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CASE REPORT

Vertigo associated with Chiari I malformation and syringomyelia

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Abstract: We report a rare case of a patient with late onset of Chiari I malformation with associated syringomyelia that was successfully treated with foramen magnum decompression. Our patient is presenting initially with vertigo and gradual weakening of the left-hand gross motor ability, gradual hypesthesia. Magnetic resonance imaging demonstrated a Chiari I malformation with syringomyelia. Posterior fossa decompression, C1 laminectomy and duroplasty were performed. After surgery, the vertigo completely resolved *(Fig. 2, Ref. 13).*
Full Text (Free, PDF) www.bmj.sk.
Key words: vertigo, Chiari I malformation, syringomyelia, MRI.

The Chiari malformation type I (CMI) being a malformation of the posterior cranial fossa often leads to syringomyelia. CMI is a congenital disorder characterized by caudal displacement of the cerebellar tonsils through the foramen magnum into the spinal canal. Syringomyelia is characterized by the presence of cystic cavities inside the spinal cord. The cavities are usually located inside the cervical cord, although they can extend upwards and/or downwards (1–3). The signs and symptoms in patients presenting cerebellar syndrome may lead to a misdiagnosis of insufficiency of the vertebrobasilar circulation. The MRI is the method of choice in the diagnosis of CMI (4–6). Conservative treatment is not recommended as the surgical procedure stops the progress of the disease with clinical improvement being the rule (2, 7). Our study provides evidence that the main benefit of the surgical management in patients with CMI with syringomyelia is to arrest the progression of the disease.

Case report

44 year old woman, complaining of vertigos, insecurity and instability over the last three and a half years. In the neurological status a painful tension of the paravertebral musculature at retroflexion was detected. The myotatic reflexes are symmetrical, there are no pathological reflexes. In the Romberg test, mild oscillations were observed. In the Utenberg test, there is an insecurity, similar as in the Babinski-Weill test. The left-hand gross motor ability is weakened, the left 4th and 5th finger flexors are weakened. Hypaesthesia on the left side from C4 to C8 was detected. X-rays of the cervical spine indicate arthrotic changes, arthrosis of small articulations, with narrowing of the intervertebral space (i.v.) C2–3 and congenital block. The neck vessels Doppler showed the left vertebral artery to be more gracile. There is an evident applanation of the systolic peak and vascular noise as an indication of external pressure caused by spondylodegenerative changes. The electromyography (EMG) indicates a moderate chronic radicular bilateral lesion of the C7 root and an initial right-side lesion of the C6 and C8 roots. The computerised tomography (CT) was performed by a spiral CT apparatus Simens Somatom EMOTION 2000. In the imaging of C5 to C7, at the C5–C6 level there is a lesser mediolateral left-sided protrusion of the i.v. disc, reducing the frontal subarachnoidal space. The lateral recessuses are free. The somatosensory evoked potentials (SSEP) of median nerve and tibial nerve are normal.

After a year and a half, the discomforts increased, the vertigo became more intensive, in both non-physiological positions and sudden movements of the head. In antigravitational tests, the patient reports intensive subjective feeling of weight in the upper extremities. The musculus biceps brachii reflex is weakened bilaterally, with increased myotatic reflexes in legs, but there are no pathological reflexes. Hypaesthesia of the right side of the face and in the left dermatomes C4 to C8 was reported. The left hand gross motor ability is significantly weakened. The left 3rd and 4th finger flexors are weakened. In Romberg test, oscillations with no clear lateralisation were detected. SSEP of the median nerve and tibial nerve show evoked potentials of extended latencies and lowered amplitudes. MRI of the cervical and the thoracic spine, performed by Simens Simphony 2002, 1.5 T, shows from the C1 vertebra caudally to the visible thoracolumbar segment, that is, to Th12, signs of syringomyelia with evi-

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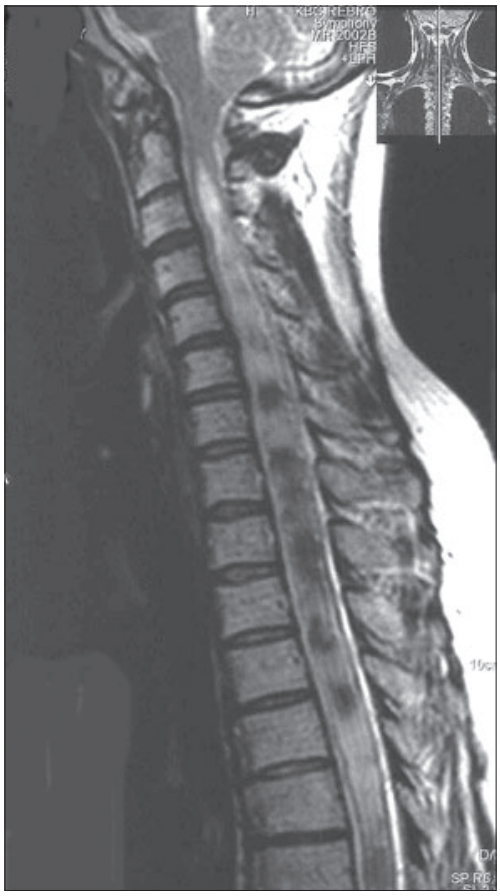

Fig. 1. MRI of the craniocervical passage and cervical spine – lowered cerebellar tonsils, Chiari I and syringomyelia of cervical spine (T2-weighted, sagital).

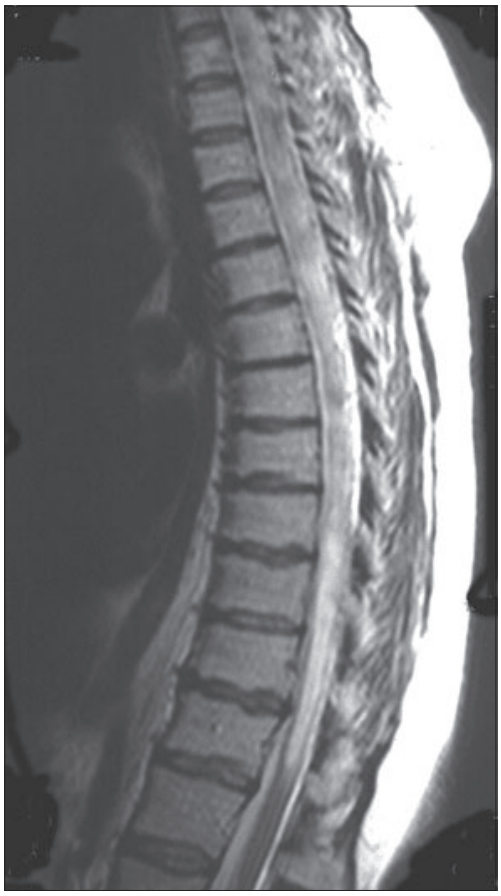

Fig. 2. MRI of the thoracic spine – syringomyielia (T2-weighted, sagital).

dent fusiform cystic widening running practically along the entire medulla. There are visibly lowered cerebellar tonsils, corresponding to the Chiari I syndrome (Figs 1 and 2).

According to the clinical status and neuroradiological examination, the patient was indicated for surgery. The uniform posterior craniovertebral decompression consisted of a small suboccipital craniectomy, a C-1 laminectomy, microsurgical reduction of the cerebellar tonsils, and dural closure with a synthetic dural graft to increase the cerebrospinal fluid space at the craniocervical junction.

The postoperative recovery went with no complications, and physical therapy was administered. Three months after the surgery, a follow-up clinical examination was performed, where the patient denied vertigo and instability. In antigravitational tests, extremities were maintained normally. She reports hypaesthesia of the right side of the face and in the dermatomes C4 to C8 at left. The left-hand gross motor ability is mildly weakened. The left 3rd and 4th finger flexors are mildly weakened. In Romberg test stabile. Six months after the surgery, the neurological findings are identical. She denies vertigos and instability. The overall condition is significantly improved, as well as the life quality according to the patient's personal assessment.

Discussion

Syringomyelia is characterized by the presence of cystic cavities inside the spinal cord, with an estimated incidence of 8.4 new cases/year/100 000 people. Four different main types may be described in descending order of frequency: associated with Chiari I malformations, associated with vertebral trauma, associated with vertebral trauma, associated with basilar invagination and associated with hydrocephalus (8, 9).

The type I Chiari malformation consists of an caudal descent of the cerebellar tonsils through the foramen magnum towards the spinal cervical channel (10). Signs and symptoms in patients with CMI including headache, neck pain, nystagmus, vertigo, weakness, spasticity, atrophy, numbness, pain and temperature dissociation, diplopia, dysphagia, and sphincter dysfunction (4).

Authors present experience with neurological manifestations, positional vertigo and dizziness, in which MRI showed a type I Chiari malformation (10). In our patient, the main symptom was vertigo, other neurological disorders to have been noticed only in the course of gradual development of the disease. Degenerative changes of the cervical spine caused neurological lesions. Hypoplasia of the vertebral artery was detected. Vertigo made

the dominant symptom that gradually increased. MRI of the brain, the cervical and the thoracic spine showed syringomyelia and lowered cerebellar tonsils. In agreement with this work, numerous authors confirm MRI to be the method of choice in diagnostics of syringomyelia and Chiari type I malformations (9–11). Posterior craniovertebral decompression and selective placement of a syringosubarachoid shunt in patients with Chiari I malformation and syringomyelia is an effective and safe treatment (11–14).

In our case, following the surgery the patient had no more vertigo and the neurological status significantly improved. Vertigo with Chiari I malformation and syringomyelia was successfully treated only surgically.

References

1. Aydin S, Hanimoglu H, Tanriverdi T, Yentur E, Kaynar MY. Chiari type I malformations in adults: a morphometric analysis of the posterior cranial fossa. Surg Neurol 2005; 64 (3): 237—241.

2. Di Lorenzo N, Cacciola F. Adult syringomielia. Clasification, pathogenesis and therapeutic approaclus. J Neurosurg Sci 2005; 49 (3): 65—72.

3. Fox B, Muzumdar D, DeMonte F. Resolution of tonsillar herniation and cervical syringomyelia following resection of a large petrous meningioma: case report and review of literature. Skull Base 2005; 15 (1): 89—97.

4. Dones J, De Jesus O, Colen CB, Toledo MM, Delgado M. Clinical outcomes in patients with Chiari I malformation: a review of 27 cases. Surg Neurol 2003; 60 (2): 142—147.

5. Bidzinski J, Michalik R. Clinical patterns of Arnold-Chiari malformations. Neurol Neurochir Pol 1998; 32 (5): 1181—1188.

6. Colak A, Boran BO, Kutlay M, Demirican N. A modified technique for syringo-subarachnoid shunt for treatment of syringomyelia. J CLin Neurosci 2005; 12 (6): 677—679.

7. Takigami I, Miyamoto K, Kodama H, Hosoe H, Tanimoto S, Shimizu K. Foramen magnum decompression for the treatment of Arnold Chiari malformation type I with associated syringomyelia in an elderly patient. Spinal Cord 2005; 43 (4): 249—251.

8. Liu B, Wang ZY, Xie JC, Han HB, Pei XL. Cerebrospinal fluid dynamics in Chiari malformation associated with syringomyelia. Chin Med J 2007; 120 (3): 219—223.

9. Plaza Mayor G, Baron Rubio M, Herraiz Puchol C, Lopez Lafuente J, Aparicio Fernandez JM. Neuro-otological manifestations as presentacion of type I Chiari malformation. An Otorrinolaringol Ibero Amer 2006; 33 (6): 613—622.

10. Alzate JC, Kathbauer KF, Jallo GI, Epstein FJ. Treatment of Chiari I malformation in patients with and without syringomyelia: a consecutive series of 66 vases. Neurosurg Focus 2001; 11 (1): E3.

11. Hida K, Iwasaki Y. Syringosubarachnoid shunt for syringomyelia associated with Chiari I malformation. Neurosurg Focus 2001; 11 (1): E7.

12. Liebenberg WA, Georges H, Demetriades AK, Hardwidge C. Does posterior fossa decompression improve oculomotor and vestibulo-ocular manifestations in Chiari 1 malformation? Acta Neurochir (Wiena) 2005; 147 (12): 1239—1240.

13. Colpan ME, Sekerci Z. Chiari type I malformation presenting as hemifacial spasm: case report. Neurosurgery 2005; 57 (2): E371.

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