Intramedullary Ependymoma with Associated Holocord Syringomyelia – A Rare Case with Limited Treatment Options

Friederike Sophie Fritzsche¹, Homajoun Maslehaty¹, Athanasios K. Petridis¹,²,³, Rashad El Habony⁴, Johannes Van de Nes⁵, Dukagjin Morina¹ and Martin Scholz¹

Abstract

Objective: We present a rare case of an intramedullary ependymoma WHO grade 2 presenting with holocord syringomyelia and discuss special features of adequate diagnosis and treatment options.

Case presentation: A 79 years old male patient presented with progressive gait ataxia and back pain. Previous operation of lumbar disc herniation and stenosis of the cervical spine did not improve the symptoms. Delayed MRI of the spinal cord showed extensive growth of an intradural mass with accompanied holocord edema. Biopsy of the intradural tumor at the level T9 revealed an Ependymoma WHO grade 2. Complete tumor resection was not possible; hence radiotherapy remained as the only adjuvant treatment option.

Conclusion: Diagnosis and adequate treatment of spinal ependymomas with accompanied holocord edema and for syringomyelia harbour some difficulties. The presented case shows a very rare clinical entity with distinct extension and associated severe progressive neurological deterioration. Microsurgical treatment techniques reached their limits in this case.

Keywords

Spinal ependymoma; Pan-spinal tumor; Holocord edema

Introduction

Spinal cord ependymomas are the most common spinal gliomas and occur most frequently in adults. The most affected level is the filum terminale followed the cervical spine. The incidence of longitudinally spinal tumors is less than 1% of all intramedullary lesions [1-3]. However, spinal cord ependymoma with accompanied holocord syringomyelia is a very rare clinical entity.

The aim of our study was to present such a rare case with an intramedullary ependymoma in the thoracic spine and discuss the clinical features, diagnostic findings and pitfalls, as well as treatment options.

Case Presentation

Patient history

A 79 years old male patient presented with progressive gait disturbance with recurrent fall incidents during the last five years. An operation of the lumbar disc herniation and stenosis of the cervical spine at the level C5/6 and 6/7 in an external clinic did not improve the symptoms. In the continuing course the mobilisation was possible to a limited extent with a wheeled walker for few meters.

In August 2011 MRI of the spine was performed in line with further aggravation of gait disturbance in terms of spinal ataxia, sensory disorders and progressive back pain, but without bladder dysfunctions.

MRI showed holocord edema, which was assumed to be the consequence of an intramedullary tumor. As differential diagnosis spinal vascular malformation, such as AV-fistula with venous congestion inside the spinal cord was discussed, which was ruled out by spinal DSA.

Repeated cranial and spinal MRI 11 months later revealed distinct progress of the spinal cord edema with extension from the medullary conus to the brainstem (Figures 1A and 1B). T2-weighted sequences showed the partly in homogen, hyperintense mass in the whole spinal cord from the cerebromedullar junction to the conus medullaris (Figure 1B). T1-weighted images with contrast showed a lack of contrast enhancement of the tumor. However, a slight contrast enhancement could be interpreted as tumor capsule (Figure 1C). The patient underwent detailed and extensive examination concerning a paraneoplastic origin of the lesion. Additional CT scan of the abdomen and thorax, as well as a lumbar puncture were without indicative findings. Therefore, the patient was admitted to our department for biopsy of the suspicious lesion.

Surgery

Biopsy was performed at level T9 because of the localized maximum extent of the tumor mass at this level. Under the microscope a midline incision of the dura was performed. The myelon appeared

Figure 1: MRI of the spinal cord; A: T2-weighted image shows the partly in homogen, hyperintense mass in the whole spinal cord from the cerebromedullar junction to the conus medullaris. B: MRI shows the progress of the mass in the spinal cord during 11 months 2011-08 (left bolt) until 2012-07(right bolt). C: T1-weighted sequence with contrast agent.
distended with cystic formations. Healthy tissue is not exactly definable. Intraoperatively, a clear and secure separation between normal cord and tumor mass was not possible (Figure 2). Due to the diffuse extension of the tumor, complete resection was not possible. Since the patient already suffered from massive gait disability but still intact bladder and rectal control, we decided to limit the procedure to a biopsy to obtain histological work-up.

Histopathology

Histopathological work-up revealed Ependymoma WHO grade 2. The tumor cells were GFAP and EMA positive, which signalizes reactive gliosis. The tumor-cells were not reacting on cytokeratines (Klon MNF116), synaptophysin or NeuN. P53-accumulation or IDH-1-R132H-mutation-specific reactions were not visible (Figure 3).

Postoperative course

This case was discussed extensively in our interdisciplinary tumor-conference. Since the patient suffered from severe gait disability but still had bladder and rectal control, we decided against gross tumor resection to prevent procedure related damage of the spinal cord. Chemotherapy was not an option, since there were no mitoses shown in the histological results. Radiotherapy of the spinal cord remained the only treatment possibility, which was refused by the patient at the moment and will be discussed again, if his present condition deteriorates.

Discussion

Since spinal ependymomas with accompanied holocord syringomyelia and edema are extremely rare findings, adequate diagnosis and therapy can be deceitful.

Diagnosis is usually made by MRI. However, correct allocation of the tumor might be difficult, due to heterogeneous contrast enhancement of the intramedullary mass. In the literature hypo- and hyperintense lesions in T1- and T2-weighted images are described as well [4]. Furthermore it is to differentiate between a widespread tumor in terms of a holocord tumor, or circumscribes tumor mass with accompanied holocord edema or syringomyelia.

Due to the heterogenous symptoms of spinal intramedullary lesions, holocord pathologies can lead to different clinical manifestations, which can complicate their exact attribution to the affected level. Considering the symptoms, Kucia and co-workers discussed 67 cases of spinal cord ependymomas and concluded the most common symptoms to be pain and dysesthesia, followed by weakness and numbness [4,5].

Considering the management strategy it is to decide between conservative and operative procedure. Aghakhani et al. postulate that surgery should be considered carefully in patients without significant neurological deterioration, to prevent postoperative aggravation of the clinical condition. The authors conclude that preoperative complaints improved in 30% of the cases, maintained in 60% and deteriorated in 10% [6]. In contrast to this opinion other authors prefer an aggressive surgical strategy to obtain complete tumor resection [7,8].

Boström et al. concluded in their study, that patients harbouring distinct spinal ependymomas should primarily been treated microsurgically [2]. Nagasawa et al. discussed the degree of resection as the most significant predictor of progression-free-survival. The authors conclude that total excision of the tumor can be performed in a curative manner and should be considered as the treatment of choice in those lesions [3].

However, holocord lesions are more specific and the surgical strategy is more complicated. Gunes and Ozdemir conclude that total resection of holocord tumors is very difficult and has been
described in only four cases in the present literature [4]. According to this, Pluchino et al. presented a case of holocord ependymoma, which could be removed completely with two surgical procedures [9]. However, aggressive surgical resection of large tumors is associated with a high overall complication rate and might lead to deterioration of the symptoms, though the overall outcome is excellent with low recurrence rate in small tumors, which has been resected completely [3]. In addition, resection of large tumors may become complicated by cord edema, arachnoid fibrosis or capillary neovascularization, which can lead to cord rotation or asymmetrical enlargement [3].

As an adjuvant therapy model, postoperative radiotherapy can be discussed which may improve the outcome of the patients with subtotal resected tumors [10]. Infrequently radiotherapy was able to decrease the rate of tumor progression in myxopapillary ependymomas, regardless of the extent of resection [3].

However, it should be noted that radiotherapy can lead to reactive gliosis and fibrosis, as well as progressive adhesion of the ependymoma to the spinal cord and destruction of microvasculature [3]. Furthermore the dose of radiation is discussed controversially [3,5].

Conclusion

Diagnosis and adequate treatment of spinal ependymomas with accompanied holocord edema and/or syringomyelia harbour some difficulties. The presented case shows a very rare clinical entity with distinct extension and associated severe progressive neurological deterioration. Microsurgical treatment techniques reached their limits in this case. Since complete tumor resection was not possible due to the extensive growth, we performed biopsy of the tumor tissue to obtain histological work-up. According to the results of the histopathology of an Ependymoma grade 2, adjuvant radiotherapy remained the only option, which was refused by patient.

References