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Shoulder pain as one of the symptoms of syringomyelia

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ABSTRACT

Syringomyelia is typically a progressive chronic condition caused by a disruption of normal cerebrospinal fluid flow. Earlier diagnosis is associated with better outcomes, because although the progression of neurological deficits usually stabilizes after intervention, many patients still remain at least partially symptomatic.

In this paper, we describe the case of a 33-year-old female patient with syringomyelia. The patient reported to the Department of Neurology due to sensory disorders and shoulder pain for several months. On admission, neurological examination revealed right hand muscular deficit, abolition of sensation of temperature and asymmetry of tendon reflexes. A few weeks earlier, outpatient magnetic resonance imaging (MRI) of the cervical spine showed the features of Arnold-Chiari malformation, syringomyelia and C5/C6 and C6/C7 discopathy. During hospitalization, MRI of the thoracic spine was performed, and the syringomyelic cavity C2–Th6 was revealed. The patient in stable condition was discharged home and referred to the neurosurgery department. The patient underwent a medial sub-occipital craniectomy and, a

month later, was admitted to the neurological rehabilitation department due to paresis and sensory disturbances of right upper limb. As a result of the physiotherapy, the right arm's motor function and general physical condition improved.

Keywords: syringomyelia, Arnold-Chiari malformation

INTRODUCTION

Syringomyelia is an etiologically diverse disease caused by a disruption of normal dynamics of cerebrospinal fluid flow [1]. The term “syringomyelia” literally means “cavity within the spinal cord” [2]. The disease is typically a progressive chronic condition with exacerbations and remissions. Although the progression of neurological deficits usually stabilizes after intervention, despite improvement, patients often still have disease symptoms, but less severe [1, 2].

Most lesions are located within the spinal cord between C2 and Th9, but they can go down to the conus medullaris or extend up to the brainstem. The prevalence in western countries is estimated about 8.4 per 100,000 persons [3, 4].

Acquired syringomyelia is considered to be caused by impaired circulation and dynamics of cerebrospinal fluid (CSF). In the case of Chiari-I malformations, this disorder is caused by the downward herniation of the cerebellar tonsils, obstructing the normal flow of CSF from the cranial to spinal compartment during a normal heart cycle, coughing or laughing. New syrinx formation or enlargement of already existing syrinx in a child with spinal cord involvement may also be due to the accompanying Chiari-II malformation, also causing impediment to normal CSF flow dynamics, as with Chiari-I malformations [1].

Most syringes are present from birth, and others grow during life. Other congenital anomalies of the spine, which cause narrowing of the spinal canal, can also lead to cerebrospinal fluid flow disorders with resulting syringomyelia [5].

If a syrinx is asymptomatic, it is only discovered on incidental imaging of the spinal cord. In symptomatic patients, bilateral motor and sensory symptoms and signs that do not involve the head or face can be present as in any other spinal cord lesion. There is often a loss of the ability to feel hot or cold, especially in the hands. The severity and duration of the symptoms often change, and each patient experiences a different combination of these symptoms, usually

varying depending on the location of syrinx in the spinal cord. MRI can easily identify the extent and location of any intramedullary cavity [1].

In patients with asymptomatic syringomyelia, observation seems justified, although sometimes it can be quite difficult, especially in cases where large syrinx extends to the brainstem. However, it should be noticed that spontaneous regression of syringomyelia has been observed, although rarely. Non-surgical treatment of symptomatic syringomyelia consists in the treatment of pain and paresthesia with the help of painkillers, antidepressants, antiepileptics, or in the maintenance of functional abilities by rehabilitation and physical medicine [1].

In this paper, we analyse the case of a patient diagnosed with Arnold-Chiari malformation, syringomyelia and discopathy.

CASE REPORT

A 33-year-old patient was admitted to the Department of Neurology due to sensory disorders occurring for several months (mainly sensation of temperature on the right forearm and paresthesia in the fingers and toes) and shoulder pain. On admission, neurological examination revealed a discrete deficiency of right hand strength, abolition of sensation of temperature, disturbed sensation of touch on the upper right limb and asymmetry of tendon reflexes (right <left). The results of laboratory tests at the time of admission to the department are presented in Table 1. A few weeks earlier, outpatient magnetic resonance imaging (MRI) of the cervical spine with assessment of the cranio-vertebral border showed the features of Arnold-Chiari malformation, syringomyelia and C5/C6 and C6/C7 discopathy (Figure 1 and Table 2). During hospitalization, MRI of the thoracic spine was performed, and the syringomyelic cavity C2 to Th6 was revealed (Figures 2, 3 and Table 2). The patient was consulted neurosurgically and qualified for surgery. The patient in stable condition was discharged home and referred to the neurosurgery department.

In the neurosurgery department, the patient underwent a medial sub-occipital craniectomy. A month after neurosurgical operation, the patient was admitted to the neurological rehabilitation department due to paresis and sensory disturbances of right upper limb. As a result of the physiotherapy used, the right shoulder pain subsided, the right arm's motor function and general physical condition improved (Table 3). Rehabilitation under the direction of a general practitioner and continuation of physical exercises were recommended.

Table 1. The results of laboratory tests at the time of admission to the department

Parameter	**Value**
ALT [U/L]	25,5
AST [U/L]	22,2
Total bilirubin [μmol/l]	10,5
CK-MB [ng/ml]	1,36
CRP quantitatively [mg/l]	0,54
APTT [sec]	31,7
eGFR [ml/min/1,73 m^2]	107,56
GGTP [U/L]	15
Glucose [mg/dl]	92,4
Serum creatinine [μmol/l]	59
K^+ [mmol/l]	3,66
Na^+ [mmol/l]	143
Troponin T [ng/ml]	0,005
PT [sec]	12,7
INR	0,96
HCT [%]	40
RBC [$x10^6$/μL]	4,8
HGB [g/dl]	14,6
WBC [$x10^3$/μL]	6,7
MCV [fL]	83,3
MCH [pg]	30,4
PLT [$x10^3$/μL]	321

Table 2. Descriptions of imaging studies and electroneuronography

Diagnostic examination	**Results**
MRI of the cervical spine – before hospitalization (Figure 1)	The cerebellar tonsils were pointed: right approx. 2 mm below, left 4 mm below the level of the foramen magnum. In the spinal cord from the level of the C2 to the Th2 (in the visible range) were revealed areas with fluid signal, located in the upper segment, somewhat eccentrically – more on the right side, in the thoracic segment of the spine occupying the central part of the spinal column. Dimension in the axial plane of approx. 9 x 6 mm – most likely syringomyelic cavities. The image probably corresponds to advanced syringomyelia (contrast-enhanced imaging was indicated). Spinal cord dilatation in the visible range of the examination. Elimination of cervical lordosis with the formation of small kyphosis in the C3-C5 segment. The tendency to frontal slide at C4-C5. Degenerative changes in the form of slight partial dehydration of the C5-C6, C6-C7 intervertebral discs. Posterior wide-base herniate of the intervertebral disc C4-C5, C5-C6, C6-C7 with a decrease in the anterior volume reserve of the spinal canal – the most severe changes at C5-C6. Signal of the bone marrow of cervical spine components was correctly. Conclusions: Features of Arnold-Chiari malformation in the form of the pointed shape of the cerebellar tonsils and low position of the cerebellar tonsils and syringomyelia (for

	checking after contrast-enhanced imaging). Increase in spinal cord volume. Discopathy of the intervertebral discs C5-C6, C6-C7. Abolition of cervical lordosis and presence of small kyphosis.

MRI of the thoracic spine – during hospitalization (Figures 2, 3)	Within the upper part of the thoracic segment, the syringomyelic cavity is visible up to the C2 region of the cervical segment. Lower cavity limit at Th6 level. Slightly below a small widening of the spinal cavity to 2 mm (Th7-Th8). After administration of the contrast agent, no enhancement was seen within the described changes. Focus of steatosis in the Th12 vertebral body. A small amount of fluid in the pleural cavities. Other illustrated structures with no obvious abnormalities. No other changes were visible. Conclusions: syringomyelic cavity in the area described above.
Electroneuronography (ENG)	Normal motor, sensory and F wave conduction in the median nerves. Normal conduction in the peroneal and sural nerves. Quite significant extension of the average F wave latency in peroneal nerves; moderate reduction in the frequency of this wave in the right nerve. Conclusion: electrophysiological features of the L-S root syndrome right> left, without other changes.

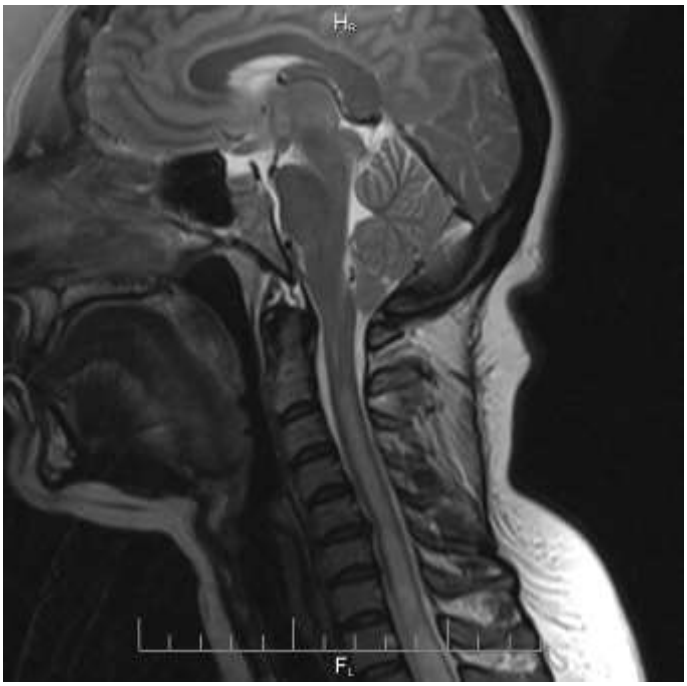

Figure 1. In the spinal cord from the level of the C2 to the Th2 (in the visible range) were revealed areas with fluid signal, located in the upper segment, somewhat eccentrically – more on the right side, in the thoracic segment of the spine occupying the central part of the spinal column; dimension in the axial plane of approx. 9 x 6 mm – syringomyelic cavities [authors' material]

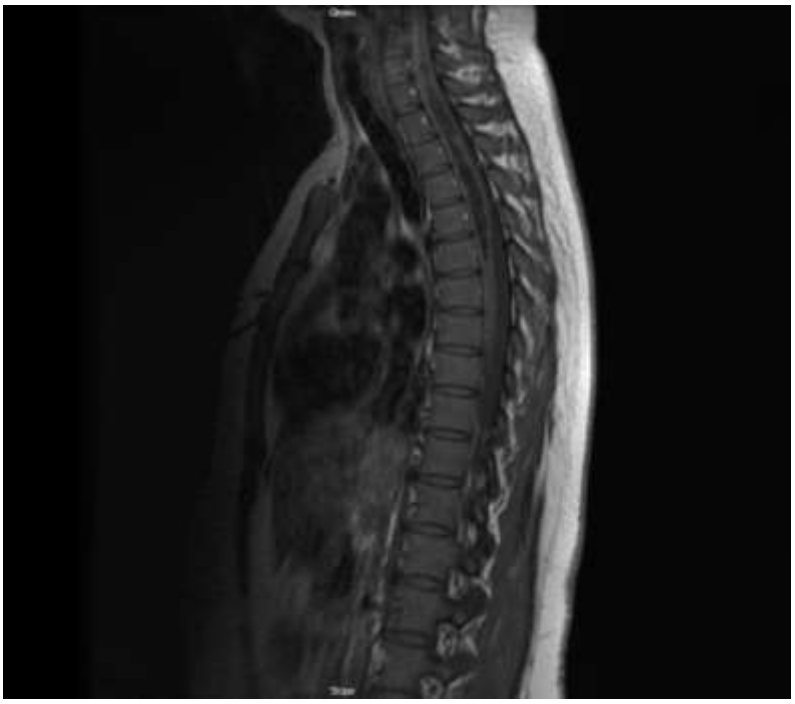

Figure 2. MRI of the thoracic spine (during hospitalization) – within the upper part of the thoracic segment, the syringomyelic cavity is visible up to the C2 region of the cervical segment; lower cavity limit at Th6 level [authors' material]

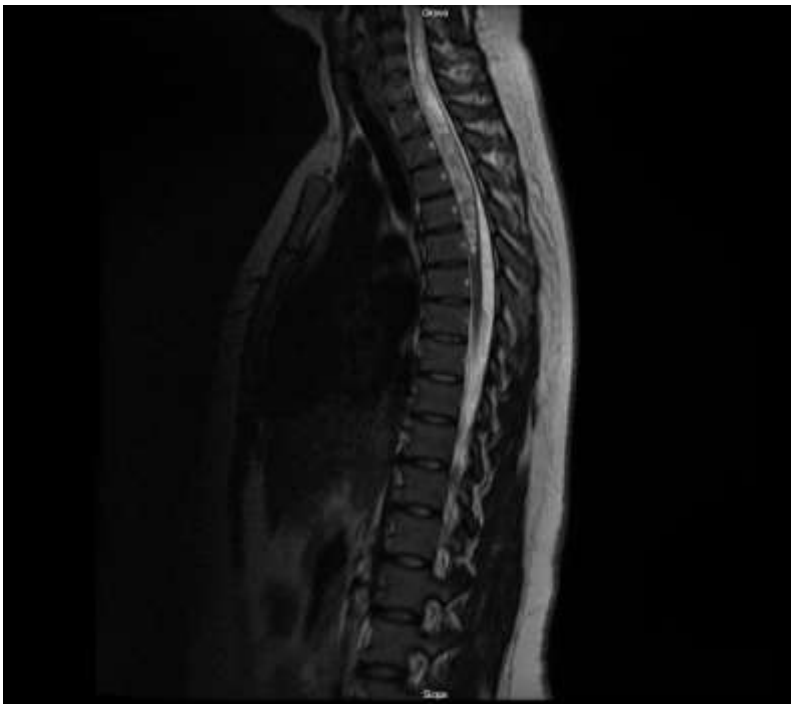

Figure 3. MRI of the thoracic spine (during hospitalization) – within the upper part of the thoracic segment, the syringomyelic cavity is visible up to the C2 region of the cervical segment; lower cavity limit at Th6 level [authors' material]

Table 3. Comparison of the patient's condition on admission to the neurological rehabilitation department and during discharge home

Type of scale	**On admission to the neurological rehabilitation department**	**Discharge home**
Right shoulder pain – Numeric Rating Scale (NRS)	4	0
Barthel scale	19 points	20 points
Rankin scale	3 points	1 point

DISCUSSION

It is crucial to first identify the etiology when assessing a patient with syringomyelia, because the therapeutic strategy will be based on causal changes rather than on syrinx itself. This means that hydrocephalus or shunt malfunction should be excluded by performing an MRI of the head and the tethered cord should be excluded by MRI examination of the lumbosacral spine. In the absence of a cause, there are not many alternatives. Nowadays, the widespread availability of MRI results in many accidental discoveries of the intramedullary cavity, and in a large proportion of these patients, a causal change or mechanism cannot be found, even using all available radiological methods [1, 6, 7].

The central canal of the spinal cord is present at birth and usually becomes progressively obliterated, but it can also persist To be able to diagnose a "persistent central canal", there should be no factors that would predispose to syringomyelia, and the syrinx should be located at the junction of the ventral one-third and dorsal two-third of the spinal cord [1].

Chiari malformation is known to be associated with syringomyelia in many patients [8-14]. Among the patients with Chiari malformation who underwent surgery, syringomyelia was found in 60% to 85% of these patients [15-17]. Many surgeons believe that the presence of syrinx may be an indication for surgery; however, these reports probably overestimate the incidence of syrinx in patients with Chiari malformation [18-20].

In the research by Strahle et al. [21], the majority of patients had undergone MRI of the cervical spine to screen for syringomyelia, half of them had undergone imaging of the whole spine – 22.9% of patients with Chiari malformation also had a syringomyelia.

In the case of shoulder pain, extensive differential diagnosis should be considered, which may be supported by the location of the patient's pain. Examples of diseases that should be included depending on the location of the pain are listed in Table 4 [22].

Table 4. Examples of diseases that should be included in the differential diagnosis depending on the location of shoulder pain [22]

Location of shoulder pain	**Diseases for differential diagnosis**
Lateral shoulder pain	cervical radiculopathy, subacromial impingement syndrome, rotator cuff tendonitis, adhesive capsulitis, subacromial bursitis, full-thickness or partial-thickness tears of the rotator cuff tendons, multidirectional instability, proximal humeral fracture
Anterior shoulder pain	rotator cuff disease, glenohumeral osteoarthritis, acromioclavicular arthritis, acromioclavicular separation, biceps tendonitis, adhesive capsulitis, anterior instability, proximal humeral fracture
Posterior shoulder pain	posterior instability/dislocation, cervical radiculopathy, suprascapular nerve entrapment, rotator cuff disease, labral tear, glenohumeral osteoarthritis, proximal humeral fracture

Nonspecific shoulder pain	• Cervical radiculopathy: pain below the elbow, numbness or weakness, decreased reflexes • Polymyalgia rheumatica: older patient, bilateral shoulder pain, full range of motion, no weakness, claudication, fatigue • Glenohumeral osteoarthritis • Rheumatoid arthritis: stiffness and other joint involvement • Consider pulmonary, gastrointestinal, and cardiac causes of diaphragm irritation or referred pain

CONCLUSIONS

Syringomyelia can be found in imaging studies in symptomatic patients as well as accidentally in asymptomatic patients. Shoulder pain and disturbances in sensation of temperature occurring in the described patient required extensive differential diagnosis, taking into account primarily orthopedic diseases. Neurological deficits usually stabilize after surgery, although some symptoms will still be present.

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