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*Anna Mosiewicz[1], Radosław Rola[1], Barbara Mosiewicz-Madejska[2], Robert Kaczmarczyk[1], Tomasz Trojanowski[1]

# Intramedullary cavernomas – an own experience and a review of the literature

## Naczyniaki jamiste rdzenia kręgowego – doświadczenie własne i przegląd literatury

[1]Chair and Department of Neurosurgery and Paediatric Neurosurgery, Medical University in Lublin
Head of Department: Professor Tomasz Trojanowski, MD, PhD
[2]Chair and Department of Neurosurgery and Paediatric Neurosurgery, Students Medical Association, Medical University in Lublin
Protector of Association: Anna Mosiewicz, MD, PhD

**Keywords**

cavernoma, spinal cord, vascular malformation

**Słowa kluczowe**

naczyniak jamisty, rdzeń kręgowy, malformacja naczyniowa

Summary

**Introduction.** Cavernous malformations constitute relatively rare vascular malformations of the centralnervous system (CNS). However, recently they are being diagnosed more often thanks to theintroduction of nuclear magnetic resonance.

**Aim.** The aim of this work was to analyse course of the disease of patients treated in our Department and to review literature on this subject.

**Material and methods.** Over the past ten years only three patients with spinal cavernomas were treated in the Department of Neurosurgery of the Medical University in Lublin. Following these cases we decided to review literature on symptomatology, clinicalcourse, diagnostics and treatment of intramedullary cavernomas.

**Results.** Being more prevalent in the brain, cavernomas hardly ever occur in the spinal cord. The most frequent location of cavernomas of the spinal cord is its thoracic part. Spinal cavernomas are difficult to diagnose due to their low incidence and nonspecific clinical course. Clinical manifestation of the disease varies from discrete neurological symptoms to rapid progression of motor and sensory deficits. However, the most common course of the disease is slow progression of neurological deficits due tomicrobleedings from the cavernoma.

**Conclusions.** The only effective way of treatment and prevention from further neurological deficits is surgery.

Streszczenie

**Wstęp.** Naczyniaki jamiste są stosunkowo rzadkimi zmianami w centralnym układzie nerwowym. Jednakże ostatnio są one diagnozowane coraz częściej dzięki wprowadzeniu do diagnostyki rezonansu magnetycznego.

**Cel pracy.** Celem pracy była analiza przebiegu choroby u pacjentów leczonych w naszej Klinice oraz przegląd literatury na ten temat.

**Materiał i metody.** W ciągu ostatnich 10 lat w Klinice Neurochirurgii Uniwersytetu Medycznego w Lublinie było leczonych tylko 3 pacjentów z naczyniakiem jamistym rdzenia. Opisy tych przypadków stały się przyczynkiem do analizy opisywanych w literaturze objawów, obrazu klinicznego, procesu diagnostycznego i leczenia naczyniaków jamistych rdzenia.

**Wyniki.** Naczyniaki jamiste częściej występują w mózgu, w rdzeniu kręgowym są one zjawiskiem niezwykle rzadkim. Najczęstszą lokalizacją naczyniaków jamistych rdzenia jest jego odcinek piersiowy. Naczyniaki jamiste rdzenia są trudne do zdiagnozowania ze względu na ich rzadkie występowanie oraz mało charakterystyczny obraz kliniczny. Spektrum manifestacji klinicznych tej choroby rozciąga się od subtelnych objawów neurologicznych do gwałtownego wystąpienia ubytków czuciowych i ruchowych. Jednakże najczęstszym obrazem choroby jest powolna progresja ubytków neurologicznych związana z mikrokrwawieniami z naczyniaka.

**Wnioski.** Jedyną skuteczną metodą terapii i zapobiegania dalszemu postępowi deficytów neurologicznych jest leczenie chirurgiczne.

**Conflict of interest**
**Konflikt interesów**

None
Brak konfliktu interesów

**Address/adres:**

*Anna Mosiewicz
Katedra i Klinika Neurochirurgii
i Neurochirurgii Dziecięcej
Uniwersytet Medyczny w Lublinie
ul. Jaczewskiego 8, 20-954 Lublin
tel./fax +48 (81) 724-45-28
mosiewiczanna@gmail.com

## INTRODUCTION

Cavernous malformations are rare vascular malformations in the nervous system, mostly found in the hemispheric locations. Nonetheless, these malformations have been described within the cerebellum, cranial nerves, spinal roots, brain stem and spinal cord as well (1). It has been estimated that cavernous malformations constitute 5-12% of all vascular malformations within CNS (2, 3) and 3-5% of them are located in the spinal cord (3-5).

Pathogenesis of cavernomas remains unclear, however most of the existing theories suggest their developmental character (2, 4). Some imply that an angioblastic mesoderm dysplasia might occur during embryogenesis (2). Interestingly, they often coexist with developmental venous anomalies. It has been suggested that venous malformations play some role in the development of cavernous malformations.

Familiar occurrence as well as multiple localizations within the brain and spinal cord have been described (5-7). Candidate genes responsible for the development of cavernomas were identified in cases with familiar occurrence (4). Reports also exist on "de novo" generation of cavernomas following radiotherapy for other spinal lesions (8).

First case of intramedullary cavernoma was described by Hadlich in 1903 based on post mortem examination (9). Schultze performed the first successful surgery for intramedullary cavernoma in 1912 (9). Only 19 cases were described in the literature up to the mid-eighties of the last century (4, 9). It was most likely related to the efficacy of the available diagnostic tools i.e. myelography that had not been able to detect intramedullary lesions like small cavernous malformations. Spinal angiography had not been able to detect these lesions either. Detectability of cavernomas both, in the spine as well as in the brain (in particular those with previous hemorrhage) increased significantly with introduction of diagnostic MRI (4, 6, 9). Zevgaridis et al. (9) performed the largest to date analysis of 117 intramedullary cavernomas published in the literature between 1903 and 1996 and, based on their personal experience, described symptomatology, diagnosis and outcome of intramedullary cavernoma treatment.

## OWN SERIES

### 1 SG

32-year-old male suffered from slow, progressive paraparesis of the lower extremities over a period of more than 12 years prior to the Department admission. Initially symptoms involved left, but subsequently involved right leg as well. He also reported transient difficulties with miction initiation that worsened over last few months prior to admission. Persistent neck pain, radiating to the left shoulder with concomitant numbness of the upper extremities was also reported. Paraparesis of the lower extremities, particularly the right one, has aggravated over the last year – patient was barely able to ambulate with elbow crouches.

Neurological examination revealed severe, spastic paraparesis of the lower extremities with clonic reflexes and bilaterally positive Babinski sign along with sensory deficits below Th6 level.

Spinal MRI revealed central, cervical disc herniation at C5/C6 level with reduction of the anterior buffer space with coexisting small, T2-hiperintensive lesion, possibly ischemic (fig. 1). Intramedullary, MRI revealed a T2-hiperintensive area approx. 17 x 9 mm in diameter that showed no contrast enhancement at the Th5 level. Similar smaller lesion was found at Th2 level (fig. 2).

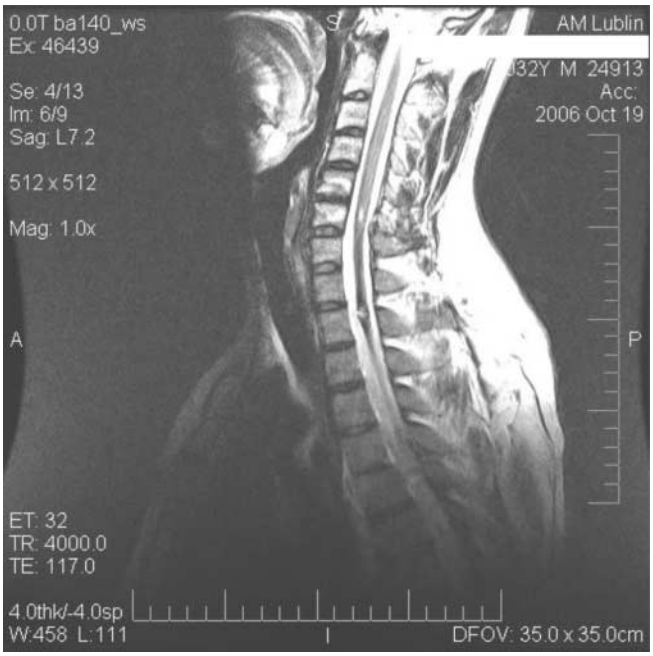


**Fig. 1.** MRI of patient number one

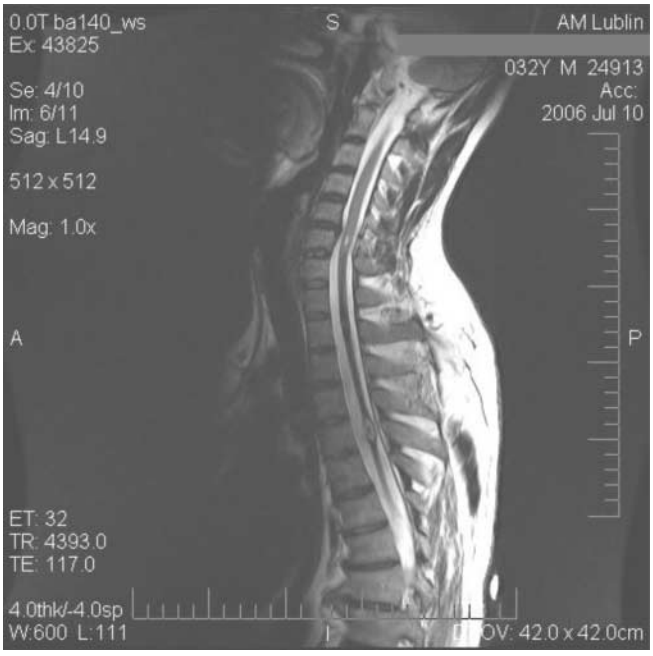


**Fig. 2.** MRI of patient number one

Primarily, the patient underwent C5/C6 discectomy m. Smith-Robinson followed by an intervertebral, polycarbonate cage implantation. In result, neck and shoulders pain subsided along with urinary disturbances. Paraparesis of the lower extremities remained stable. Consequently, the patient underwent surgery three months later.

Initially Th5/Th6 laminectomy was performed; dural sack was incised in the middle.

Thick, yellowish spinal cord following midline incision 3-4 mm under the surface revealed red-gray lesion built of enlarged, tortuous veins. Cavernoma dissected from the surrounding relatively easily, yellowish tissue showed increased consistency when compared to normal spinal cord. A gross total resection was achieved. In the early postoperative period paraparesis transiently aggravated; it has returned to preoperative level within days. Patient required permanent bladder catheterization. Histopathology confirmed cavernoma weaving.

**2 BT**

27-year-old male presented with 3-month history of right shoulder pain and right, upper extremity numbness. On admission to the Department he showed mild, right-sided hemiparesis, more prominent in the upper extremity. Additionally, patient reported slight sensory deficit on the right. Neurological examination revealed significantly elevated patellar reflexes, predominantly on the right, bilateral foot clonus and bilaterally positive Babinski sign.

MRI examination revealed intramedullary lesion at C5 level (fig. 3). Subsequently, the patient underwent C5/C6 laminectomy; dural sack was incised in the middle.

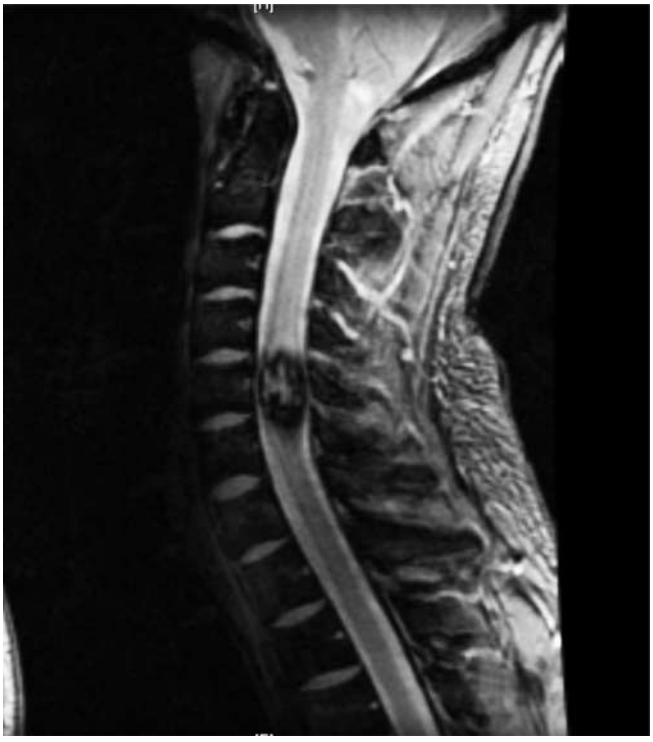

**Fig. 3.** MRI of patient number two

Yellowish spinal cord was slightly elevated; following midline incision 2 mm under the surface revealed red-gray, grape-like lesion that macroscopically resembled cavernoma. Cavernoma was surrounded by yellowish spinal cord and easily dissected from it. Cavernoma occupied most of the transverse section of the spinal cord and was removed in a piecemeal fashion under the microscope. Its midline part contained hemolysed blood. There were no complications in the early postoperative period and the patient was discharged from the Department without any changes in his neurological status. Histopathology confirmed cavernoma weaving.

**3 JT**

73-year-old male presented with 3-month history of progressive dysarthria, dysphagia and mild right-sighted hemiparesis. On admission to the Department neurological examination revealed severe dysarthria, mild right-sided hemiparesis, right-sided hemiatrophy of the tongue, upper extremities dyscoordination, more pronounced on the right side. Still, the patient was able to ambulate. MRI examination disclosed intramedullary, pathological lesion 19 x 9 x 8 mm at C1 level (fig. 4).

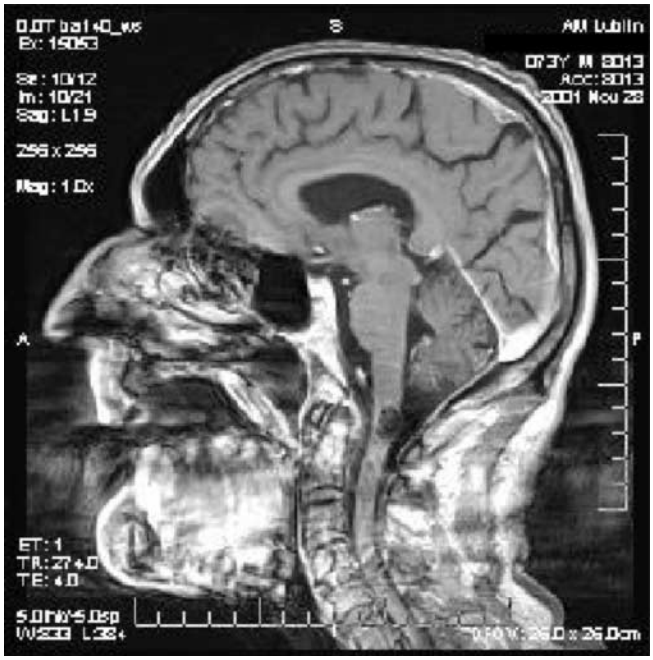

**Fig. 4.** MRI of patient number three

Based on clinical symptoms and radiological findings he was qualified for surgery. It involved suboccipital craniectomy with C1 laminectomy. Upon approach, on the surface of the spinal cord at C1 level a bluish-gray vascular lesion was found and it was subsequently dissected from the surrounding spinal cord. Malformation contained a blood clot in the middle. Immediately after surgery right upper extremity paresis deteriorated to very severe level while dysphagia and dysarthria subsided. Patient was discharged from the hospital in good condition, ambulating independently with permanent,

profound right-sided upper extremity paresis and mild lower extremity paresis. Again, histopathological examination confirmed cavernoma weaving.

## DISCUSSION

A number of publications stresses out that cavernomas are amongst the rarest spinal cord vascular malformations (1-6, 10, 11). The Department of Neurosurgery in Lublin has treated over seventy patients with cavernous malformations in the brain and only 3 patients with cavernomas in the spinal cord. It has been estimated that the ratio of brain to spinal cord cavernomas averages 10:1 (10). In our case it reached 23:1.

Symptomatic spinal cord cavernomas have been found in patients aged 12 to 88 years old. Similarly to brain cavernomas the incidence peaks in the forth and fifth decade, predominantly in women (1, 9, 10). Some authors estimate that cavernomas are found 1.5 to 2 times more often in women than men (1, 9, 12, 13).

Zevgaridis et al. (9) in their analysis proved that spinal cord cavernomas are most common in the thoracic spine (54%) then in the cervical spine (39%) while only 7% of cases were found in the lumbar spine. Similarly, Ogilvy et al. (1) described 36 cases of the spinal cord cavernomas with predominant location in the thoracic spine followed by the cervical and then lumbar spine.

One of our patients was diagnosed with two cavernomas, both of them in the thoracic spine after 12-year symptomatic period. The other patients were diagnosed relatively early, after no more than 4 months. In patients analyzed by Zevgaridis et al. (9) symptomatic periods varied from 1 week to 600 months and averaged 57 months.

Interestingly, no statistically significant differences were found between patients diagnosed with spinal cord cavernomas prior to MRI and after its implementation as a diagnostic tool. Clinical course varies from sudden onset to slowly progressive symptoms that developed over the years as it was in the first of our cases. Based on clinical presentation Zevgaridis et al. (9) divided their cohort into three groups. First group, which included 30% of patients presented with numerous, discrete neurological symptoms that ameliorated over the time and patients regained their health. These patients were usually diagnosed with multiple sclerosis and other demyelinating diseases or transverse myelitis (5, 9). Second and the largest group of patients presented with slowly progressive neurological symptoms. Slowly progressive spinal cord injury results from microbleedings from cavernoma with concomitant glial reaction to the blood components (2). Cavernomas that contain thin-walled pathological vessels filled with blood sometimes rupture and bleed into surrounding neural tissue due to blood pressure increase. Such repetitive microbleedings might result in “mass effect” and spinal microcirculation disturbances. Additionally a hyalinization of vascular walls within malformation occurs with subsequent clotting within the lumen of pathologic vessels that constitute a cavernoma (1). Direct toxic effect of hemosiderin on spinal neural pathways is also considered as one of the cause of progressive myelopathy. The third group of patients (26%) present with acute onset of the disease with pain resulting from spinal cavernoma hemorrhage and rapidly progressing motor and sensory deficits followed by bladder and bowels dysfunctions. In cases with cavernomas located superficially a subarachnoid hemorrhage might occur (1, 4, 9). This group might be subdivided into two additional groups – first (16% of patients) that develop spinal deficits within hours or days and second (10% of patients) in which it takes weeks or months to occur. Our second and third case might be allocated in this group. Interestingly, trauma, pregnancy and excessive physical activity might result in deterioration in patients with spinal cavernomas (4).

It has been estimated that 60-70% of spinal cavernomas eventually bleed and annual risk of hemorrhage ranges from 1.4 to 1.6% (9, 13). Brain cavernomas have significantly lower hemorrhage incidence at 10 to 36% (9, 14).

Spinal cavernomas are difficult to diagnose due to their low incidence and variable clinical course as described above (4). Numerous cases have been described where time period from onset to proper diagnosis ranged in years (4, 9). Similar pattern occurred in the first of our patients whose time course of the disease was 12 years.

Such difficulties have often been related to the imperfection of radiological methods implemented over the years. Myelography in majority of patients with small, intramedullary cavernomas showed no abnormalities. Only selected cases showed widening of the spinal cord or, in cases with exophytic growth, disturbances in contrast flow. Introduction of computed tomography combined with myelography improved diagnostics. Sometimes a calcification within the cavernoma might be found (6). Despite the fact that cavernomas are vascular malformations spinal angiography shows marginal utility in this disease (6). A breakthrough in diagnostics of spinal cavernomas came with magnetic resonance introduction. A radiological presentation of cavernomas is fairly characteristic – they usually present as distinct lesions with hypointense borders and often a hyperintense center in T2-weighted sequences (6).

Macroscopically cavernomas are usually well-margined deep blue or red-purple lesions that resemble raspberry or mulberry. Surrounding neural tissue is usually yellowish owing to the presence of hemosiderin deposits from previous hemorrhages.

All of our patients intraoperatively presented similar pattern. Histopathology reveals engorged, thin-walled vessels with single-layered endothelium and no neural tissue in-between. In majority of the cases macrophages loaded with hemosiderin or calcifications might be found as natural sequelae to hemorrhage from cavernoma (6).

Microsurgical removal is advocated for patients with symptomatic cavernomas (1, 4, 5, 9-11). Radiotherapy is inefficient in spinal cavernomas and is not recommended in their treatment (2). Only surgical removal restrains disease progression, prevents future hemorrhages and, in many cases, results in amelioration of the previous symptoms (6, 9). Surgical technique is similar to the one implemented for intramedullary tumors. In two of our cases laminectomy followed by posterior midline myelotomy was necessary in order to access the lesions. In the third described case cavernoma was superficial and easy to identify. Most difficult cases include those with ventral location in the cervical or thoracic spine where transthoracal approach with corpectomy or cervical corpectomy are often needed in relation to cavernoma location (4). Another clinical challange involves asymptomatic cases of spinal cavernomas. A question arises then whether these patients should be treated surgically or followed up with serial MRI studies (11). We faced similar dilemma in patient with multiple spinal cavernomas. This patient a year after surgery still requires rehabilitation while small Th2 level cavernoma remains asymptomatic.

For that reason we offered conservative treatment with periodic MRI imaging.

Despite the fact that these lesions are rare, we have mostly favorable outcomes of surgical treatment. Clinical symptoms ameliorated in patient with large cervical cavernoma, patient with multiple cavernomas, cervical discopathy and profound paraparesis remained stable, while the oldest, 73-year-old patient deteriorated: he had the upper extremity paresis, while speech and swallowing disturbances waned. Likewise Zevgaridis et al. (9) reported clinical improvement in 66% of cases, stable condition in 28% while only 6% of patients deteriorated.

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