Surgical management of symptomatic spinal cord and intracerebral cavernomas in a multiple cavernomas case

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Abstract: Multiple cavernous malformations are associated with familial cases and are present in 10-20% of all cavernoma cases. 5% of cavernomas are located intramedullary and of these only 10% present multiple cavernomas. With the availability of echo gradient MRI the cases of multiple cavernomas are diagnosed earlier and it is not rare that it uncovers multiple cavernomas in cases where only a single lesion can be identified on regular MRI sequences. We present the case of a 55 years old woman presented with a two years history of mild backache, followed by progressive lower legs motor deficit and urinary retention. The spine MRI showed an intramedullary T2/3 lesion and the cerebral MRI established the diagnosis of multiple cavernomas. One year after the intramedullary cavernoma was operated with success, she developed generalized seizures and a new cerebral MRI showed bleeding and volume growth of one right temporal pole cavernoma. The cerebral lesion was resected successfully and the patient was discharged free of seizures. This familial type multiple cavernomas cases should be screened and followed with repeated brain and spine MRI's every year.

Key words: multiple cavernous malformations, spinal cord, seizures, motor deficit

Introduction

Multiple cavernous malformations are associated with familial cases and are present in 10-20% of all cavernoma cases. 5% of cavernomas are located intramedullary and of these only 10% present multiple cavernomas. Reports of multiple cavernomas are yet rare in literature and they usualy focus on intracranian lesions. After the MRI was introduced in clinical practice the study of cavernous malformations shifted from a few anecdotal case reports to large series, some of them larger than 300 cases. With the availability of echo gradient MRI the cases of multiple cavernomas are diagnosed earlier and it is not rare that it uncovers multiple cavernomas in cases where only a single lesion can be identified on regular MRI sequences (1, 6, 9).

Spinal intramedullary cavernomas are at times difficult to differentiate from other

intramedullary tumors but a few MRI characteristics like the hypointense hemosiderin ring surrounding the lesion should orient the diagnosis. Compared to intracranian lesions the compactness of white matter long tracts and grey matter nuclei intramedullary makes dangerous lesions of the cavrnomas in this location. This is the reason why most of the patients with both spinal and intracranian CM have the intramedullary lesion removed first. (3, 16, 17)

We present a young woman's case of multiple cavernomas located both at spinal level and intracranian levels, with emphasis on the surgical technique for removal of two intramedullary and temporal lesions.

Case presentation

A 55 years old woman presented with a two years history of mild backache. A few months before admission she noted mild lower legs motor deficit progressing to difficulty ambulating without help and urinary retention. The patient had previous surgery for a thyroid tumor and hysterectomy and she received medication for mild arterial hypertension.

On admission she showed 3/5 motor weakness and mild sensory deficit in both lower limb. Her ankle and knee reflexes were diminished and she had difficulty urinating.

The MRI scan showed an enlarged spinal cord at the T3-T4 level with an inhomogeneous central signal and a peripheral hyperintensity prolonged from T2 to T4 disk. An area corresponding to T3 disk abnormal serpentine vessels were observed. No angiography was performed because the brain MRI confirmed the diagnosis of multiple cavernomas.

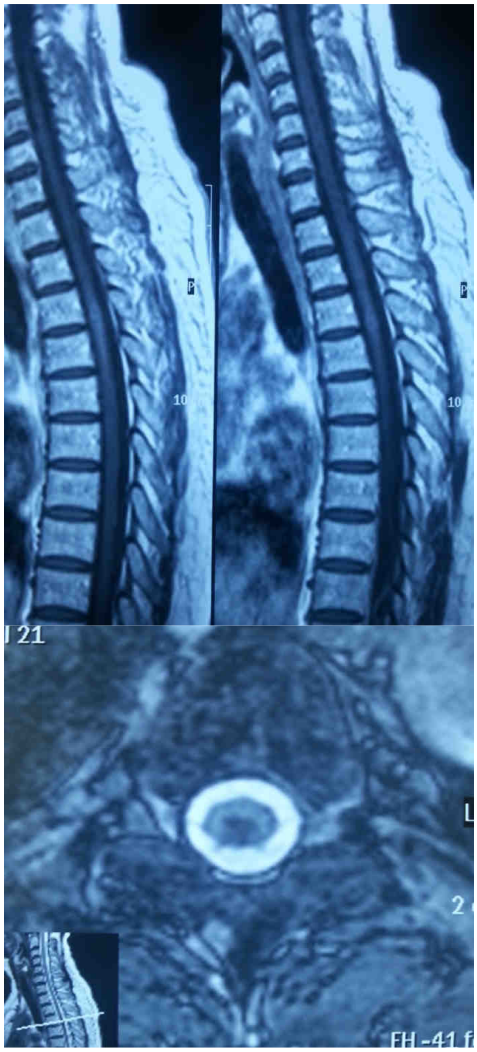

Figure 1 - T2-T3 intramedullary lesion, a hypointense signal in T2 indicates old bleeding

First surgery

The patient was placed prone on the operating table with the head resting in horseshoe. A midline incision over the involved level and a laminectomy centered on and continued one level above and one level below the lesion were performed. The operative microscope was used. The spine was

not considered instable as the articulars were left intact at all levels. After opening the sura in the midline the arachnoid was incised and suspended to the dura from T2 to T4 and then the pia was opened and stitched laterally to the dura in a few suture points. With gentle moves of the baionet the medulla was split uncovering a hemosiderin rich gliosis tissue. Deep to this tissue a tangled vessels reddish blue tumor was identified and gently dissected circumferential from the surrounding gliosis. An afferent vessel coming from the anterior spinal Artery was coagulated and cut before complete resection. The pia was approximated with a few interrupted sutures and the arachnoid was closed with continuous 6.0 and the dura was tightly closed with 4.0 surjet.

The patient's deficit persisted postoperatively. It took 6 months of motor recovery for the patient to be able to ambulante with a cane and to completely recover the bladder function.

As none of the cerebral lesions were symptomatic the patient was treated conservatively for 1.4 years. As she suffered a few temporal seizures she was readmitted and a new MRI was performed.

The echo gradient sequence showed a big 2/3 cm hypointense lesion in the right temporal pole and many infracentimetric lesions distributed both infratentorial and supratentorial bilateral, the biggest of the infratentorial lesions was located at the left Posterior medulla level, pand of the supratentorial lesions, in the inferior parietal lobule. The patient was operated for resection of the right temporal pole cavernoma that increased in size due to rebleeding and manifested with seizures.

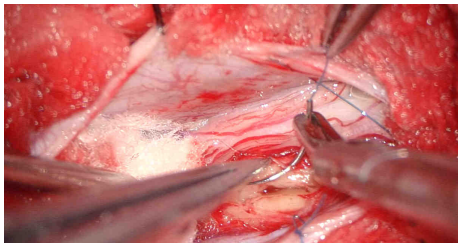

Figure 2 - After the dura has been opened over the tumor, the pia is retracted by interrupting sutures

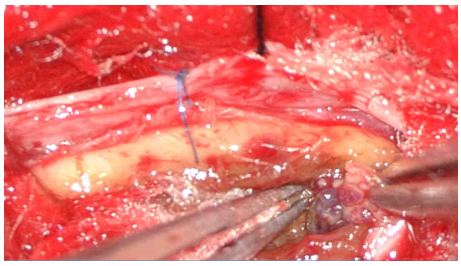

Figure 3 - The strawberry like lesion is found surrounded by a yellow, hemosiderin gliosis tissue

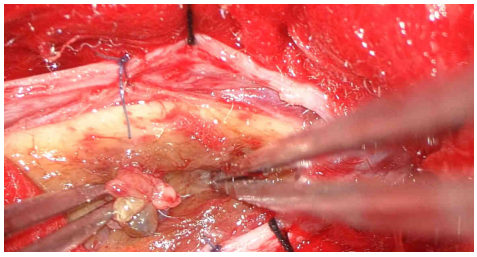

Figure 4 - Complete removal of the tumor using extracapsular dissection

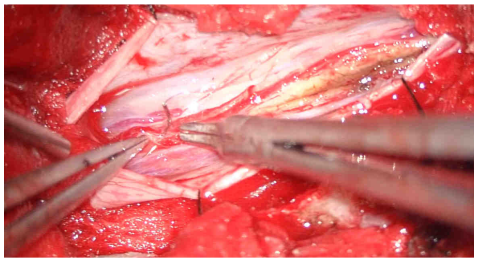

Figure 5 - The pia is sutured after hemostasis was achieved

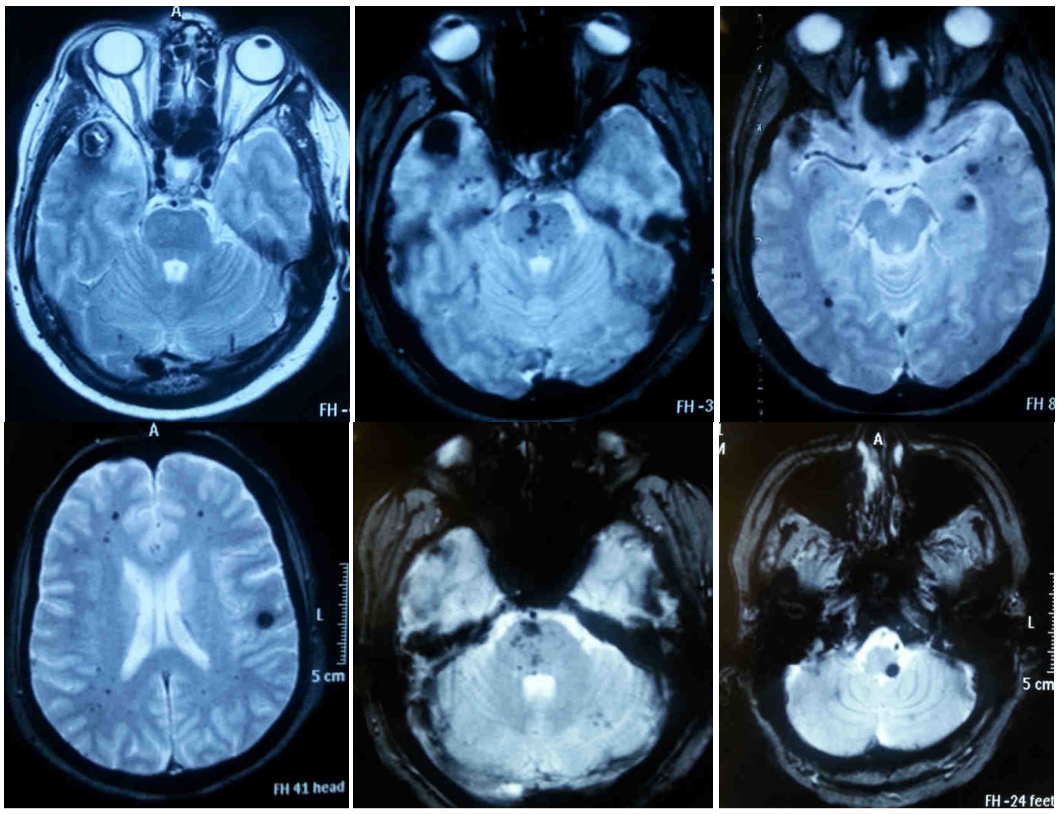

Figure 6 - Echo gradient MRI sequence showing multiple cavernomas the biggest of the in the right temporal pole. Many small cavernomas are located in the brain stem

Second surgery

After elevating the right shoulder the head was rotated 45 degrees to the left and fixed with tape to the horseshoe. A curved incision starting 2 cm in front of the tragus and continued inside the hairline towards the midline was performed to allow the execution of a frontotemporal craniotomy. The scalp and the temporalis muscle were dissected with care in separate plans to avoid injury to the superficial temporal Artery of to the facials nerve. A small craniectomy was performed using a perforator and gigli saw and after removing the bone the sphenoid wing was drilled flat with the middle fossa floor. The dura was incised with superior pedicle and a subtemporal route was choosen to open the basal cysterns. When a good brain relaxation was obtained, an approach through the Sylvian fissure was used to see the discoloration area the cavernoma created in the inferior temporal girus. Using the hemosiderin as landmark the cavernous malformation was approached and dissected circumferentialy using suction and cottonoids. After coagulating the feeder coming from an M3 branch the lesion was removed as a single piece with the surrounding hemosiderin ring attached. The lesion bed was inspected for a good hemosthasis and the dura was tightly sutured and suspended. An epidural drain was left in place under the bone flap and the scalp was sutured.

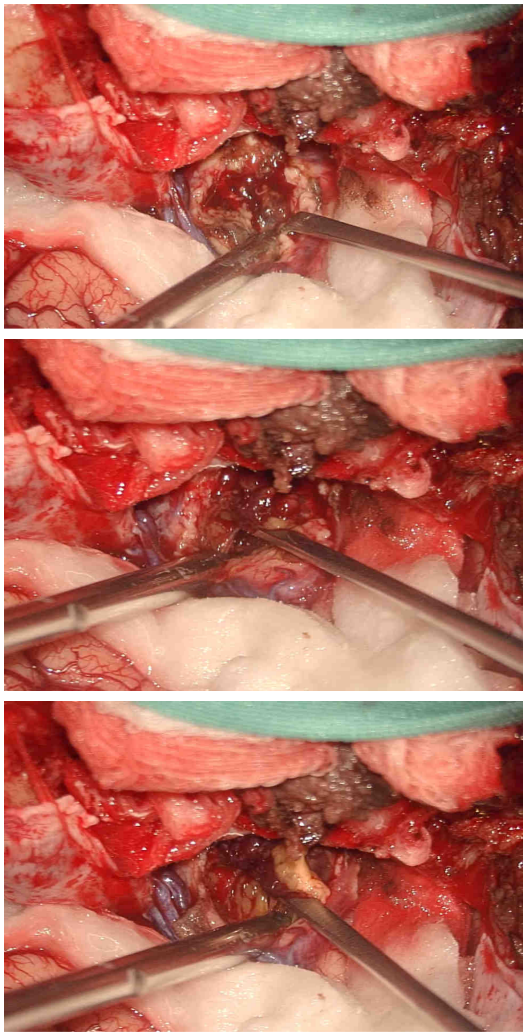

Figure 7 - Using a Sylvian fissure corridor and a small corticotomy in T 1 the cavernoma is identified and circumferentially disected along with the hemosiderin rich gliosis ring

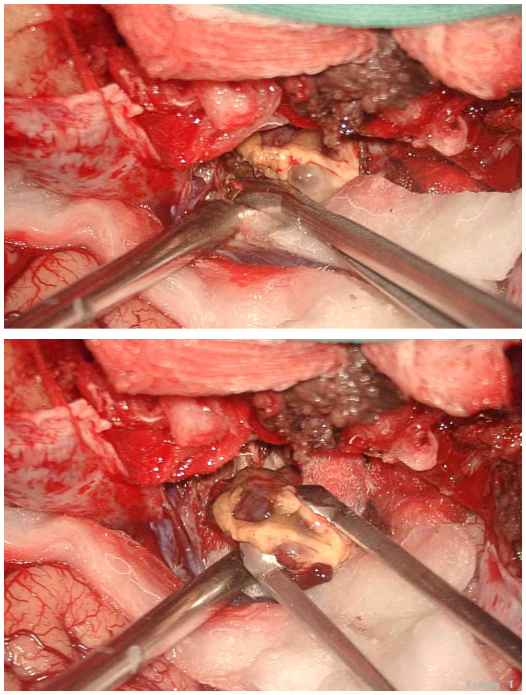

Figure 8 - Using cottonoids to create a plane between the lesion and the parenchima, the cavernoma is resected in one piece

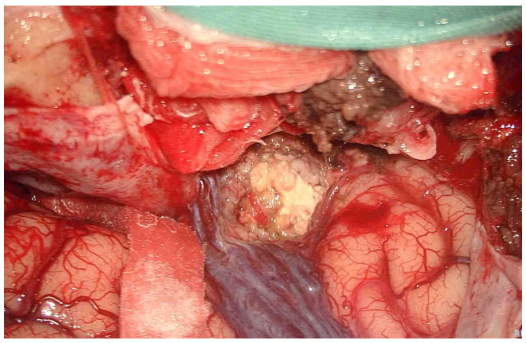

Figure 9 - The cavernoma bed after resection and hemostasis

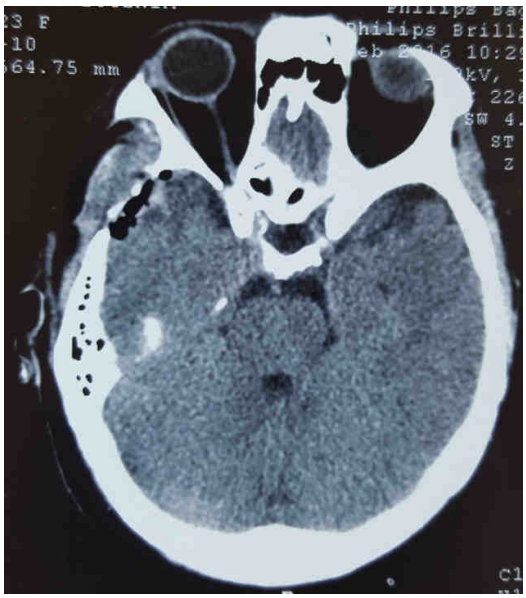

Figure 10 - Postoperative CT scan, showing total resection of the cavernoma

The postoperative course was uneventful with no deficits or seizures. The patient was discharged in good health one week after the surgery.

Discussion

Intramedullary cavernous malformations are rare lesions with only few cases reported in literature yet the association between intramedullary location and the presence of multiple cavernomas is extremely rare. (2, 4, 7) The most common site for intramedullary lesions is the thoracic cord, but they have been described at cervical level and cauda equina. (5, 8, 12) In most of the cases the slowly progressive myelopathy is the usual presentation a fact that is characteristic for low flow vascular malformations with tendency to repeated bleeding episodes, such as these. In the case of supratentorial location the most common presentation are partial or generalized seizures preceded by a few years history of headache. (11, 13) The cavernomas present as mixed signal intensity masses surrounded by a decreased signal intensity rim yet this was not the case here as both the spinal and the temporal lesions had bled recently.

Given the high density of eloquent areas intramedullary these lesions are better observed if asymptomatic. (14, 15)

Cases like this with multiple cavernomas will unfortunately never be cured of the disease and will come to the attention of the neurosurgeon every time one small lesion will encounter acute growth following a bleeding episode.

Conclusion

Multiple cavernoma cases present difficulties in the strategic long term management and surgery as the only curative option should remain the ultimate alternative when the new neurological deficits arise. This reported case is illustrative for the need to perform both cerebral and spinal MRIs in patients suspected for multiple cavernomas and these patient's family members should be investigated to rule out this pathology.

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