Acta Neurochir (Wien) (2004) 146: 187–191
DOI 10.1007/s00701-003-0177-0

Acta Neurochirurgica
Printed in Austria

Case Report

Spontaneous resolution of a Chiari I malformation associated syringomyelia in a child

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Published online January 8, 2004

Summary

A child with complete spontaneous resolution of a Chiari I malformation associated Syringomyelia without surgical intervention is presented. The child was followed clinically by serial magnetic resonance imaging (MRI) and remains neurologically stable after 8-years of follow-up. To our knowledge, only 6 pediatric cases with spontaneous resolution of a spinal cord syrinx documented by MRI without surgical intervention have been reported. This case is of interest in the light of the postulated theories to explain spontaneous resolution of syringomyelia.

Keywords: Syringomyelia; Chiari I malformation; spontaneous resolution; spinal cord.

Introduction

Traditionally, Chiari I malformation is defined as a congenital rather than an acquired defect. Chiari described type I malformations as ''elongations of the tonsils and medial part of the inferior lobes of the cerebellum which go along with the medulla into the cervical canal''. These malformations are associated with syringomyelia in 50% to 75% of patients [24]. More recently, cases of ''acquired'' Chiari I malformation have been documented in patients with supratentorial mass lesions [36] and after removal of cerebrospinal fluid (CSF) via a spinal shunting procedure or multiple lumbar [8, 39, 44] punctures.

Since the development of magnetic resonance imaging (MRI), Chiari I malformation and syringomyelia is being diagnosed with increasing frequency in the pediatric population [4, 9, 10, 19, 29, 37].

The natural history of Chiari I malformation is unpredictable. A clinical spectrum has been described in this age group.

We present a 6-year-old girl with Chiari I malformation and syringomyelia from C3 to T10 on MRI and thoracic scoliosis with convexity to the right. The neurological examination was normal. Eight years after her initial examination the MRI demonstrated complete and spontaneous resolution of syringomyelia and improvement of Chiari without surgery. Spontaneous resolution of syringomyelia and Chiari I malformation is rare and may be observed only if CSF flow obstruction resolves spontaneously. In young children, a period of observation is therefore warranted. If the tonsils rise together with the child's continuing cranial growth, surgery may not be required. If the child's symptoms persist or progress however, surgery should be recommended.

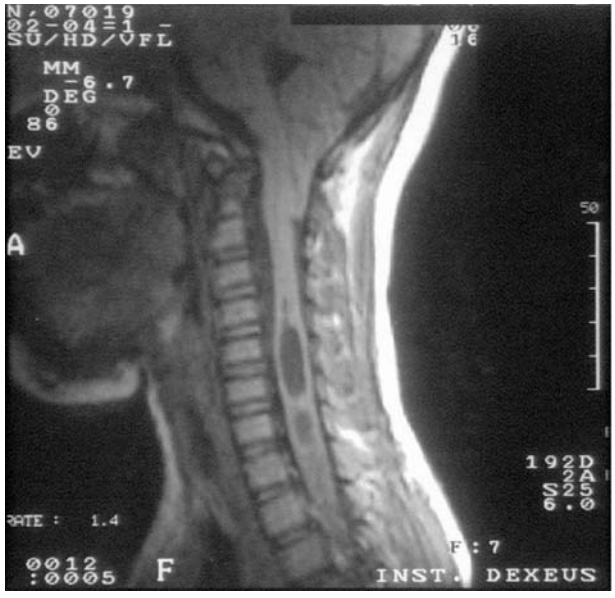
Clinical presentation

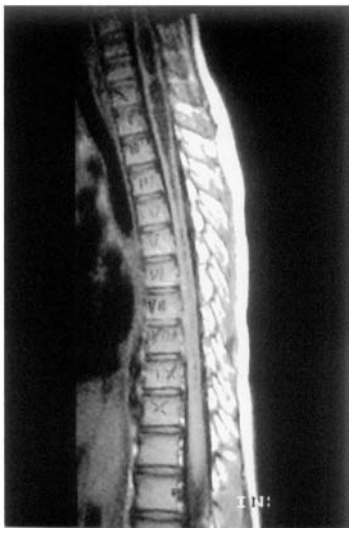
A 6-year-old girl presented with a 6-months history of scoliosis, without any other symptomatology. No personal and family history report was documented. The neurological examination revealed a right dorso-lumbar scoliosis without any neurological disorder. The child had a 30° scoliosis in that time. A corset was prescribed. The patient exhibited progression of her curvature from 30 to 48° in 8 years. MRI revealed a Chiari I malformation with syringomyelia between C3 and T10 (Figs. 1 and 2). No hydrocephalus was observed. Periodical follow up was done either by the neurosurgeon, the orthopedist and the radiologist.

The neurophysiological somatosensory evoked potentials study (SEP) revealed an increase of latency and decrease of posterior tibial muscle amplitude and the median nerve somatosensory evoked potentials revealed a lower amplitude of N13.

No symptomatology appeared, therefore only clinical observation was undertaken. MRI was repeated 6 years later, which demonstrated a near complete spontaneous resolution of syringomyelia (Fig. 3).

Another MRI was done 2 years later, which revealed a complete resolution of Chiari I malformation associated syringomyelia.





Figs. 1 and 2. Sagittal T1-weighted MRI of the cervical and thoracic spine showing a Chiari I malformation and a large cervico-thoracic syrinx

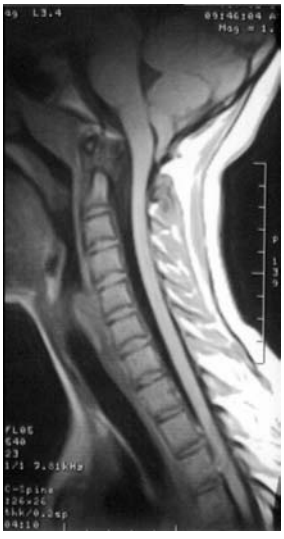


Fig. 3. Sagittal T1-weighted MRI showing almost complete resolution of the syrinx

Orthopedic surgery for the scoliosis was performed at 14-years of age. The patient remains asymptomatic.

Discussion

The Chiari type I malformation, as originally described in 1891, is defined as a congenital herniation of the cerebellar tonsils through the foramen magnum associated with a normal hindbrain [6, 7]. More recently, cases of ''acquired'' Chiari I malformation have been documented in patients with supratentorial mass lesions [18] and after removal of CSF via a spinal shunting procedure or multiple lumbar [17] punctures. The Chiari I malformation is associated with syringomyelia in 50% to 75% of patients [11].

To our knowledge, there have been 6 pediatric [1, 2, 40–42, 48] (Table 1) and 6 adult [5, 20, 21, 32, 33, 38] cases reported dealing with spontaneous resolution of

Table 1

Author	Age	Syrinx location	Initial Chiari I?	Follow-up	Neurological symptoms at follow-up	Chiari I malformation resolved?
Sudo, 1990	14 years	cervicothoracic	yes	2 1/4 years	unchanged	yes
Yeager, 1992	19 months	cervical	no	2 years	asymptomatic	–
Avellino, 1996	5 years	cervicothoracic	yes	6 years	asymptomatic	improved
Avellino, 1998	5 years	cervicothoracic	yes	11 years	unchanged	improved
Sun-J, 2000	11 years	cervicothoracic	yes	2 years	asymptomatic	improved
Sun-P, 2001	7 years	cervical	yes	6 years	unchanged	yes
Guillen, 2001	6 years	cervicothoracic	yes	8 years	asymptomatic	improved

syringomyelia as documented by MRI and no surgical intervention. Sudo *et al.* [40] reported a case of a 14-year old male with a cervicothoracic syrinx and Chiari I malformation. At 2 years of follow-up, the patient improved neurologically, and spontaneous resolution of the syrinx and Chiari I malformation were noted.

Yeager and Lusser [48] reported the case of an asymptomatic 19-month-old girl with a cervical syrinx which completely and spontaneously resolved at 2 years follow-up. Avellino *et al.* [1] reported an asymptomatic 5-year-old boy with cervical and thoracic syringes as well as Chiari I malformation without any likely antecedent history who experienced spontaneous resolution of the syringes and improvement of the Chiari I malformation over a 6-year period and a 5-year-old female with cervicothoracic syrinx and Chiari I malformation. The patient improved neurologically and spontaneous resolution of the syrinx at 11 years follow-up was observed.

Sun [41, 42] reported 2 cases. The first involved a 11 year-old male with a cervicothoracic syrinx and Chiari I malformation, the patient remained neurologically unchanged, and near disappearance of the syrinx and a persistent Chiari I malformation were observed at 2 years follow-up. The second involved a 7 year old male with a cervical syrinx and Chiari I malformation. Over a 6-year period, the Chiari I malformation persisted, and near disappearance of the syrinx was noted.

From reviewing the literature, resolution of the syrinx was associated with improvement in the Chiari I malformation in all the reported pediatric cases who had an initial Chiari I malformation. In contrast, in the reported adult cases, resolution of the syrinx was associated with improvement in the Chiari I malformation in 2 of the 7 cases with complete resolution.

In our case, the mechanism by which the syrinx was greatly reduced in size is unknown. A possible explanation may be that the patient has grown in height and skull size over a 6-year period. Perhaps a change at the craniovertebral junction and cervical cord anatomy lead to normalization of the craniospinal differential pressure with subsequent correction of the physiopathology and dissappearance of the syrinx [1].

To date, very few cases of spontaneous resolution of syringomyelia in patients with Chiari I malformation have been documented accurately by MRI.

There is evidence that syringomyelia develops in association with obstruction of CSF flow at the foramen magnum in Chiari I malformation [3, 22, 23, 35, 43]. In children, the growth of the cranium continues after the cerebellum has reached more than 90% of its final size

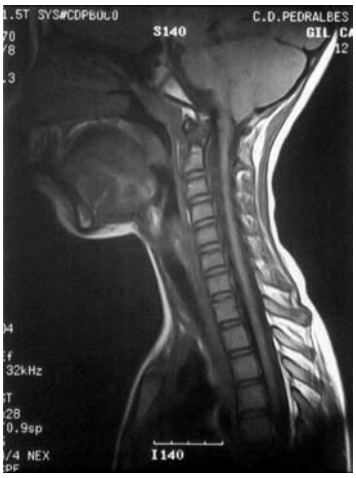


Fig. 4. Sagittal T1-weighted MRI eight years later showing a complete ressolution of Chiari I malformation associated syringomyelia

by 2 years of age [34]. Therefore, the tonsils may regress intracranially in relation to the growing cranium [28] to the extent that free CSF passage is established, allowing the syrinx to decrease in size or even disappear completely.

A second possible mechanism involves changes in venous flow in the posterior fossa. Girard [16] demonstrated that Chiari I malformation is a regular feature in children with vein of Galen aneurysms due to veno-occlusive disease. After successful embolization of the venous malformation, the Chiari I and Syringomyelia – if present- disappeared. One may speculate that venous thrombosis involving the posterior fossa may cause descent of the cerebellar tonsils into the foramen magnum and that, after recanalization, the tonsils may reposition intracranially.

Another possible mechanism of spontaneous resolution involves supratentorial lesions such a hydrocephalus or other space-occupying processes.

Decompression of the syringomyelia into the spinal arachnoid space may occur through a tear in the spinal cord caused by the increased pressure within the syringomyelia during Valsalva-like maneuvers. MRI has been used to document this in one case [38].

The pathogenesis of syringomyelia associated with Chiari I malformation still is not fully understood. Gardner's hydrodynamic theory [12–15] and Williams's hypothesis of craniocervical pressure dissociation [45–47], postulate communication between the fourth ventricle and the syrinx via the obex and the central canal. Neuropathological studies [25, 26, 30] have shown that such communication does not exist in the overwhelming majority of patients.

Welch *et al.* [44] showed a descent of the cerebellar tonsils in patients who underwent lumbo-peritoneal shunting, and therefore assumed that the difficulty in balancing intracranial and intraspinal pressure could cause such anomalies.

These authors stressed the possibility of a secondary, and not only congenital, origin of Chiari I disease.

In 1994 Oldfield [31] proposed that the Chiari anomalies could induce a piston-like motion affecting the cerebellar tonsils, causing a systolic wave in the CSF flow, which would affect the spinal cord inducing CSF leakage through interstitial and perivascular spaces.

In 1995 Milhorat [27] suggested that constriction of the aperture of the upper end of the central canal leads to noncommunicating syringomyelia.

Conclusions

The natural history and clinical course of syringomyelia are extremely variable. Spontaneous resolution of Chiari I malformation associated syringomyelia can occur. However, the cause of the resolution remains unclear. Although in children the resolution of syringomyelia is often accompanied by an improvement of the Chiari I malformation, this is not the case in adults. Our case contributes further to the growing body of evidence that the natural history of Chiari type I malformation associated with syringomyelia is not necessarily towards progression. Asymptomatic children with Chiari type I malformation and syringomyelia may benefit from conservative management with neurological and MRI follow-up [35].

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Comment

In this case report, the authors from Barcelona, Spain describe spontaneous resolution of a cervicothroacic syrinx in a six year old girl who presented with scoliosis and a Chiari-I hindbrain malformation. Although this finding has been described previously, it is an infrequent occurrence and the present article adds to our understanding of the natural history of Chiari-I and hydrosyringomyelia. Guillen and Costa remind us that progression of the spinal cord cavitation is not inevitable in children with Chiari-I.

The manuscript is well-written and well-illustrated. I think it will be of interest to the readers of *Acta Neurochirurgica.*

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