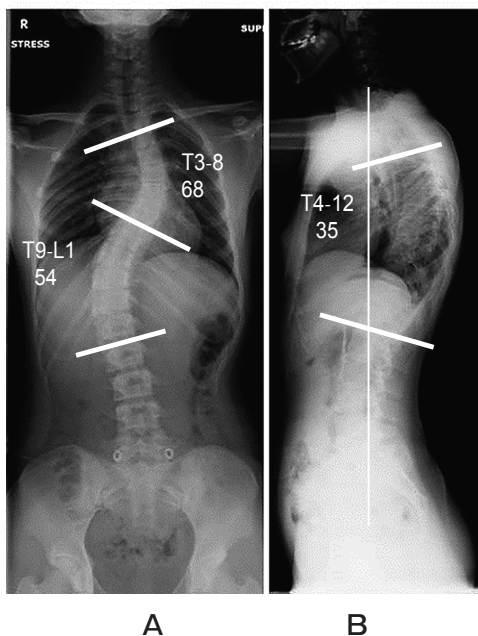


Fig. 1 Preoperative A-P, lateral, and bending radiography. A, The coronal Cobb angles were  $45^\circ$  in T3–8 and  $54^\circ$  in T9–L3; B, Lateral radiography shows the  $35^\circ$  kyphosis angle.

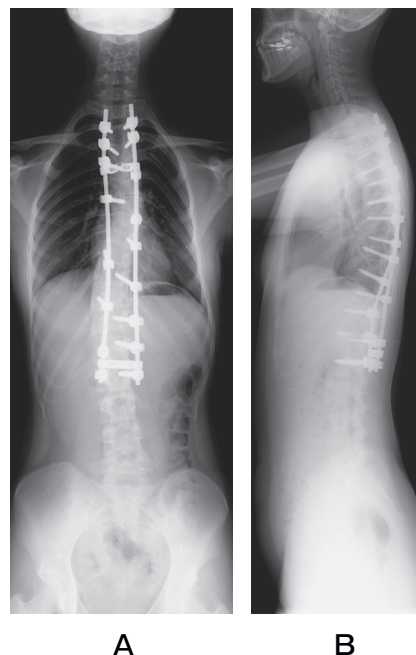


Fig. 3 Postoperative A-P, lateral radiography. A, The coronal Cobb angle was corrected from  $63^\circ$  to  $18^\circ$ ; B, The sagittal alignment became normal.

headache and numbness of both arms; we performed syringosubarachnoid shunting. After the syringosubarachnoid shunting, the syringomyelia was reduced and her symptoms were relieved (Fig. 4). Eight years after her first operation, she has maintained good spinal balance and there have been no symptoms of syringomyelia.

## Discussion

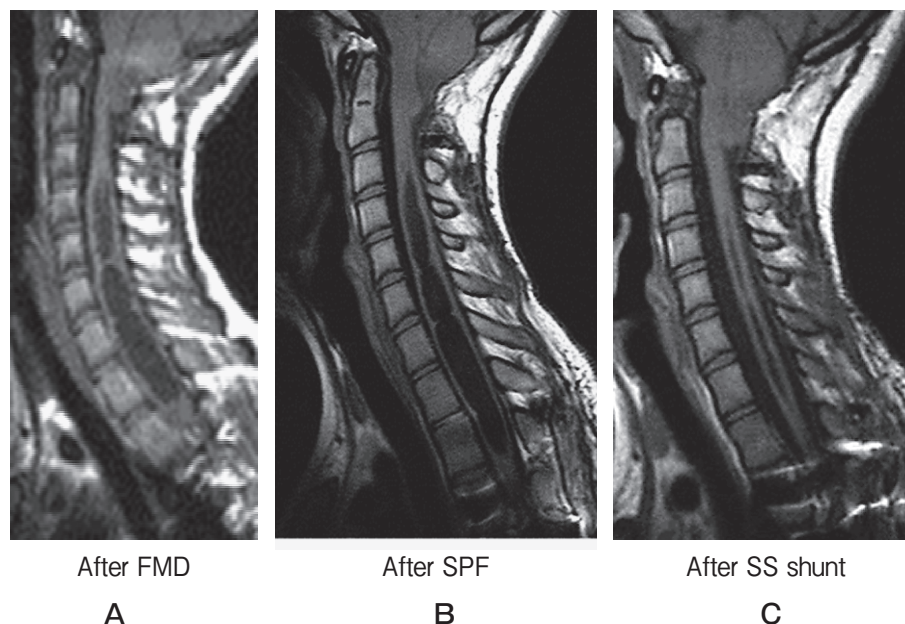
### *Relationship between scoliosis and CM-I.*

A subset of patients with idiopathic scoliosis may have an underlying neurological abnormality (most commonly syringomyelia with CM-I) despite a normal history and physical examination [5, 6]. CM-I is associated with syringomyelia in 50% to 75% of patients [10], and scoliosis has been reported in more than two-thirds of patients with CM-I associated with syringomyelia [5]. Asymptomatic children with CM-I and syringomyelia may benefit from conservative management with neurological and MRI follow-up [6, 11] because the natural history and clinical course of syringomyelia are extremely variable, and the spontaneous resolution of CM-I associated syringomyelia can occur in some cases [12, 13].

However, there is still controversy as to whether

early surgical intervention for syringomyelia is beneficial in preventing scoliosis curve progression in patients with mild to intermediate scoliosis. Some reports state that the syringomyelia does not affect the progression of scoliosis associated with CM-I [6], and some authors do not recommend surgical intervention for scoliosis in patients with CM-I and syringomyelia until neurologic problems occur [5]. On the other hand, there are some reports that approx. 50% of patients with CM-I-associated scoliosis improve or stabilize after foramen magnum decompression [1, 2], sometimes with complete straightening of the scoliosis [14]. We recommend early cervicomedullary decompression for a patient whose scoliosis curve is more than 25°, because bracing is necessary for such a patient.

**Surgical intervention for CM-I.** Patients with CM-I present most commonly with a chief complaint of impaired oropharyngeal function, scoliosis, headache or neck pain, sensory disturbance and weakness [14]. There are several treatments for patients with syringomyelia associated with CM-I, such as foramen magnum decompression with or without obex plugging [7] and placement of a syringoperitoneal [15], syringosubarachnoid [9] or thecoperitoneal shunt [16]. The most common and safest procedure is



**Fig. 4** Postoperative MRI. **A**, After the foramen magnum decompression; **B**, After the segmental pedicle screw fixation for scoliosis correction; **C**, After the syringosubarachnoid shunting, the syrinx was markedly reduced.

foramen magnum decompression, because shunt procedures may induce arachnoiditis and shunt dysfunction in as many as 49% of patients [17]. However, shunting can achieve early reduction in cavitory size. Since the surgical outcome of foramen magnum decompression is very reliable and safe for CM-I, it was our first surgical choice. However, it may not reduce the syrinx in all cases, necessitating further treatment such as the syringosubarachnoid shunting we performed in the present case.

### ***Surgical intervention for scoliosis with CM-I.***

The literature emphasizes the importance of the early diagnosis of CM-I malformation with syringomyelia in very young children with scoliosis [18]. The physical examination should concentrate on identifying an abnormal gag reflex for detecting the syrinx. Even if the syrinx is documented at an early stage, the timing for performing a foramen magnum decompression is difficult to determine. Most surgeons advocate addressing CM-I and syringomyelia as an initial step in managing scoliosis [1] for the purpose of halting curve progression and avoiding iatrogenic neurological deterioration during deformity correction.

If the syringomyelia is large and does not diminish after surgical intervention, it is difficult to perform the scoliosis correction because of the possible complication of paraplegia after scoliosis surgery. However, when the spinal deformity continues to progress, as in the present case, the patient cannot wait for the syrinx to gradually reduce. A recent report recommends a one-stage deformity correction without treating the syrinx [19].

In conclusion, spinal deformity is an important clinical manifestation of Chiari I malformation with a large syringomyelia, but foramen magnum decompression is sometimes ineffective for a patient who has CM-I associated with a large syringomyelia. In that situation, the patient needs syringosubarachnoid shunting to reduce the size of the syrinx and its inherent symptoms. To achieve rigid fixation, posterior segmental pedicle screw fixation was beneficial for the present patient with CM-I and syringomyelia.

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