Benign neurosurgical disease

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Case presentation and history:

Here we shall describe a case of a 39 years old male. Medical history was uneventful except for arterial hypertension treated by indapamide 2.5mg once a day. This patient suffered severe headaches and back pain in 2011. Neurological examination was normal at that time according to reports from outside hospital. Both brain CT and MRI were normal. Patient suffered other three identical episodes of severe headaches and back pain during following 14 months. CT angiography was performed as well and was negative. Lumbar puncture showed signs of subarachnoid hemorrhage, spectrophotometry showed pure bilirubine curve with maximum in range of 450 to 460 nm.

Then our Department was consulted. Cerebral CT angiography and MR angiography were available at that time, both were well performed and were negative.

What should be performed at this moment? Cerebral DSA? Or cerebral DSA + spinal angiography?

We considered that both investigations should be performed in setting of negative MRI of the spine. Would the reader of this case follow the same strategy?

We asked our colleagues to perform MRI of spine before angiography. MRI of cervical spine and up to 6th thorax vertebra was negative. Then we scheduled the patient for angiography with request to add MRI of lower thorax spine and lumbosacral region.

This MRI disclosed multiple extramedullar intradural contrast enhancing lesions. The biggest two tumours were at level of L1 and L2. Tumour was found in sacral canal as well. Several smaller tumours were found in lumbar region, at level of Th 8 and. (Fig. 1-4)
It is questionable whether this finding answers the episodes of severe headaches and back pain of the patient. Nevertheless, it is quite obvious that these tumours caused subarachnoid hemorrhage during the last clinical episode. It seems to be probable that everybody would consider surgery at this time. We add the information that lumbar puncture did neither reveal tumour cells nor signs of inflammation.

What would be the differential diagnosis? Ependymoma? Multiple neurinomas? Metastatic disease? Multiple paragangliomas?

We suspected that ependymoma would be the most probable diagnosis due to typical location of tumours and contrast enhancement.
What would be the goal of the surgery? Resection of both the lesions in lumbar and sacral region? Or simple biopsy? Or resection of the major lesions in lumbar region?

We did not consider all tumours as radially resectable. We decided for the resection of tumours at level L1 and L2 at the first step.

**Procedure and outcome:**

Surgery was done in standard fashion under electrophysiological monitoring. Signs of older subarachnoid hemorrhage were found after dura opening. One tumour growing at level of conus medullaris and the other one growing at fillum terminale were resected. Tumours looked like ependymomas. Spreading of minimal tumour layer was observed on several roots of cauda equina.

Postoperatively, patient had paresthesias in the right lower extremity, not in clear radicular distribution. Patient was discharged home on postoperative day 6.

Histological examination revealed typical myxopapillary ependymoma (grade I) (Fig. 5).

Fig. 5:

What should we do then? Histological findings reveal benign lesion. On the other hand, there was a multiple spreading of tumours. We found radical resection of all tumours not to be feasible.
We discussed the case at our neurooncological committee and decided for watch and wait strategy due to typical histological finding of GI ependymoma.

MRI of brain and whole spine was performed 3 months later. No tumour intracranially was found. New small tumours at level of surgery (L1 and L2) were found. Tumour at level L5 showed signs of minor growth. New tumour at level C7 was found as well (Fig. 6-7).
Patient was neurologically stable.

What to do at that stage? New surgery to gain new histological sample? To perform radiotherapy for histologically benign disease?

We decided for spinal radiotherapy due to disseminative fashion of disease and signs of growth within 3 months after surgery.

Last follow-up was performed 6 month after surgery and 1 month after radiotherapy. Patient was neurologically stable and MRI showed no signs of tumour growth.

We select on purpose this case to be presented. This is a recent case with an opened end. We are fully aware that we offer more questions than answers. We believe that this kind of a case may rise a discussion and that we get important feedback as well as advise from readers.

Plans et al. described a case of intracranial dissemination of filum terminale myxopapillary ependymoma and reviewed literature on this topic. They found 23 cases in literature describing dissemination of myxopapillary ependymoma.

Tarapore et al. presented a very interesting study on spinal ependyomomas this year. They analyzed 134 patients with spinal ependymomas over the last 25 years. Their conclusions are surprising to us. In their study, grade II ependymomas had significantly longer progression.
free interval than grade I ependymomas (14.9y versus 6y). In this study postoperative radiotherapy was performed in all partially removed GI ependymomas. These findings question the value of grading system of spinal ependymomas.

Conclusions:
We should reconsider our belief that GI ependymoma represent a benign disease provided that further studies confirm the conclusions of Tarapore’s study. Taking into account the conclusions of Tarapore’s study, we probably should have performed radiotherapy immediately after the surgery.

References: