Intramedullary spinal cord tumour (hemangioblastoma) - Clinical case presentation

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Abstract

Hemangioblastomas represent cca. 1 - 2,5% of all the intracranial tumours and only 2 -3% at the level of the spinal cord. The symptomatology, usually insidious, is directly proportional with the size of the tumour. The native magnetic resonance imaging (MRI) scan and the use of a contrast agent is the investigation of choice for diagnosing intramedullary tumours. Authors report the case of a 51 years old man patient admitted in our institution for an intramedullary spinal cord tumor, developed insidious. The lesion was completely removed. Postoperatory outcome was excellent, and the histopathological result was: hemangioblastoma.

Keywords: intramedullary spinal cord tumor, hemangioblastoma, surgery, von Hippel-Lindau

General Data

Intramedullary tumours are rare and account for cca. 4% of all the tumours of the central nervous system, among them, the most common being ependymomas and astrocytomas. Ependymomas are more commonly seen in adults, while astrocytomas are predominant in children. Other types of intramedullary tumours (cavernomas, hemangioblastomas, lipomas, etc.) occur much more rarely (1, 2, 4).

Hemangioblastomas represent cca. 1 - 2,5% of all the intracranial tumours; 85% of these, develop at the level of the posterior cranial fossa and only 2 -3% at the level of the spinal cord, the rest being located in the supratentorial (3, 4).

The symptomatology, usually insidious, is directly proportional with the size of the tumour, its location and the presence of the peritumoral edema and syringomyelia (9).

The most common symptoms (8, 11) are:
- Local pains
- Motor deficits (paraparesis, tetraparesis)
- Sensitivity disorders (paraesthesia, dysaesthesia)
- Radicular pain
- Urinary disorders

The native magnetic resonance imaging (MRI) scan and the use of a contrast agent is the investigation of choice for diagnosing intramedullary tumours. It can highlight both the highly vascularised tumour components and the cystic fluidal components. The medullary cord can be enlarged by an edema or vascular congestion. There can be found haemorrhagic foci and peripheral deposits

of hemosiderin. Siringomyelic cavities in hypersignal T2 and izosignal T1 can be associated, depending on the protein level.

An angiography can be useful for diagnosing the lesion, as well as for identifying the vascularisation and the drainage vessels (4, 5, 7).

Intraoperative monitoring of the motor and sensory evoked potential represents a useful procedure for an adequate functional postoperative result.

Case presentation

Fifty-one year-old patient is hospitalised at the Neurosurgery Clinic I of "Bagdasar-Arseni" Emergency Hospital in Bucharest because of cervical pains, incomplete tetraplegia with C5 Frankel D level and symptoms appearing insidiously for cca. 2 months that did not ameliorate.

An MRI of the cervical spine showed a tumoral intramedullary mass eccentrically and posterolaterally situated on the left in the medullary cord, in the 2/3 posterior, of ovoid shape, cca. 11/12 mm in size, with an izo – hipointense T1 inhomogeneous signal, with increased gadolinium uptake in T1, with perilesional edema which led to the tumefaction of the medullary cord. The lesion is in contact with the leptomeninges and presents intramedullary cystic masses, hidromyelic cavities cranially extended up to the level of the medulla oblongata/corticobulbar tract and caudally up to 1/3 medium of the vertebral body T6.

Thinking of the Von Hippel-Lindau disease, we expanded our investigations: the abdominal CT-scan did not show pathological lesions on the examined segments and the native cerebral MRI and with contrast did not reveal any pathological processes. The ophthalmological examination was within normal limits.

Preoperative preparation

Preoperatively, he was administered a steroid-type anti-inflammatory (Dexamethasone) and cephalosporin antibiotics at the incision of the tegument (2 g iv cefort). A urinary catheter was inserted and it was removed 2 days post operation.
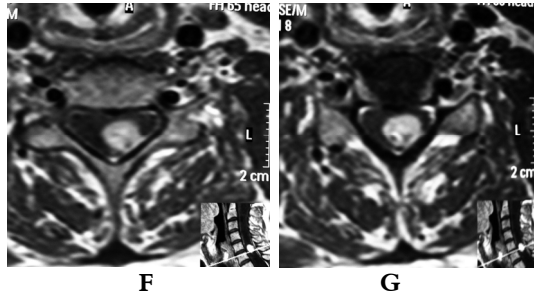

A   B

C   D

E
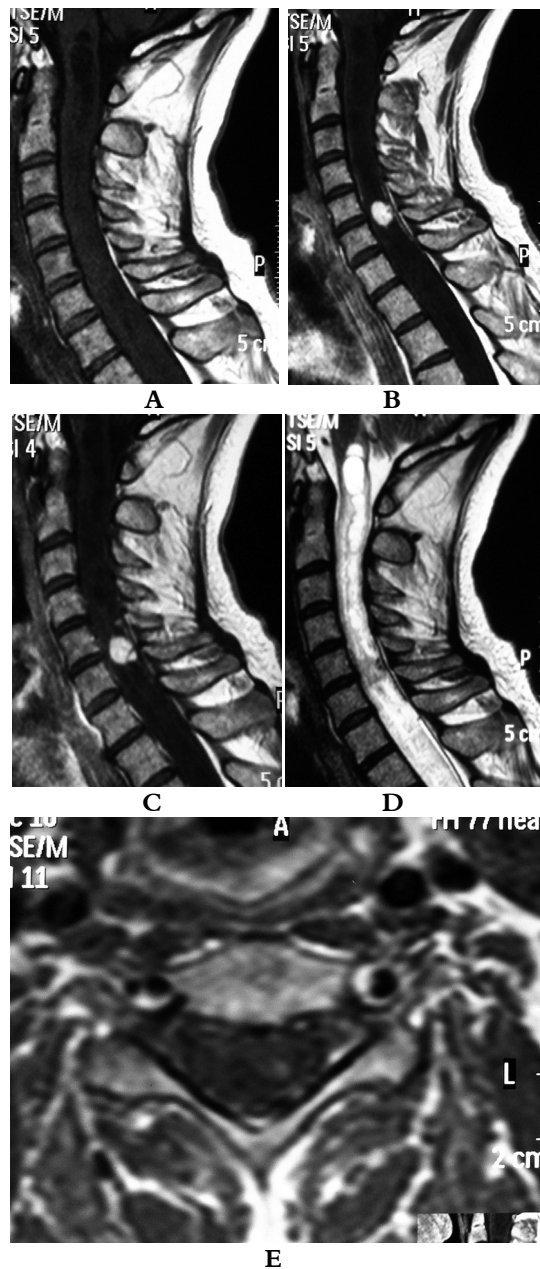
Figure 1 MRI cervical spine: **A -** sagittal section T1; **B, C** sagittal section T1+K ; **D** – sagittal section T2 ; **E** – axial section T1 ; **F, G** axial section T1+K

The surgical intervention

The patient was in the prone position, and the operative field was sterilized with Betadine. A median incision was made into the skin and after pulling the muscle apart from the bone, a rigorous hemostasis followed. C5 level was radiologically found, after which a C5 and C6 laminectomy was performed and a thorough hemostasis in the epidural space, with electrocoagulation and Surgicel. The dura mater was incised along the midline and held elevated with Vicryl 4.0 suture. The dissection in the subdural space and especially the dissection of the tumour were performed using an operating microscope.

The arachnoid was carefully incised and moved to the side. The inspection of the medullary surface showed an exteriorised tumour under the pia mater which dissected the posterolateral medullary cords on the left side and was covered with dilated subpial veins. The tumoral mass was red, bloody, 11/9/12 mm in size, very well vascularised (Figure 2 A, B). The pia mater was peritumorally incised and the veins were partially coagulated with care, partially dissected to reveal the tumour capsule. A 6.0 suspension suture on the pia mater was useful for dissecting the tumour.

Once made visible, the interface between the tumour and the medullary parenchyma is circumferentially delimited through dissection. Coagulation must be used at really small values so as not to tear the peritumoral vessels. The dissection of the tumour is done with cottonoids that are circumferentially placed, around the tumour. The easy, repeated coagulation of the tumour capsule causes the slow shrinking of the tumour and the interface between the tumor and the spinal cord was more easily exposed. After the complete dissection was performed, the tumour is totally ablated, en bloc, and the vascular pedicle being coagulated and sectioned at the end (Figure 2 C, D). The arachnoid was sewn with separate monofilament 6.0 sutures and the dura mater in the watertight manner with Vicryl 4.0.

From a histopathological point of view at the optical microscopy (objective 10x, haematoxylin and eosin stain) a tumoral mass showed up composed of a cell population, rod-shaped, small in size, mitotically inactive, closely connected with numerous blood vessels, capillaries or arterioles, for the most part monolayers.

The cells have an uncertain histogenesis, different histological origins being assigned to them, from the vascular cells passing through the glial cells, to the neuronal cells. Thanks to this, the tumour is put in the category of tumours with uncertain histogenesis (Figure 3 A). In Figure 3 B, using the haematoxylin and eosin staining method (objective 40x), an abundant vascular network of capillaries uniformly distributed among the tumour cells can be seen. Final histopathological examination: HEMANGIOBLASTOMA.
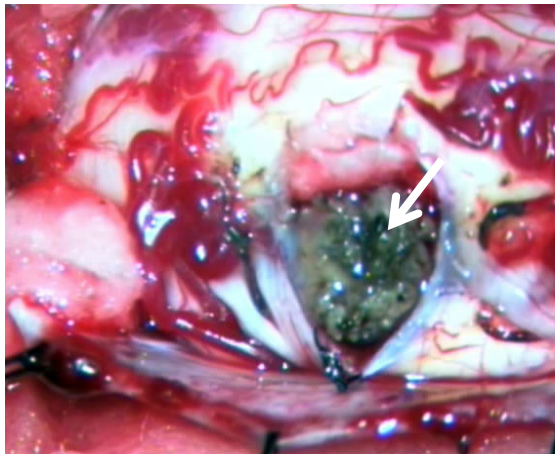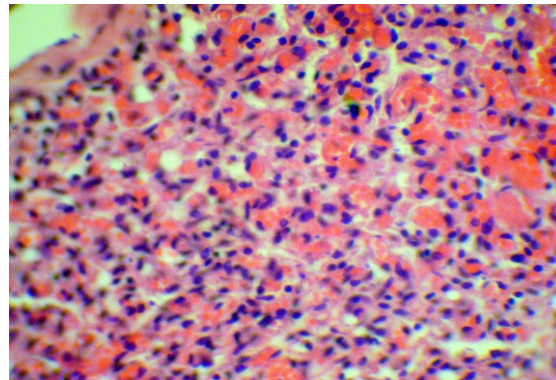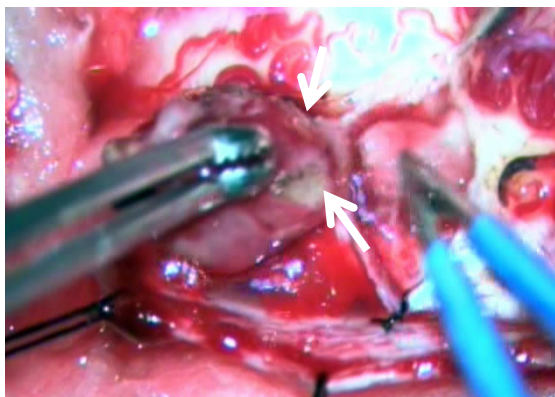
A

B

C
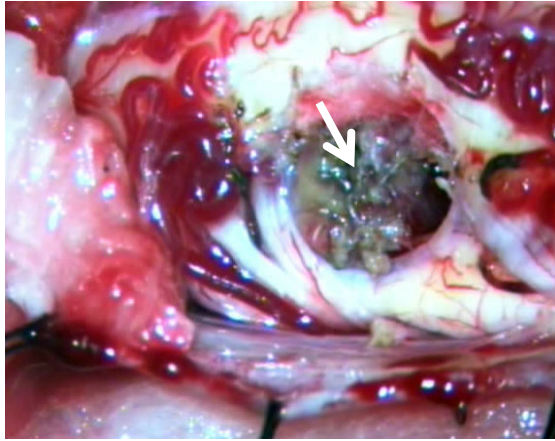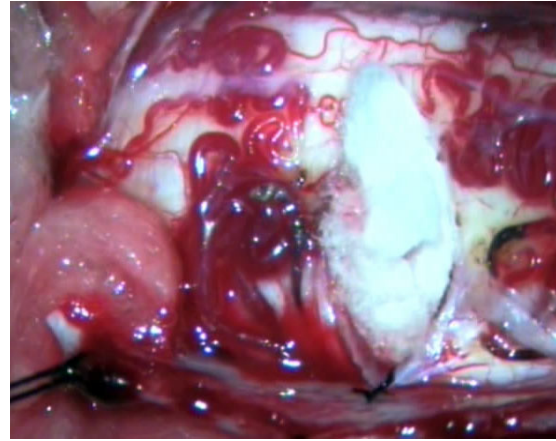
D

Figure 2 Intraoperatory pictures: **A, B, C** – the dissection of the tumour from the surrounding glial tissue and the en bloc; d – aspect after the total ablation of the tumour
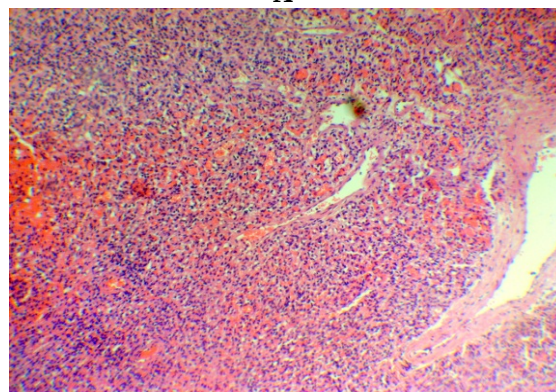

A

B

Figure 3 Histopathological aspect: **A** (10x objective) haematoxylin and eosin stain; **B** (40x objective) haematoxylin and eosin stain

Postoperative evolution

Postoperator, the patient was surveyed another 24 hours in the intensive care unit (ICU) where the administration of Dexamethasone 1v/day and Clexane 0.6 ml, 1v/day continued. 24 hours after the operation, the drainage tube was removed and precocious mobilisation and kinetotherapy commenced to prevent deep venous thrombosis. Preoperative deficits were gradually cured after the operation.

Five days after the operation an MRI is taken of the cervical spine, check-up which confirms the total ablation of the tumour and the diminishing of the siringomyelic cyst (Figure 4). Seven days after the operation, the patient was released from hospital, as he was neurologically ameliorated. There were no postoperative complications.

Discussions

Hemangioblastomas are, histologically speaking, very well vascularised benign tumours, most frequently found in the posterior cranial fossa. They can develop spontaneously or within the von Hippel Lindau disease (3).

Figure 4 Cervical spine MRI contrast (postoperative aspect)

The von Hippel Lindau disease is a rare autosomal dominant genetic syndrome, characterised by the growth of retinal angiomas, hemangioblastomas at the level of the brain and spinal cord, renal and pancreatic carcinomas, pheochromo-cytomas, polycythaemia (6, 10).

As is stated above, they are benign tumours according to the WHO classification, although they can disseminate via the cerebrospinal fluid pathways after surgery. They can be solid or cystic tumours, with a highly vascularised mural nodule.

If the hemangioblastoma is joined by a siringomyelic cyst, a puncturing of the cyst is performed together with draining the liquid, moving on afterwards to dealing with the tumour nodule. The dissection and ablation of the intramedullary tumours is done only under the operating microscope, this ensuring proper lighting and optimal magnification, this way the demarcation line between the tumour and the surrounding glial tissue becoming apparent.

It is essential that the ablation must be done en bloc, unlike other types of intramedullary tumours (ependymomas, astrocytomas) where a debulking of the tumour can be useful.

For tumours associated with intratumoral cysts, the ablation of the tumour nodule and opening the cyst suffices (1). In principle, the siringomyelia does not require draining. If after the spinal MRI more lesions are found (von Hippel Lindau disease), the lesion responsible for the symptoms must be dealt with.

In case of cervical lesions, the patient should preferably be extubated in the ICU where he will be monitored at least 24 hours. Mobilisation must be done as early

as possible in order to prevent deep vein thrombosis.

The most common complications that arise with such lesions are: intramedullary hematoma (if the tumour has not been entirely ablated), epidural hematoma, arachnoiditis, meningocele or cerebrospinal fluid fistula. Postlaminectomy cervical kyphosis is more common with children and it can be prevented by performing laminoplasty or laminotomy (1).

Conclusions

Spinal hemangioblastomas belong to the group of intramedullary tumours, along with ependymomas and astrocytomas, a rarely encountered entity in neurosurgical spinal pathology. They are highly vascularised tumours and curing is possible only with total ablation through a microsurgical approach. The pia mater is incised in the place where the tumour is exteriorised from the medullary parenchyma and the ablation of the tumour is performed en bloc, unlike the other intramedullary tumours (ependymomas, astrocytomas) where the myelotomy is median and the tumour is sometimes ablated using the intratumoral debulking procedure.

The postoperative results depend not only on the experience of the neurosurgeon, but on the preoperative neurological status as well.

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