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Intracranial myopericitoma. A case report of a rare tumour in a rare location in an AIDS patient

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ABSTRACT

Myopericytoma is a benign, soft tissue tumor probably derived from perivascular myoid cells. They are usually found in subcutaneous tissues in the extremities. Intracranial localization of myopericytoma is exceedingly uncommon. We report a 43 years old male patient with incidentally found myopericytoma of the posterior fossa. Patient was operated and tumor was completely removed. Patient was subsequently diagnosed with acquired immunodeficiency syndrome due to human immunodeficiency virus infection. One year after operation tumor showed no signs of recurrence, but patient developed progressive symptoms of AIDS and started highly active antiretroviral therapy (HAART). Connection of intracranial myopericytoma appearance and HIV/AIDS has been reported before, but clear connection is yet to be elucidated.

INTRODUCTION

Myopericytoma represents a rare benign lesion most frequently found in soft tissue. However, it may arise in any other location. Clinical, neurological and radiological features are unspecific with symptoms arising due to compression of surrounding structures. Surgical resection is the treatment of choice with excellent prognosis. Pathology examination provides the definite diagnosis, but the differential diagnosis is wide. Intracranial location is exceedingly uncommon, and we found only several case reports described in the literature^{2,7}. Connection of intracranial myopericytoma appearance and human immunodeficiency virus infection has been reported before, but causal connection is yet to be elucidated¹.

We present a case of a 43 years old male with incidentally found myopericytoma of the posterior fossa. One year after the operation patient was diagnosed with acquired immunodeficiency syndrome due to human immunodeficiency virus infection.

Keywords

AIDS/HIV,
myopericitoma,
intracranial tumour



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CASE REPORT

We present a case of a 43 years old male gynecologist who was first examined 6 years ago because of typical migraine headaches, when brain computed tomography (CT) showed small dural based extra axial hyperdense lesion in the posterior region of foramen magnum without significant compressive effect, which was confirmed with brain magnetic resonance imaging (MRI). The patient was motivated for further intermittent follow-up and consecutive MRI showed no signs of lesion expansion and patient remain neurologically intact without symptoms and any complains for almost 6 years. However, patient skipped the last two follow-ups since he was clinically suspected for COVID-19 infectious disease, although three consecutive PCR tests were negative. Shortly after resolution of respiratory symptoms patient abruptly developed ataxia and gait disturbance with severe neck pain after which he was admitted in our emergency department and brain CT scan showed significant enlargement of extra axial lesion in the posterior cranial fossa with signs of cerebellar and fourth ventricle compression, but without signs of hemorrhage nor hydrocephalus (Figure 1).

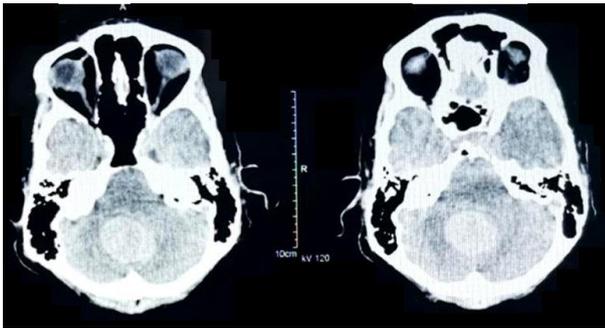


Figure 1. First brain CT after development of signs and symptoms of cerebellar dysfunction, showing tumor in the cerebellum.

Patient was transferred to Neurosurgical clinic of Clinical center of Serbia. At department admission he presented with cerebellar symptomatology including ataxia, asthenia and astasia-abasia complex without muscle weakness except of mild bilateral thigh flexors weakness. Also, global hyperreflexia with atypical plantar reflex bilaterally was noted. His past medical history was insignificant. Brain MRI showed large intradural extra-axial ovoid shaped tumor measuring 36 x 31 x 33 mm in diameter (LL x AP x CC) located at the posterior midline aspect of foramen

magnum and with signs of cerebellar and fourth ventricle compression (Figure 2). Patient was prepared for neurosurgical intervention, and MRI angiography was performed to visualize V3 and V4 segments of vertebral arteries, and fortunately they were distant from the tumor.

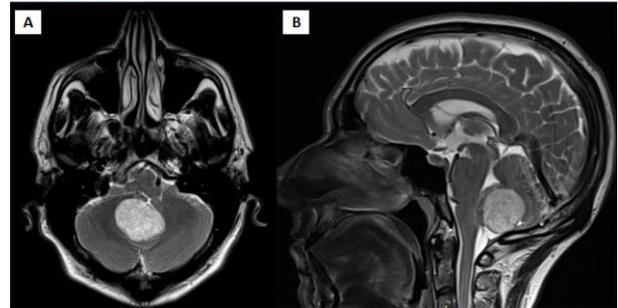


Figure 2. Brain MRI showing tumor in the posterior aspect of foramen magnum: (A) axial plane, (B) sagittal plane.

On the day of surgery, extremely rare situation occurred. Namely, four consecutive transfusion cross-matching blood tests using activated Papain showed positive inter-reaction. However, after our Blood Transfusion Institute provided enough units of blood, we proceeded with intervention, and the patient was operated in the sitting position. The skin was incised on the midline from the occipital protuberance down to the upper cervical region. The midline plane was opened between the posterior muscles, up to the occipital protuberance and down to the spinous process of C2. Bone opening was performed using a drill and Kerrison rongeurs. We decided to perform small right unilateral approach instead of large medial bone opening, since we wanted to avoid occipital sinus injury. Posterior margin of foramen magnum has been also removed. The dura was incised in a Y-shaped fashion and retracted with stitches. After dura opening and retraction of right cerebellar hemisphere, a tumor with insertion on the midline portion of dura was brought into view. The plan was to be perform tumor debulking with the ultrasound aspirator, however since tumor was highly vascularized, we decided to coagulate and liberate dural insertion, after which the tumor was removed in one piece (Figure 3).

Operation and early postoperative period were uneventful. Blood loss during operation was minimal, and there was no need for a blood transfusion. Patient recovered well, and preoperative cerebellar symptomatology was less

pronounced. Control brain CT showed complete tumor removal. Patient was discharged on the 4th postoperative day. The conjugation of morphological, histopathological and immunohistochemical studies allowed the final diagnosis of intracranial myopericytoma. After 6 months patient was without symptoms, and control brain CT showed no signs of tumor recurrence. However, one year from operation patient suffered rapid weight loss, several episodes of recurring fever and profuse night sweats with extreme and unexplained profound fatigue. After investigations patient was diagnosed with acquired immunodeficiency syndrome due to human immunodeficiency virus (HIV) infection. Patient recently started highly active antiretroviral therapy (HAART).

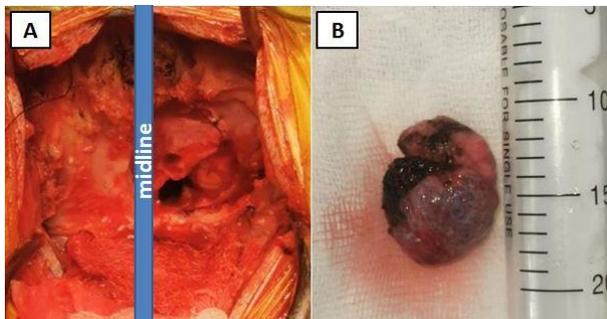


Figure 3. (A) Intraoperative image showing right unilateral approach during dura opening phase, (B) tumor after removal.

DISCUSSION

Myopericytoma is a benign tumor originating from the myoid perivascular cells⁶. It is often diagnosed in middle-aged patients, and most commonly arises in subcutaneous tissue of distal extremities or sometimes in retroperitoneal space. Granter et al. described the first case of myopericytoma and called it perivascular myoma³. According to current nomenclature, these tumors correspond to a continuum morphological spectrum, including myofibroma, myopericytoma and glomangiopericytoma⁵. First case of intracranial localization of myopericytoma was described by Rousseau et al. Since then only several cases of intracranial localization have been described⁸. We present a case of an intracranial myopericytoma with even more rare localization in the posterior cranial fossa. To our knowledge this is the second case of myopericytoma with posterior fossa localization.

In the described cases patients had a wide range of age, with slight female predominance⁷. Our patient is a 43 years old male patient. The histological findings were similar in all cases from the literature with tumor composed of large thick walls vessels with myxoid changes, lined by elliptical and oval/spindle endothelial cells⁷. Differential diagnoses included meningioma, solitary fibrous tumor/haemangiopericytoma and angioleiomyoma. However, in all cases, as well as in our patient, immunohistochemical studies confirmed diagnosis of myopericytoma.

Regarding tumor origin, some studies showed unusual molecular changes such as t(7;12)(p22;q13) and del(6)(q12q15)⁶. Also, some authors postulate a probable relation between Epstein-Barr infection in AIDS patients and myopericytoma development^{1,4}. Our patient was probably HIV positive many years before operation, and AIDS symptoms appeared soon after intracranial surgery. Although reports on this peculiar association have been described in the literature, more data are needed to clearly show a connection between these entities.

Intracranial myopericytoma is a low-grade slow growing tumor, with excellent overall survival. In all cases described in the literature only one patient died and it was due to other cause⁷. One year after the operation, our patient is without neurological symptoms and without signs of tumor recurrence. However, his general condition is worsening due to AIDS.

In conclusion, intracranial myopericytoma is a rare benign neoplasm, successfully treated by surgery and with excellent follow-up. Connection of intracranial myopericytoma appearance and HIV/AIDS is yet to be elucidated.

REFERENCES

1. Calderaro J, Polivka M, Gallien S, Bertheau P, Thiebault JB, Molina JM, Gray F. Multifocal Epstein Barr virus (EBV)-associated myopericytoma in a patient with AIDS. *Neuropathol Appl Neurobiol* 2008;34:115–7. doi: 10.1111/j.1365-2990.2006.00865.x.
2. Dray MS, McCarthy SW, Palmer AA, Bonar SF, Stalley PD, Marjoniemi V, Millar E, Scolyer RA. Myopericytoma: a unifying term for a spectrum of tumours that show overlapping features with myofibroma. A review of 14 cases. *J Clin Pathol*. 2006;59(1):67-73. doi: 10.1136/jcp.2005.028704.
3. Granter SR, Badizadegan K, Fletcher CD. Myofibromatosis in adults, glomangiopericytoma, and

- myopericytoma: a spectrum of tumors showing perivascular myoid differentiation. *Am J Surg Pathol.* 1998;22(5):513-25. doi: 10.1097/00000478-199805000-00001.
4. Lau PP, Wong OK, Lui PC, Cheung OY, Ho LC, Wong WC, To KF, Chan JK. Myopericytoma in patients with AIDS: a new class of Epstein-Barr virus-associated tumor. *Am J Surg Pathol.* 2009;33(11):1666-72. doi: 10.1097/PAS.0b013e3181aec307.
 5. Lindberg MR. *Diagnostic pathology of soft tissue tumors.* 2nd edn, 2015:344-5. Section 8.
 6. Mentzel T. Myopericytoma, including myofibroma: WHO classification of soft tissue and bone tumors. 4th edn, Lyon: IARC, 2013.
 7. Oliveira RC, Velasco R, Santos GR, Rebelo O. Intracranial myopericytoma: a tumour in a rare location. *BMJ Case Rep.* 2018;2018:bcr2017223111. doi:10.1136/bcr-2017-223111
 8. Rousseau A, Kujas M, van Effenterre R, Boch AL, Carpentier A, Leroy JP, Poirier J. Primary intracranial myopericytoma: report of three cases and review of the literature. *Neuropathol Appl Neurobiol.* 2005;31(6):641-8. doi: 10.1111/j.1365-2990.2005.00681.x.