Deformation of the cervicomedullary junction and spinal cord in a surgically treated adult Chiari I hindbrain hernia associated with syringomyelia: a magnetic resonance microscopic and neuropathological study

Case report

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The lower brainstem and cervical spinal cord from an ordinarily treated case of Chiari Type I hindbrain hernia associated with syringomyelia was examined using high-resolution magnetic resonance microscopy and standard neuropathological techniques. Magnetic resonance microscopy allows total screening and visualizes the disturbed internal and external microanatomy in the three orthogonal planes with the resolution of low-power optical microscopy. An additional advantage is the in situ visualization of the shunts. Afterwards the intact specimen is still available for microscopic examination.

Part of the deformation of the medulla is caused by chronic tonsillar compression and molding inside the foramen magnum. Other anomalies, such as atrophy caused by demyelination, elongation, and unusual disturbances at the level of the trigeminal and solitary nuclear complexes contribute to the deformation.

At the level of the syrinx-free upper part of the cervical cord, anomalies of the dorsal root and the dorsal horn are demonstrated.

KEY WORDS • Chiari I malformation • syringomyelia • magnetic resonance imaging
The present case is of particular interest because several anomalies were found, which is unusual in the classic descriptions of an adult patient with Chiari I malformation.

Case Report

This 60-year-old woman presented for the first time in 1980 complaining of pain in both arms and hands, especially on the left side. Her complaints had been progressive over a period of more than 10 years.

Examination. Neurological examination revealed sensory disturbances in the left side of the body; these were more pronounced in the shoulder and arm. Pain and thermal modalities, rather than tactile sensation, were heavily impaired in the left dermatomes C3–8. There were burn lesions of the left thumb and index finger complicated by osteomyelitis in the metacarpal bones. Proprioception was disturbed in the left arm and in both of the legs; it was most prominent in the left leg, resulting in an atactic gait. Tactile and pain sensations were diminished in the left side of the face with a nearly abolished left-sided corneal reflex. There was a weakness in the left leg without pyramidal signs. The patient had a minimal scoliosis. There were no cutaneous signs of an underlying neural closure defect. Ophthalmological examination revealed a horizontal nystagmus and an abducent paresis on the right side (perhaps a paralytic eye from birth). Somatosensory evoked potentials were disturbed on the left side, indicating a lesion in the cervical cord or located more cephalad. An MR image showed a Chiari I malformation with a syringomyelia from C3–T11 (Fig. 1 left). The largest cyst was situated in the cervicothoracic region. The conus medullaris had a normal position and there was no myelodysplasia.

First Operation. In 1987 the patient underwent a posterior fossa decompression and a syringoarachnoidal shunt was placed at the level of C-4 (Fig. 1 right).

First Postoperative Course. The patient’s postoperative MR image showed a rounded ectopic tonsil and partial collapse of the syringes, indicating a satisfactory cranio-cervical decompression. However, no substantial recovery was noticed. The patient’s pain worsened and the ataxia slowly progressed.

Second Operation. In a second operation performed in 1989 a lumboperitoneal shunt was placed, followed by a syringopleural shunt with a T-drain at the level of C5–6.

Second Postoperative Course. The cysts never completely collapsed. The patient died unexpectedly in 1991, a few weeks after she left the hospital where she had been treated for panaritium followed by general sepsis.

Autopsy Findings

At autopsy the spinal cord and brainstem were separated from the brain, which appeared normal. An ectopia of the right tonsil stretching upward to the lower margin of C-1 was noticed. The position of the left tonsil was normal. The cerebellum, which appeared macroscopically to be normal, was removed from the brainstem; the medulla oblongata and the overlying right tonsil were then separated from the pons. The spinal cord showed some deformation on the dorsal side and scarring of the pia-arachnoid on the side of operation. More particularly, the spinal cord did not show signs of neural closure defects. The lumbo-peritoneal shunt was removed. The other shunts were left inside the spinal cord. General autopsy revealed a pulmonary embolism as the cause of death.
Microanatomy of Chiari I malformation

High-Field MR Microscopy Imaging and Neuropathological Control

The medulla oblongata was separated from the spinal cord, which was cut into 3-cm pieces. The cervical spinal cord and the cervicothoracic transition were scanned without prior specimen fixation. The thoracic cord and the medulla oblongata were fixed for 2 weeks in 10% formalin before MR investigation was performed.

Inherent to MR imaging, sections could be made in the three orthogonal planes. For the transverse images, 2-mm slices were taken with a 17-× 17-mm field of view and a 350 × 350 data matrix, corresponding to an inplane resolution of 40 × 40 μ. For the longitudinal views 1-mm slices were selected with a 38-× 17-mm field of view and a 700 × 320 data acquisition matrix. Acquisition parameters of 2500 msec repetition time and 18 msec echo time were used to produce essentially spin-density images; 32 excitations were made for the transverse and 16 for the longitudinal images, resulting in an acquisition time of 4 to 8 hours for the collection of slices in each direction. The limiting probe dimensions were 23 mm in diameter and 40 mm (the height of the coil homogeneity) in height.

The MR microscopy images of the pathological specimen were systematically compared with reference images of normal specimens. All MR findings were correlated with data obtained by neuropathological examinations. Serial sections of the entire spinal cord and the lower brainstem were routinely examined by light microscopy using standard histological stains such as the Klüver–Barrera and trichrome Masson.

Results

Medulla Oblongata

In comparison with MR microscopy images of the normal specimen, the longitudinal MR sections showed a mild elongation of the medulla oblongata. Because the clinical MR image obtained during life showed a normal positioning of the pontomedullary transition, the elongation was restricted to the medulla. The length of the olive was increased approximately 3.5 mm (Fig. 2). A normal value for the length of this nucleus on MR imaging is approximately 14 mm. The right side was situated in a ventrodorsal direction, asymmetrically flattened by the descended right tonsil, with its diameter decreased to approximately two-thirds of normal size. The left half of the medulla had an overall reduced size. The olivary nucleus was flattened in a ventrodorsal direction and appeared slightly elongated in longitudinal sections. The cell rows appeared normal on the MR microscopy sections (Fig. 3 upper). Correlated neuropathology sections, however, showed a very mild neuronal cell loss (Fig. 3 lower). The accessory olivary nuclei were also flattened. The descended right cerebellar tonsil, which appeared pointed on the preoperative images, had a normally rounded shape on the MR microscopy sections, which is mostly seen on postoperative imaging after suboccipital craniectomy.2,4 The tonsil did not show any changes in signal intensity. Neuropathological examination showed cellular loss in its cortical layers.

Axial MR microscopy images obtained from the right side showed the nuclei gracilis, cuneatus, and cuneatus accessorius as they are normally depicted. However, their rostral boundaries could not properly be delineated. On the left side, the nucleus cuneatus and gracilis were hypoplastic and had an increased signal intensity. The left nucleus cuneatus accessorius could not be identified. The fibrae arcuatae internae of the left side, which form the right medial lemniscus, were absent. The right medial lemniscus was hypoplastic and exhibited an increased signal intensity. On the left side, the medial lemniscus had a
normal size and signal intensity. Histopathological examination revealed that the increased signal intensity and hypoplasia of the left medial lemniscus corresponded to important fiber loss and gliosis or demyelination. Neuropathological differentiation between gliosis and demyelination could not be made. Neuropathological investigation also confirmed the hypotrophy of the nuclei cuneatus and gracilis and the absence of the fibrae arcuatae internae and nucleus cuneatus accessorius. In the lower parts of the medulla, the right pyramidal tract was smaller than the left, exhibiting a slightly increased signal intensity that corresponded neuropathologically with demyelination of the corticospinal tract. This corresponded clinically with the patient’s weakness in the left arm and leg during life. The medial longitudinal fascicle appeared normal at any level.

The MR microscopy images also showed a disturbance of the internal microanatomy of cranial nerve nuclei inside the left half of the medulla. The trigeminal complex was hypoplastic, whereas its laminar arrangement could not be recognized. The entire complex was dorsally displaced. The left nucleus of the solitary complex was absent although its tract could be identified. In the central gray matter, left and right hypoglossal nuclei could be identified bilaterally by their lower signal intensity. The nucleus ambiguus was normal. Neuropathological investigation confirmed all these features and revealed no addition information. On the compressed right side the anatomical relationships were well preserved.

**Syringomyelic Spinal Cord**

The most prominent abnormal feature seen on the MR microscopic images was the presence of large, well-demarcated areas of high signal intensity in the central gray matter of the spinal cord from C-4 down into the lower thoracic regions (Fig. 4). These areas corresponded to the syringes. At some levels the syringes were deformed by collapse and were not bordered by a cell layer. At the cervical level the cysts extended into the left posterior horn up to the pia mater. In the MR microscopy sections, as well as in the corresponding neuropathology sections, no communication with the subarachnoid space could be observed.

The central canal could be identified from the obex down to the upper end of the most rostral cyst corresponding with root level C-4 in the transverse as well as in the longitudinal MR images. The high signal intensity of the lumen of the canal indicated (microscopically) a fluid-filled open canal. The surrounding layer of low signal intensity correlated with a histopathologically normal ependymal layer. From the level of C-7 downward the central canal remained ventrally separated from the posterior horn up to the pia mater. In the MR microscopy images, as well as in the corresponding neuropathology sections, no communication with the subarachnoid space could be observed.

**Fig. 4.** Axial magnetic resonance microscopy image of a section through the cervical cord at the C4–5 level showing the partially collapsed syrinx (S) extending into the gray matter of the left posterior horn (arrows).

**Fig. 5.** Upper: Magnetic resonance (MR) microscopy image of a section through the syrinx-free C-2 segment. The structures of the left posterior horn are hypoplastic despite the absence of a syrinx. The right dorsal root and the dorsal root entry zone (arrows) have an abnormal size and course, due to a congenital abnormality. Lower: Photomicrograph of a neuropathology section corresponding to the MR microscopy image above, showing, apart from some remnants of the substantia gelatinosa, the absence of posterior horn structures. The abnormal dorsal root as well as the abnormal dorsal root entry zone (arrows) correspond to the MR microscopy images. Klüver–Barrera stain, original magnification × 100.
syrinx; the ependymal layer then widely opened into a syrinx. A remnant of the ependymal layer was present in the ventral part of the cavity. From this point upward the syringes actually presented as cavities extending in a craniocaudal direction inside the gray matter, which was parallel with the central canal. The leptomeninges were not fibrotic around the cord or at the craniocervical junction.

The dorsal part of the cord was heavily deformed due in part to the collapse of the syringes and in part to the deformation and hypotrophy of the dorsal white matter. The left cuneate and both gracile fascicles exhibited an increased signal intensity due to demyelination or gliosis. These findings were confirmed by neuropathological investigations, which cannot differentiate between demyelination and gliosis. An area of high signal intensity, containing the fasciculus gracilis and fasciculus cuneatus, bordered the dorsolateral extension of the syringes. The posterior horn and dorsal root entry zone could no longer be identified. The correlating histopathological sections showed only some remnants of the substantia gelatinosa. The entering dorsal roots were thin and hypoplastic. The upper portion of the cervical cord up to the C2–3 root level was syrinx free. However, we found identical abnormalities of the posterior horn and dorsal root entry zones and these were on the same side (Fig. 5).

On the right side, the dorsal roots shown entering the dorsal root entry zones appeared hypertrophic. These roots had an increased amount of large-caliber myelinated fiber bundles entering the cord and spreading fanwise into the right dorsal white columns before disappearing inside the base of the ventral horn. The corresponding sagittal MR microscopy sections showed an abnormal arrangement of clustered dorsal roots entering the cord between the dorsolateral sulcus and the top of the posterior horn.

Inside the gliotic deformed areas of the dorsal white matter tracts, chaotic networks of densely packed abnormal microvessels were also seen surrounding the syringal cavities. The walls of these vessels appeared irregular and thickened by an accumulation of abnormal amounts of collagen fibers inside the adventitia, when viewed with additional collagen staining. This hypervascularity with abnormal intrinsic microvessels was also seen in the corresponding areas of the syrinx-free upper segments.

In the MR microscopy sections the shunts inside the syringes were visualized (Fig. 6). At the C4–5 root level, a syringoperitoneal shunt was seen lying properly inside the left dorsolateral extent of the syrinx. At the lower C7–8 level the syringopleural T-drain was seen entering the cord at the left dorsolateral area lying inside the syrinx, whereas the distal and proximal intramedullary arms of the drain perforated the cyst wall and penetrated into the ventral horn of the right side. In the area of penetration, multiple spots of low signal intensity were seen surrounding the drain. These spots were remnants of blood. At the surface of the cord the shunt insertion sides were covered and surrounded by dense arachnoidal scarring.

**Discussion**

*Magnetic Resonance Neuropathology*

Magnetic resonance microscopy fills the visualization “gap” between the operative microscope and the high-
power optical microscope; in addition it provides information on the microscopic level about poorly understood MR characteristics exhibited during the patient's lifetime. In clinical MR imaging, longitudinally extending fluid cavities are often surrounded by areas of high signal intensity,\(^5,10\) which may extend beyond the immediate neighborhood of the cavities. These areas have been misdiagnosed on clinical MR images as tumor, leading to biopsy procedures.\(^2,3,9\) It is presumed that they correspond to zones of demyelination or gliosis.\(^10\) The exact neuropathological correlation is not known and myelomalacia has also been mentioned.\(^10\) Magnetic resonance microscopy imaging at the level of the cysts localizes the areas partly in the remnants of parenchyma surrounding the cysts and partly in the dorsal white matter tracts. Neuropathological correlation reveals gliosis in the parenchyma surrounding the cysts and in the adjacent dorsal white matter tracts. Serial consecutive slices trace the changes in white-matter tracts up to the nuclei gracilis and cuneatus. This explains how wallerian degeneration of long white tract areas of high signal intensity can be seen on clinical MR images, even rostral to the upper syrinx in the syrinx-free segments of the cervical spinal cord.\(^2,10,39\)

The 1- or 2-mm MR microscopy slices enlarge the spatial views in a section, in contrast to the 6-\(\mu\)m standard neuropathology slices, and produce an unfamiliar view of the abnormal microvessels inside the pathological tissue surrounding the syringoeal cavities. These vessels have been described before.\(^1,2,8,29,38,45\) Their appearance in MR microscopy images is comparable to their appearance on postmortem microangiography of the spinal cord in post-traumatic syringomyelia.\(^40\) The microvessels are not only seen in the gliotic remnants of the parenchyma surrounding the cavities\(^1,45\) but are also present in the entire area of the changed gliotic dorsal white matter.\(^29,38\) Based on the presence of these vessels with hypertrophied adventitia, an underlying vascular malformation has already been assumed by some authors.\(^2,7,29\) Although the origin of these vessels and their relation to the underlying pathological tissue is not understood, their widened perivascular spaces should play an important role in the passage of cerebrospinal fluid from the subarachnoidal space into the spinal cord. This should lead to the formation of cavities.\(^1,3,11,47\)

An illustrative example of the advantages of sagittal MR microscopy imaging sections is the direct visual demonstration of the elongation of the medulla oblongata and its olivary nuclei. These nuclei show no changes in signal intensity and their corresponding neuropathological findings are normal. This finding can be related to the discussion of the caudal descent of the brainstem below the foramen magnum in Chiari I malformation cases. This descent has often been described, although for the most part it has been poorly documented, even in surgical descriptions.\(^1,2,8,21,33\) Caudal displacement of the medulla with a normally positioned pons is difficult to demonstrate on clinical MR imaging as well as in postmortem examinations.

Another advantage of MR microscopy lies in the visualization of draining catheters inside the syringes in the excised spinal cord. Correct assessment of the position of the shunt inside the spinal cord is very difficult even postmortem. In this case, there was damage to the spinal cord by the T-arm of a syringopleural shunt; the T-arm had penetrated into the ventral horn. Remnants of an old hemorrhage surrounding the shunt suggest a traumatic insertion. Postoperative rotational movements of the shunt inside the syrinx have previously been reported to cause damage to the spinal cord.\(^46\)

### Deformation of the Medulla Oblongata

In contrast to the Chiari II malformation, severe brainstem syndrome occurs infrequently in Chiari I malformation, especially in adult cases.\(^6,7,23,33,34\) Our patient only exhibited sensory deficits on the left side of the face, which frequently occur in Chiari I patients,\(^2,12,42\) and motor weakness in the left leg as signs of brainstem involvement. Nevertheless, the lower brainstem was heavily deformed. This deformation was caused by compression and molding inside the foramen magnum due to single tonsill herniation caused by elongation, by hypotrophy of the lemniscal system, and by anomalies inside some cranial nerve nuclei.

Elongation of the medulla with symmetrical elongation of both olivary nuclei without neuronal cellular loss contributes considerably to the deformity of the medulla. Its origin is unknown, although chronic compression, probably occurring since birth, is likely to cause tissue changes. This elongation differs from the caudal displacement of the entire malformed brainstem, fourth ventricle, and parts of the cerebellum, which is seen in Chiari II cases.\(^6,30,37\)

The unilateral hypotrophy of the entire lemniscal system, caused by transynaptic degeneration, results in deformation of the internal microanatomy. The deformation of the medial lemniscus is not caused by syringobulbia or clefts inside the medulla, which has often been reported.\(^1,4,22,39,43\) The deformed area in the hypotrophic nucleus gracilis and nucleus cuneatus differs from the impressive deformity in the same area shown in Chiari II malformation caused by the overhanging of this dorsal part of the descended medulla over the spinal cord, which is kept in place by the dentate ligaments.\(^2,4,5,7\)

Anomalies of cranial nerve nuclei in Chiari I malformation have only been microscopically described in the presence of syringobulbia of clefts.\(^1,4,7\) In Chiari II malformation they occur more frequently and are thought to be caused by maldevelopment.\(^1,3,14,21,28,41,47,50\) Nuclear anomalies inside the trigeminal and the solitary complexes, as demonstrated in this case, however, are unusual in Chiari I as well as in Chiari II malformation. No similar descriptions have been found in the literature. The origin remains uncertain and might be complex. The effects of chronic tonsillar compression or of molding inside the foramen magnum are difficult to assess, whereas mechanical forces such as distortion due to operative decompression are likely to play a role as well.

### Associated Syringomyelia

The serial MR and neuropathology sections of the spinal cord display the characteristic features of a hindbrain hermia-related syringomyelia. The central canal is microscopically open\(^1,3,12,28,31,35,46\) but not necessarily patent.\(^2,28\) It opens widely into a syrinx cavity.\(^3,49\) The syringes extend outwardly into the posterolateral gray matter\(^2,8,14,17,18,21,29,38,39,45,49\) as well as cranio-caudally independently.
Microanatomy of Chiari I malformation

from the central canal. At the level of posterolateral cyst extensions, the posterior horn structures have disappeared. Cerebrospinal fluid pulsations dissecting the gray matter are generally accepted mechanisms for syrinx extension into the posterior gray matter. A syrinx-free cervical segment between the fourth ventricle and the most rostral syrinx is also a common finding.

The finding that in this segment on the same side of the syrinx extensions the posterior horn structures have disappeared and are replaced by gliosis with an abundance of abnormal microvessels, however, is unusual. These gliotic changes are not related to a syrinx and their cause is not clear. Preexisting lesions of different origins have been the subject of many studies of syringomyelia. Changes due to multiple operations might have contributed to disturbing the microanatomy in this case.

Another unusual finding is the disarrangement of the entry zone and the course of the dorsal roots at the same level on the opposite side. Its cause also remains unclear.

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