

Spontaneous Syringomyelia Resolution at an Adult Chiari Type 1 Malformation

Kendiliğinden Emilen Syringomyeli ve Erişkin Chiari Tip 1 Birlikteliği

ABSTRACT

We present a 41-year-old man with Chiari type 1 malformation and cervical syringomyelia. Although the tonsillar herniation persisted, his syringomyelia was almost completely resolved during an eleven-year time period without surgery.

KEYWORDS: Chiari type 1 malformation, Syringomyelia, Treatment, Pathogenesis

ÖZ

Chiari malformasyonu, syringomyeli gelişimine sebep olabilir. Yazıda 41 yaşında Chiari tip 1 malformasyonlu hasta ele alınmıştır. Hastanın tonsiller herniasyonunun sürmesine rağmen, servikal syringomyelisinin 11 yıllık bir süreçte kendiliğinden hemen tamamen düzeldiği görülmüştür.

ANAHTAR SÖZCÜKLER: Chiari tip 1 malformasyonu, Syringomyeli, Tedavi, Patogenez

Fatih Ersay DENİZ

Erol ÖKSÜZ

Gaziosmanpaşa Üniversitesi Tıp Fakültesi,
Beyin ve Sinir Cerrahisi, Tokat, Turkey

Received : 16.09.2008

Accepted : 12.12.2008

Correspondence address:

Fatih Ersay DENİZ

E-mail: ersaymd@yahoo.com

INTRODUCTION

Chiari type 1 malformation is the congenital downward displacement of the cerebellar tonsils from the foramen magnum and is associated with syringomyelia in about 40% of patients (1). Syringomyelia is the presence of a cavity in the spinal cord and was first used by Ollivier d'Angers in 1827. The exact pathogenesis and the natural history of syringomyelia is not known. It may often be related to trauma, tumor, congenital abnormalities or it may be idiopathic syringomyelia (6). Although it is generally accepted that the presence of a syringomyelia with Chiari type 1 malformation is a strong indication for surgical decompression as the syrinx will progress, even if it is asymptomatic, some patients show no change in symptoms over many years.

We describe a case of spontaneous resolution of syringomyelia in an adult patient with Chiari type 1 malformation.

CASE REPORT

A 41-year-old man visited our hospital with complaints of pain in the neck and the head. He had epilepsy which was under control with drugs. He had no history of trauma and the neurologic examination was normal. The previous magnetic resonance imaging (MRI), which was performed 11 years ago, showed Chiari type 1 malformation, bulging of the C5-C6 disc and an associated cervical syringomyelia (Figure 1). No symptomatology appeared; therefore he refused to be operated. A repeat MRI was performed to evaluate the Chiari type 1 malformation and the syringomyelia. His initial MRI demonstrated a spontaneous decrease in syrinx size, without improvement of Chiari type 1 malformation. The C5-C6 disc bulge appeared unchanged (Figure 2).

DISCUSSION

Although Chiari type 1 malformation and syringomyelia association is a well known entity, the natural history of syringomyelia is unpredictable. Some patients may experience improvement or stabilization without surgery. About thirty patients where spontaneous resolution of syringomyelia was documented by MRI without surgical intervention have been reported. As results of surgery are often poor after neurological deficit development, a prophylactic decompression is generally considered.

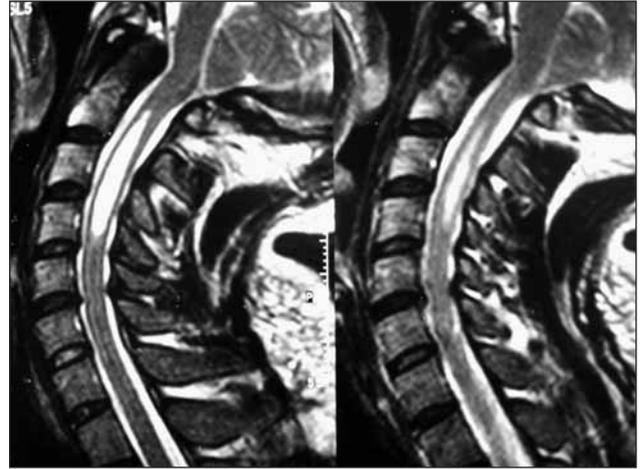


Figure 1. Sagittal T2-weighted MRI showing Chiari type 1, Syringomyelia from C2 to C4, C5-6 disc bulging.



Figure 2. Sagittal T2-weighted MRI showing Chiari type 1, near complete resolution of syringomyelia, C5-6 disc bulging.

Heiss et al. prospectively studied 20 adult patients with Chiari type 1 malformation and syringomyelia association, and concluded that the disappearance of the abnormal shape and position of the tonsils after simple decompressive extraarachnoid surgery suggests that the Chiari type 1 malformation of the cerebellar tonsils is acquired, not congenital (3). Girard et al. reported eight children of vein of Galen aneurysm and Chiari type 1 association. They concluded that posterior fossa hydrovenous congestion is a result of inadequate venous drainage and that the tonsillar descent is reversible if adequate venous drainage is reconstituted following therapeutic embolization of the fistula (2). Hoffman et al. detected descent of the cerebellar tonsils in patients who underwent lumbo-peritoneal shunting, and it was thought that the

difficulty in balancing intracranial and intraspinal pressure could cause such anomalies (4). The hydrodynamic theory, the craniospinal dissociation theory, and the tonsillar piston theory rely on the cerebrospinal fluid (CSF) pressure pulsations or venous pressure changes in the cranium and spinal subarachnoid space during coughing, straining, or valsalva manoeuvres and expanding the syrinx (6). Oldfield et al. investigated 7 patients of Chiari type 1 associated with syringomyelia and proposed that the descended tonsils obstruct the CSF flow to and from the spinal compartment at the foramen magnum thus explaining the appearance of syringomyelia. They also reported that none of the patients had evidence of a patent communication between the fourth ventricle and the syrinx on MRI and intraoperative ultrasound studies (5). Regression of the tonsils intracranially due to a growing cranium at a child or due to an atrophy development at an adult may cause to improve CSF circulation and syringomyelia resolution. Santoro et al. determined that fissuring of the cord parenchyma is instrumental in the spontaneous resolution of syringomyelia (7).

In our case, the mechanism of the spontaneous syringomyelia resolution is unknown. It may be speculated as our patient had epilepsy that there may be a communication between the syringomyelia and the subarachnoid space during an episode of increased pressure accompanying an epileptic attack, or just the opposite; a seizure free period may reveal a stability of intracranial and intraspinal pressure changes, resulting in syringomyelia resolution.

The pathogenesis of syringomyelia associated with Chiari type 1 malformation and its spontaneous resolution is not fully understood. With the increasing availability of MRI, the incidental diagnosis of syringomyelia in asymptomatic patients is becoming more frequent. The evidence that syringomyelia can spontaneously resolve makes the treatment strategy controversial. Since the outcome of surgical treatment for patients with syringomyelia is not always satisfactory and also some patients remain neurologically stable without surgery, clinical and radiological follow-up may be adequate for patients without progressive symptoms.

REFERENCES

1. Elster AD, Chen MY: Chiari I malformations: Clinical and radiologic reappraisal. *Radiology* 183:347-353, 1992
2. Girard N, Lasjaunias P, Taylor W: Reversible tonsillar prolapse in vein of Galeno aneurismal malformations. *Child Nerv Syst* 10:141-147, 1994
3. Heiss JD, Patronas N, DeVroom HL, Shawker T, Ennis R, Kammerer W, Eidsath A, Talbot T, Morris J, Eskioglu E, Oldfield EH: Elucidating the pathophysiology of syringomyelia. *J Neurosurg* 91(4):553-562, 1999
4. Hoffman HJ, Tucker WS: Cephalocranial disproportion. A complication of the treatment of hydrocephalus in children. *Child Brain* 2: 167-176, 1976
5. Oldfield E, Muraszko K, Shawker T, Patronas N: Pathophysiology of syringomyelia associated with Chiari I malformation of the cerebellar tonsils. *J Neurosurg* 80: 3-15, 1994
6. Ozişik PA, Hazer B, Ziyal IM, Ozcan OE: Spontaneous resolution of syringomyelia without Chiari malformation. *Neurol Med Chir (Tokyo)* 46:512-517, 2006
7. Santoro A, Delfini R, Innocenzi G, Di Biansi C, Trangimeni G, Gualdi G: Spontaneous drainage of syringomyelia. Report of two cases. *J Neurosurg* 79:132-134, 1993