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Technical Note & Surgical Technique

Primary spinal glioblastoma multiforme presenting with leptomeningeal gliomatosis and subarachnoid hemorrhage: A case report

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1. Introduction

De novo spinal glioblastoma multiforme (GBM) is a rare lesion of the central nervous system with a poor prognosis. It represents about 1 to 3% of all spinal cord tumours [1]. In >60% of cases Spinal GBM developed at the cervical or cardiothoracic region. [2]. Patients with these rare lesions may present with symptoms similar to more common neurosurgical conditions [3]. Leptomeningeal gliomatosis (LM) refer to the spread of malignant cells through the CSF space. High-grade glioma is the most common tumor to lead to LM in adults. Currently the incidence of LM can only be estimated [4]. Clinical presentation is varied, but most commonly includes headache, nausea and vomiting, back pain or radicular limb pain or sensory abnormalities and focal neurological deficits. Prognosis of LM is varying according to age and tumor type, is generally poor even with aggressive treatment [4,5].

Information regarding primary spinal GBM to understand this rare entity to guide the treatment is needed. We present a rare case of spinal GBM with leptomeningeal gliomatosis involving the Brainstem in a 39-year-old female.

2. Case report

A right-handed 39-year-old female was referred to our clinic for further clarification after diagnosis of subarachnoid hemorrhage (SAH) in the neurological department.

Initial symptoms were severe persistent pain in the neck and headache in the posterior side of the head for one week accompanied with nausea and vomiting. Medical history was non-contributory. Lumbar puncture was done in the neurological department before referral with 3 CSF-tubes with xanthochromic CSF. At admission in our clinic no neurological deficits were present.

At the admission, cerebral angiography, CT scan and MRI of the brain have been performed.

No abnormality has been detected except for not requiring therapy Isolate small developmental venous anomaly (DVA li.-frontal), otherwise unremarkable brain parenchyma.

Three days later, a new MRI 3-Tesla scan of the brain and total neural axis has been performed due to the worsen of the pain despite central analgesics, which demonstrated an unclear lesion intraspinal intradural with contrast enhancement in the level C4/5 ventro-lateral, T2/3 dorsolateral, L1/2 and L3 with diffuse leptomeningeal contrast until brainstem (pons) Fig. 1. Ophthalmological examination was normal.

Differential diagnoses were a granulomatous disease, neurosarcoidosis, tuberculosis, inflammatory process, lymphoma, or leptomeningeal carcinomatosis from an occult malignancy. Total body Scintigraphy, CSF diagnostic, standard tuberculosis and neurosarcoidosis diagnostics have been done. The only positive result was the protein in CSF which was 4467 mg/L. m. The number of cells was normal; no CSF cytological examination has been done. One week later the patient showed paralysis of the 6th cranial Nerve with deterioration of the orientation. A 3-Tesla MRI brain and total neural axis was performed again, it revealed extension of the previously described lesion especially in brain stem area, medulla oblongata and pons, no vertebral body enhancement or metastasis has been detected. An open Biopsy was done after laminectomy T2. Histopathology showed GBM with meningeal affection. The clinical situation had been dramatic worsened with blindness and paralysis in upper limb both side. Adjuvant therapy has been refused from the family due to sever disability and blindness. The patient died after 35 day from admission and 9 days after biopsy.

Histological examination: The tumor tissue showed all features of a glioblastoma with predominantly small tumor cells, endothelial proliferation, frequent mitotic figures and tumor necrosis. The tumor cells reacted positively with antibodies against GFAP, CD56, S100, and MAP2a, whereas EMA, IDH1, CD3, CD79a, CD99, AE1/AE3, MNF116, MelanA, Synaptophysin and TTF1 were not expressed. The tumor showed a high proliferation rate of 60% Ki67 positive cells Fig. 2.

3. Discussion

Spinal glioblastoma multiforme (GBM) is a rare clinical entity especially in adults. It accounts about 1–5% of all glioblastoma and about 1 to

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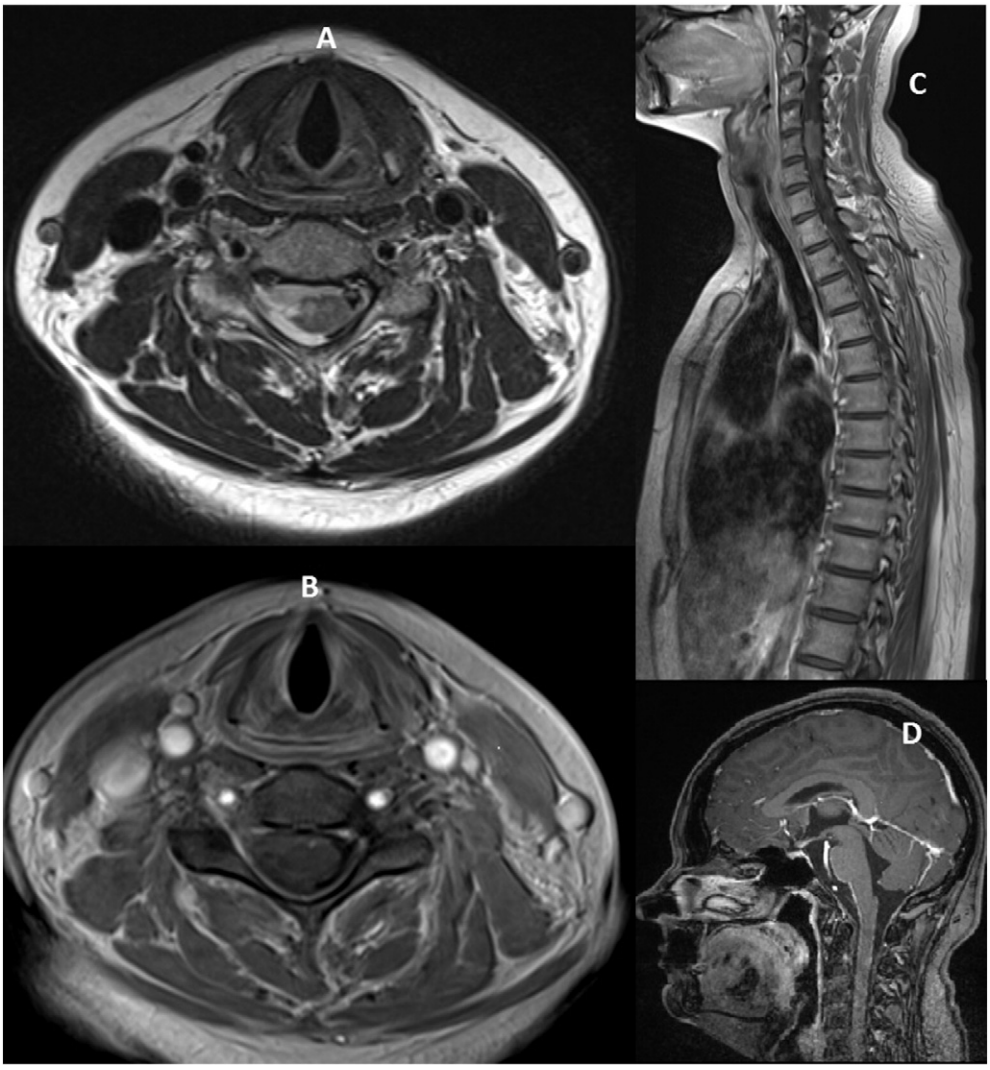

Fig. 1. A: MRI axial T2-weighted image at level C4 showing an intraparenchymal lesion with meningeal affection, B: T2 post-contrast axial T2-weighted image at level C4 showing a week enhancement with intradural intraparenchymal lesion and leptomeningeal enhancement. C: MRT sagittal T1-weighted image of the lumbar and thoracic region showed showing an intraparenchymal location at level C4 level, D: MRT of brain showing the leptomeningeal enhancement up to Brainstem.

3% of all spinal cord tumours. Available data is limited with few examples cited in a limited number of case reports [1,4].

Clinically our case reports exhibit points of interest regarding the site, clinical presentation, clinical course and radiological finding.

Regarding the site, our case showed lesions in cervical, thoracic and lumbar region. Published data showed that in >60% of cases Spinal GBM developed at the cervical or cervicothoracic region [2]. Spinal GBM is rarely in 3 regions in the same time.

Regarding to clinical presentation, our case presented with typical symptoms of SAH which laboratory confirmed. However patients with these rare lesions usually present with symptoms similar to more common neurosurgical conditions related to the site of the tumor [3]. Regarding the clinical course and radiological finding, our case showed at the time of diagnosis multiple foci of glioblastoma with Leptomeningeal gliomatosis seeding of a spinal cord to the brain, which usually appears late in the course of disease. It is rare form of spinal GBM.

The Leptomeningeal gliomatosis explain the initial symptoms of SAH in our case reported. In agreement with published data which showed that clinical presentation in case of LM is most commonly includes headache, nausea and vomiting, back pain or radicular limb pain or sensory abnormalities and focal neurological deficits.

The fatal outcome in our case with five weeks duration is in agreement with published data which showed that leptomeningeal metastases with CSF tumor dissemination almost always leading to a fatal outcome, most patients with lesions in the cervical or upper thoracic cord have significantly shorter survival [1,5].

Treatment of spinal GBM includes debunking of tumor and postoperative radiation with/without chemotherapy. Surgical management of an intramedullary mass consisted of biopsy, or cyst aspiration with decompressive laminectomy. Radical surgical removal in patients with intramedullary glioblastoma is impossible because there is not a clear cleavage plane between the glioblastoma and the medullary parenchyma.

The value of radiotherapy and chemotherapy in spinal cord GBM is a matter of debate due to the small number of cases reported in literature. However chemotherapy followed with radiotherapy is the most commonly used treatment modality in case of leptomeningeal metastasis [1,5]. Until now there is still no satisfactory treatment for spinal GBM and leptomeningeal metastasis. Limited data exist in literature to guide treatment.

4. Conclusion

Primary spinal GBM is an extremely rare entity with a poor prognosis and a short survival time. Improvements of current modes of treatment are required to improve survival and ensure better quality of life. Our

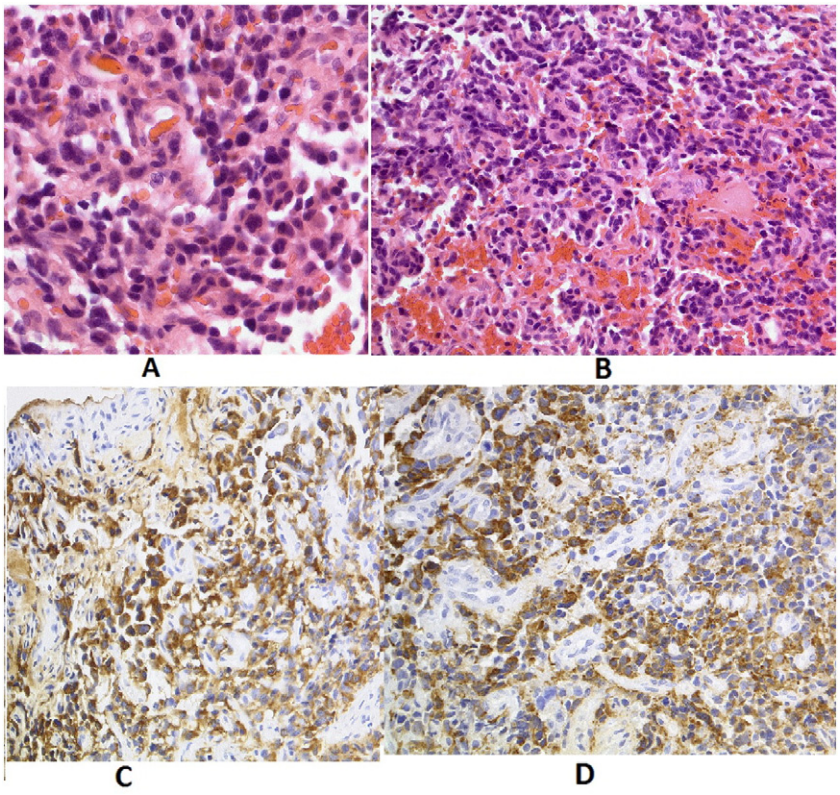

Fig. 2. A: The picture shows a malignant small cell tumor with necrosis on the left side and pathological vessels on the right side of the picture (HE, ×200 original magnification). B: malignant small cell tumor with two mitotic figures in the center of the picture is seen (HE, ×200 original magnification). C: positive Anti-GFAP reaction underlines the astrocytic line of differentiation (Anti-GFAP, ABC, ×200 original magnification). D: The tumor proliferates highly (Anti-Ki67, ABC, ×200 original magnification).

case report illustrates the importance of considering of spinal GBM as one of the differential diagnosis of SAH with brain negative angiography.

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