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Case Report

Complete paraplegia due to multiple intracerebral and spinal cavernomas

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We report on a 29-year-old male patient with multiple intracerebral and spinal cavernomas. Bleeding in the thoracic cord at admission and additional bleeding which occurred 12 days later in the cervical cord resulted in complete paraplegia below thoracic level 4 (Th4). Four years earlier multiple cerebral cavernomas had been diagnosed by magnetic resonance imaging (MRI). Based upon reported cases in the literature multiple intracerebral and spinal cavernomas are exceptional. Additionally, the clinical presentation in our case is uncommon.

Keywords: paraplegia; cavernoma; multiple; spinal; intracerebral

Clinical case

A 29-year-old male patient presented with a history of decreasing strength in the right lower extremity leading to complete paraplegia at the midthoracic level within 24 h. On admission MRI of the cervical spine was found to be normal. MRI and CT-scan of the thoracic spine showed a multisegmental, intramedullary haemorrhage throughout the levels Th4-Th10 without radiologic evidence of mass effect. The typical appearance of a cavernoma at Th6 was found on MRI (see Figure 1). Four years earlier the patient had suffered from transient trigeminal hypaesthesia and paralysis of the facial nerve. At that time multiple cerebral cavernomas had been diagnosed on MRI. Since that time, the patient had been healthy and active in sports. No history of neurologic diseases or deficits exists in the family.

Twelve days after admission the patient developed tetraparesis below cervical level 6 (C6). MRI of the cervical spine showed multisegmental expansion of the myelon with a central lesion (see Figure 2). Laminectomy and decompression at C6/7 was performed. Histopathology showed haemorrhage associated with necrosis.

Tetraparesis resolved gradually. The neurologic deficits remained stable at thoracic level Th4 with a complete paraplegia (ASIA Impairment Scale: A, motor score: 50 of 100, total sensory score: 96 of 224)[1] and associated areflexive bladder dysfunction.

During his hospital stay the patient had an epileptic seizure. Multiple infra- and supratentorial lesions with hypointense iron-storage products on T2-weighted images were observed on MRI of the brain (see Figure 3). Medication with carbamazepine was started. No further seizures and/or changes of neurologic status were noted during the rehabilitation program. Further neurosurgical intervention was considered to have no beneficial effect. The patient was discharged to his home after 4½ months of intensive rehabilitation. The patient reached complete independence related to the neurologic level (functional independence measure [FIM] 115 of 126 at discharge).[1]

Discussion

Cavernomas consist of sinusoidal vascular spaces with single layers of endothelium. They belong to the slow-flow vascular malformations of the central nervous system.[2] Fifty-seven to seventy-five per cent of cerebral cavernomas are found supratentorially.[3,4]

MRI is the most reliable non-invasive diagnostic tool for their detection, especially when associated with haemorrhage.[2,3,5] MRI studies may be negative before the occurrence of bleeding,[6] as described in our case (cervical spine). The first clinical manifestation is mostly found in young adults (mean age 35 years).[4,7] Epilepsy is the leading clinical finding in 41–62% of the cases. Hemorrhage can be found in 23–33%.[3,4,7]

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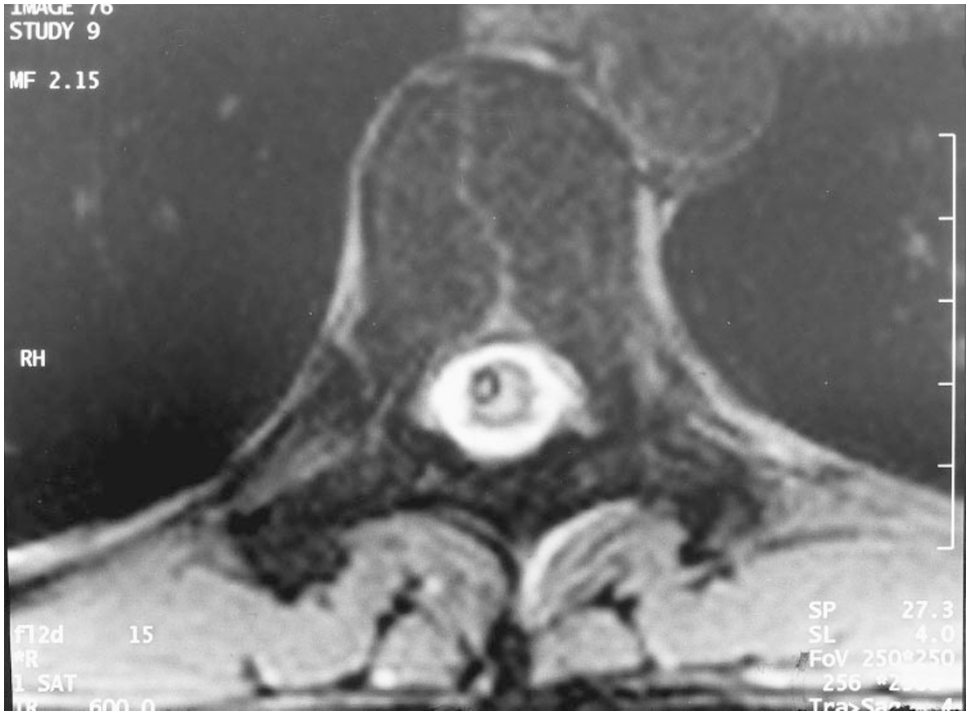

Figure 1 Axial T2-weighted image, thoracic level Th6. Typical appearance of a cavernous angioma located in the right anterior aspect of the spinal cord with markedly hypointense rim of iron-storage products *(Courtesy of Prof. Dr. M. Schumacher, Freiburg, Germany)*

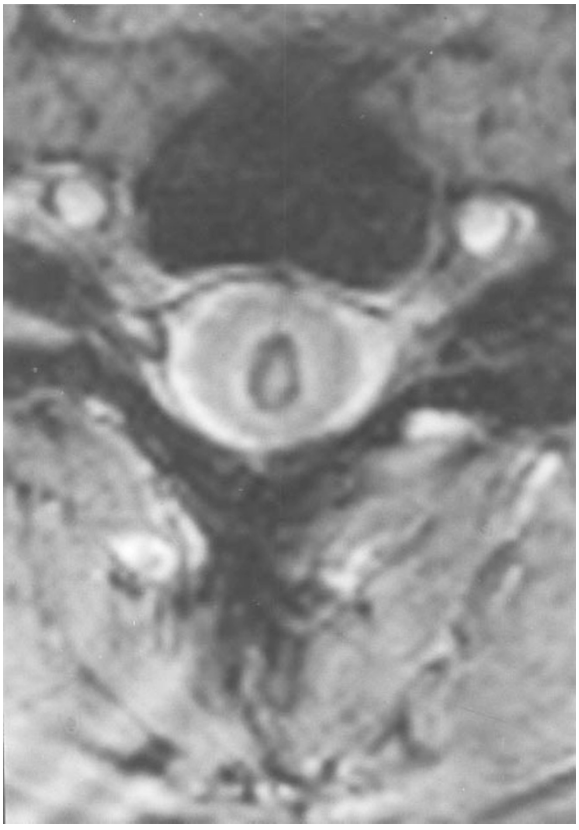

Figure 2 Axial T2-weighted image, cervical level C6. The cavernous angioma is located in the middle zone of the spinal cord *(Courtesy of Prof. Dr. M. Schumacher, Freiburg, Germany)*

Multiple intracerebral cavernomas are diagnosed in 9–16%.[2,3,7,8] Pozzati *et al*[8] isolated 18 of their 145 patients with intracerebral cavernomas with recurrent bleeding, growth and/or *de novo* appearance. Multiple appearances of cavernous angiomas, as described in our patient, are a leading risk factor for this 'aggressive biological behaviour' (others are female gender, pregnancy, previous radiotherapy).[8]

Spinal cavernomas are rarely found: Cantore *et al*[6] reported on 47 patients (six of his own patients and 41 described in the literature). Whereas cerebral cavernomas do not display a gender prevalence,[9] those of the spinal cord favour women (69%). Only one of 47 patients was found to have two spinal cavernomas. Four had additional cerebral cavernomas. Multiple intracerebral and spinal cavernomas are therefore exceptional.[6,10]

Neurosurgical treatment of singular cerebral cavernoma is mostly recommended in symptomatic patients presenting with neurological deficits due to haemorrhage and mass effect.[11–13] Whether neurosurgical resection is indicated in asymptomatic patients or in patients presenting with seizures remains controversial.[11,12,14]

Little information could be found concerning the treatment of singular spinal cavernomas.[6,15] Spetzger *et al*[15] reported a series of nine patients recommending microsurgical resection. Experience with neurosurgical treatment of multiple cavernomas–either cerebral or spinal–has not been reported so far.

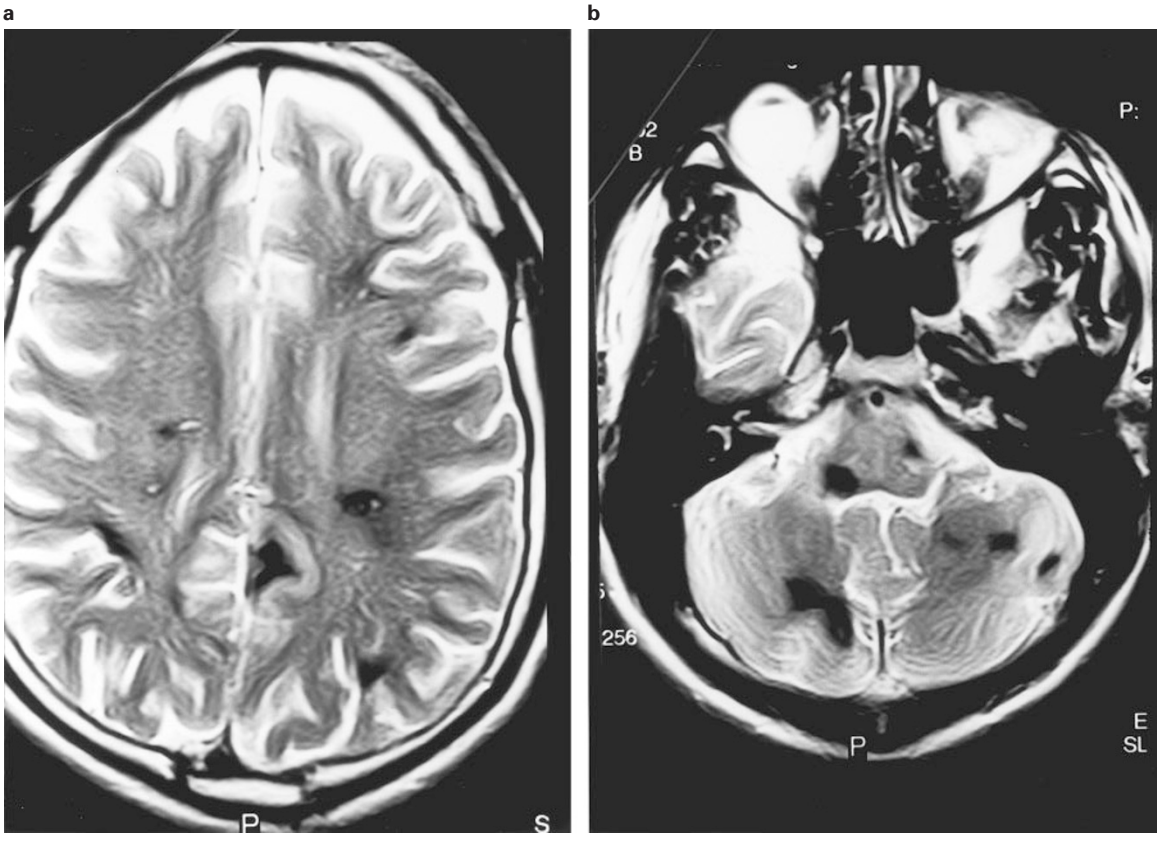

Figure 3 (**a**) Axial T2-weighted image, brain. Multiple cavernous angiomas subcortical and deep white matter. Typical feature with focal methaemoglobin, complete rim of hypointense iron-storage products and no edema. (**b**) Axial T2-weighted image, brain. Multiple cavernous angiomas of the brainstem and cerebellum

References

1 Maynard FM *et al*. International standards for neurological and functional classification of spinal cord injury. *Spinal Cord* 1997; **35:** 266–274.
2 Requena I *et al*. Cavernomas of the central nervous system: clinical and neuroimaging manifestations in 47 patients. *J Neurol Neurosurg Psych* 1991; **54:** 590–594.
3 Schefer S, Valvanis A, Wichmann W. MRT-Morphologie und Klassifikation der zerebralen Kavernome. *Radiologe* 1991; **31:** 283–288.
4 Lonjon M *et al*. Intracranial cavernomas. 30 cases. *Presse Med* 1993; **22:** 990–994.
5 Turjman F *et al*. Hemorrhagic complications of intracerebral cavernomas: Value of MRI. *J Neuroradiol* 1992; **19:** 107–117.
6 Cantore G *et al*. Intramedullary cavernous angiomas of the spinal cord: report of six cases. *Surg Neurol* 1995; **43:** 448–451.
7 Cubo DE *et al*. Cerebral cavernoma. A retrospective analysis of 24 cases. *Neurologia* 1995; **10:** 277–282.
8 Pozzati E *et al*. Growth, subsequent bleeding and de novo appearance of cerebral cavernous angiomas. *Neurosurg* 1996; **38:** 662–669.
9 Simard JM *et al*. Cavernous angioma: a review of 126 collected and 12 new clinical cases. *Neurosurg* 1986; **18:** 162–172.
10 Abid R *et al*. Brain and spinal cord cavernoma. Value of MRI and review of the literature. Apropos of a case. *J Radiol* 1993; **74:** 563–567.
11 Bertalanffy H, Gilsbach JM, Eggert HR, Seeger W. Microsurgery of deep-seated cavernous angiomas: Report of 26 cases. *Acta Neurochir* 1991; **108:** 91–99.
12 Braun V, Antoniadis G, Rath S, Richter HP. Kavernome. Indikationen zur operativen Entfernung und Ergebnisse. *Nervenarzt* 1996; **67:** 301–305.
13 Cassaza M *et al*. Supratentorial cavernous angiomas and epileptic seizures: Preoperative course and postoperative outcome. *Neurosurgery* 1996; **39:** 26–31.
14 Churchyard A, Khangure M, Grainger K. Cerebral cavernous angioma: a potentially benign condition? Successful treatment in 16 cases. *J Neurol Neurosurg Psych* 1992; **55:** 1040–1045.
15 Spetzger U, Gilsbach JM, Bertalanffy H. Cavernous angiomas of the spinal cord. Clinical presentation, surgical strategy and postoperative results. *Acta Neurochir* 1995; **134:** 200–206.