ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

www.surgicalneurologyint.com

Surgical Neurology International

Editor-in-Chief: Nancy E. Epstein, MD, Clinical Professor of Neurological Surgery, School of Medicine, State U. of NY at Stony Brook.

SNI: Neuro-Oncology

Editor
Mitsutoshi Nakada, MD
Kanazawa University, Ishikawa, Japan

SNI
Open Access

Case Report

Cerebellar liponeurocytoma with an unusual metastatic CSF spinal seeding

Uri Pinchas Hadelsberg, Lea Kahanov, Andres Vargas

Department of Neurosurgery, Hadassah University Medical Center, Jerusalem, Israel.

E-mail: *Uri Pinchas Hadelsberg - urih85@gmail.com; Lea Kahanov - leyka10@gmail.com; Andres Vargas - andresvargas236@gmail.com

***Corresponding author:**
Uri Pinchas Hadelsberg,
Department of Neurosurgery,
Hadassah University Medical
Center, Jerusalem, Israel.

urih85@gmail.com

Received : 28 July 2021
Accepted : 19 January 2022
Published : 11 February 2022

DOI
10.25259/SNI_743_2021

Quick Response Code:

ABSTRACT

Background: Cerebellar liponeurocytoma is rare intracranial tumor appearing mostly in the posterior fossa.

Case description: We hereby report a long follow-up of a case of cerebellar liponeurocytoma in a 60-year-old female. At first, she presented in March of 2010 with the symptoms of hydrocephalus and was found to have a lesion located in the fourth ventricle. The tumor was resected with a small remnant around the brainstem which grew on serial imaging. Due to slow tumor growth, the patient was treated with conformal radiotherapy and was kept under follow-up with both outpatient visits and serial brain imaging. In 2018, due to low back pain and lumbar radicular pain, a new set of images of the spine was obtained which revealed multilevel intradural tumor spinal dissemination. The patient further underwent an open spinal biopsy at the level of L5 which revealed the same pathology of the intracranial tumor. The patient went on to receive total spine irradiation.

Conclusion: This case report describes a rare metastatic phenomenon to the spinal cord of the exact same pathology and grade of an intracranial cerebellar liponeurocytoma tumor.

Keywords: Cerebellar liponeurocytoma, CSF metastasis, Radiation

INTRODUCTION

Cerebellar liponeurocytoma is a relatively rare intracranial entity. Despite there being approximately 70 cases described in the medical literature of cerebellar liponeurocytomas and despite the pathology defined as a World Health Organization (WHO) Grade II tumor with a favorable prognosis, its natural history is not completely understood. Our case report describes an unusual characteristic of tumor aggressiveness hitherto unknown, showing its potential characteristic of leptomeningeal tumor dissemination, even without the development of malignant transformation per pathology report.

CASE PRESENTATION

We present a case of a cerebellar liponeurocytoma which initially presented in a 61-year-old healthy female. The patient was admitted to the neurosurgical ward in March of 2010. At that time, the patient was admitted to the ER with a month long of general weakness and deterioration. On the day of admission, she complained of dizziness and urge incontinence. The patient's physical examination was noticeable for a slight 4+5/5 right hand weakness and general weakness in her lower limbs as well as narrow-based walking. The patient underwent an initial CT scan and MRI

[Figure 1] which demonstrated obstructive hydrocephalus and a lesion involving the fourth ventricle. The patient underwent surgery through a suboccipital midline craniotomy in March 2010. The intraoperative pathology suggested a differential diagnosis between lymphoma, a glioma, and a round blue cell tumor, with a small residual left due to proximity to brainstem. The patient was discharged and did not appear in a timely fashion to her oncology follow-up. On follow-up imaging, a small residual part of the tumor was noticed that was not seen on the immediate postoperative MRI [Figure 2]. The patient's pathology report read "cerebellar liponeurocytoma with atypical morphological features." Of notice, the patient's first follow-up in the oncology clinic was in September the same year. Due to tumor growth, the patient underwent intensity-modulated radiation therapy to the residual tumor adjacent to the brainstem for a total of 54Gy with a KPS of 90 in May 2012.

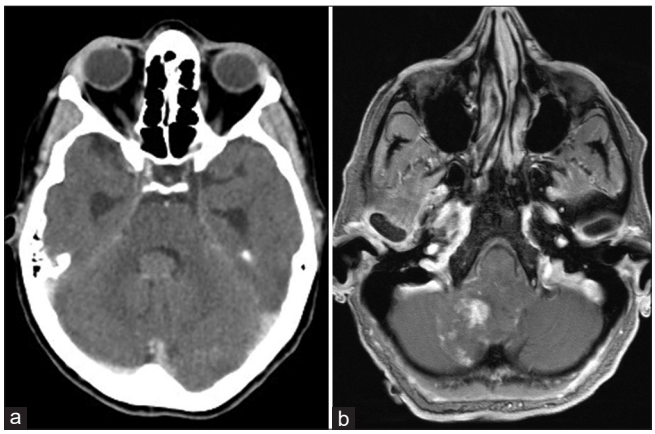

Figure 1: Axial CT and MRI. (a) A noncontrast CT representing hydrocephalus (note bilateral temporal horns) and a mass occupying the fourth ventricle. (b) An axial MRI with contrast demonstrating a paramedian hyperintense mass in the left cerebellar.

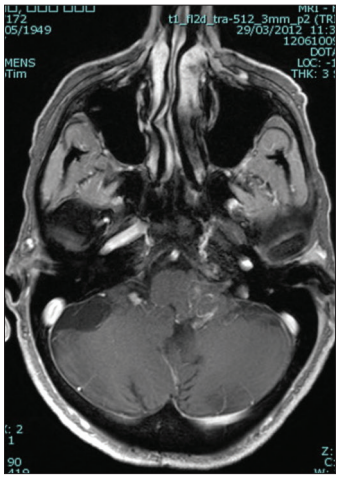

Figure 2: Axial MRI with contrast demonstrating a residual tumor in the tumor bed 3.29.2012 IM 10 SE 11.

The patient was followed up with serial MRI imaging being performed every 6 months. Due to new leg pain presented during follow-up, the patient underwent an FDG-PET in October 2017 scan to evaluate tumor metastases which demonstrated tumor spread to the cervical spinal canal. This prompted a total spine MRI [Figures 3 and 4] which revealed diffuse spinal cord involvement throughout the spinal cord from the cervical region to the cauda equina. The patient eventually underwent an open biopsy at the level of L5 was performed in January 2018 which demonstrated the exact same pathology seen in the previous surgery (without any pathological features suggestive of a malignant transformation). After reviewing the patient's charts during tumor board meetings, it was decided to treat in conservative fashion and postpone radiation or chemotherapy as she remained symptomatic, however, with

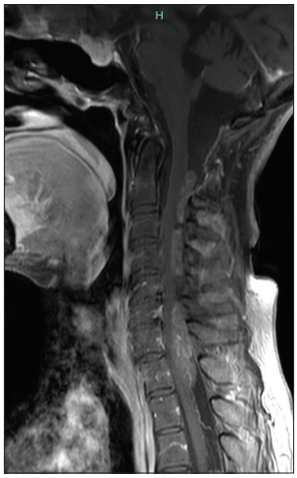

Figure 3: Sagittal cervical MRI with contrast showing diffuse tumor spread along the cervical region occupying the posterior portion of the canal with compression of the cervical spinal cord.

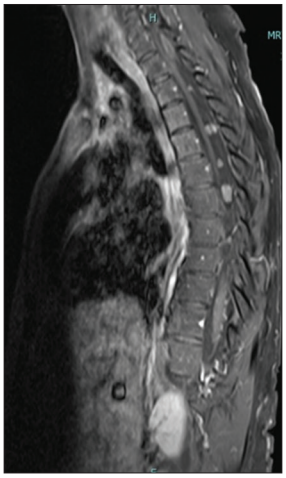

Figure 4: Sagittal thoracic MRI with contrast showing several lesions occupying the posterior part of the thoracic spinal cord. One can appreciate the lesion in the cervicothoracic spinal level as well as the T1 vertebral height.

a stable tumor burden. The patient's symptoms were minimal right leg weakness and decreased lower limb reflexes. The patient was stable during follow-up until June 2018 when she started having lower back pain with the right leg pain albeit stable imaging. Due to the deterioration of her leg symptoms, the patient underwent spinal radiation to L1-S2-3 of 30Gy in 10 fractions in July 2018. In October 2019, the patient complained of visual disturbances. Ophthalmologic examination from October 2019 revealed bitemporal hemianopsia and right eye visual deterioration with visual deterioration on follow-up. An MRI scheduled shortly after showed a new lesion in the suprasellar lesion [Figure 5]; however, due to her visual deterioration, the patient underwent radiosurgery in February 2020 to the chiasm of 30 Gy in 10 fractions.

In March 2021, the patient was seen during follow-up with stable disease both in her spine and her brain. Visual deterioration stopped after radiosurgery treatment. At present, the patient is at home with stable disease and no new neurological deterioration.

DISCUSSION

Regarding the pathology itself of a cerebellar liponeurocytoma, it was initially described as a WHO Grade I tumor. However, due to its substantial recurrences, the WHO assigned it a higher grade.[5,8] Indeed, cerebellar liponeurocytomas are rare tumors of the central nervous system (CNS), located mainly in the posterior fossa and first described by Bechtel *et al*. in 1978.[1] Historically speaking, cerebellar liponeurocytomas have only been described to occur in the posterior fossa, with reports only as late as one in 2009 describing a supratentorial origin.[1] These tumors have been reported at a wide range of ages (4–77 years) but seem to be encountered typically in adults of middle age.[4] In addition, according to Patel *et al*., there is no consensus regarding the treatment of liponeurocytoma, specifically whether chemo- or radiotherapy is a necessary part of the postoperative treatment regimen.[10]

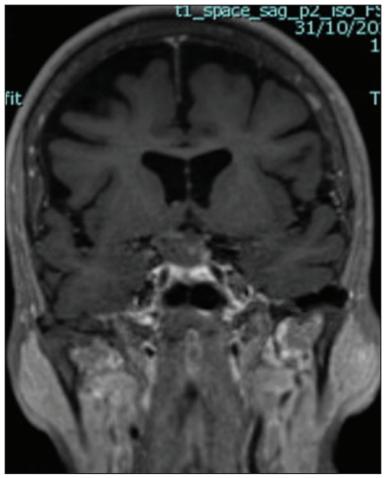

Figure 5: Coronal MRI with contrast done after radiosurgery to the suprasellar region demonstrating a suprasellar lesion.

In the pathology report, it was noted that the tumor had fragments of a neuroectodermal tumor with reactive gliosis with ill-defined neuronal rosettes and pseudorosettes. On immunostaining, the tumor stained positive for INI-1, S-100, GFAP, and vimentin, in accordance to the previous pathological reports of this tumor.[2,6] This tumor also displayed a high MIB-1 index (10–15%) in the pathological report.

In the most comprehensive review regarding these tumors, Oudrhiri *et al*. have noticed that only a bit more than 30 cases of this pathology were ever reported.[9] The authors mention the following locations of this tumor in the CNS: cerebellar hemisphere, vermis, cerebellopontine angle, and the fourth ventricle. There exists a rather recent report from 2021 by Hirono *et al*., depicting spinal dissemination from a liponeurocytoma originating from the floor of the fourth ventricle and lower vermis.[7] In this case, the patient presented with low back pain, gait disturbance, and unsteadiness. Specifically, the spinal dissemination was small multifocal thoracic metastases, with the patient ongoing a subtotal intracranial resection.

This patient received radiation to the residual tumor in the brainstem. Residual tumor left after surgery is a feasible radiosurgery target and is a practice advocated by some authors.[3] Moreover, Gembruch *et al*., in their literature review of the treatment of liponeurocytoma, mentioned that postoperative radiotherapy seems to decrease the risk of tumor recurrence.[5] They have performed a systematic review of the literature and found 73 patients with liponeurocytoma. Of those who were treated with adjuvant radiotherapy, only one patient had recurrence versus 13 patients out of 29 patients which were not treated with radiotherapy postoperatively.

CONCLUSION

Much about this type of tumor remains unknown; however, some have already suggested that a high MIB-1 index and incomplete tumor resection might represent adverse prognostic factors in patients with liponeurocytoma.[11]

Our take-home message from this case is the potential feature of seeding metastasis in cerebellar liponeurocytomas and emphasizing that total tumor resection is superior in outcome compared to subtotal resection. Radiotherapy should be offered to patients, especially in the event of a subtotal resection.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Bechtel JT, Patton JM, Takei Y. Mixed mesenchymal and neuroectodermal tumor of the cerebellum. Acta Neuropathol 1978;41:261-3.
2. Brat DJ, Parisi JE, Kleinschmidt-DeMasters BK, Yachnis AT, Montine TJ, *et al*. Surgical neuropathology update: A review of changes introduced by the WHO classification of tumours of the central nervous system, 4th edition. Arch Pathol Lab Med 2008;132:993-1007.
3. Cacciola F, Conti R, Taddei GL, Buccolieor AM, di Lorenz N. Cerebellar liponeurocytoma. Case report with considerations on prognosis and management. Acta Neurochir (Wien) 2002;144:829-33.
4. Gembruch O, Junker A, Ahmadipour Y, Sure U, Lemonas E. Cerebellar liponeurocytoma-a rare entity: A case report. J Med Case Rep 2018;12:170.
5. Gembruch O, Junker A, Monninghoff C, Ahmadipour Y, Oppong MD, Sure U, *et al*. Liponeurocytoma: Systematic review of a rare entity. World Neurosurg 2018;120:214-33.
6. George DH, Scheithauer BW. Central liponeurocytoma. Am J Surg Pathol 2001;25:1551-5.
7. Hirono S, Gao Y, Matsutani T, Ikeda J, Yokoo H, Iwadate Y. Metabolic, immunohistochemical, and genetic profiling of a cerebellar liponeurocytoma with spinal dissemination: A case report and review of the literature. Brain Tumor Pathol 2021;38:257-62.
8. Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, Burger PC, Jouvet A, *et al*. The 2007 WHO classification of tumours of the central nervous system. Acta Neuropathol 2007;114:97-109.
9. Oudrhiri MY, Raouzi N, El Kacemi I, El Fatemi N, Gana R, Maaqili MR, *et al*. Understanding cerebellar liponeurocytomas: Case report and literature review. Case Rep Neurol Med 2014;2014:186826.
10. Patel N, Fallah A, Provias J, Kha NK. Cerebellar liponeurocytoma. Can J Surg 2009;52:E117.
11. Xu L, Du J, Wang J, Fang J, Liu Z, He Y, *et al*. The clinicopathological features of liponeurocytoma. Brain Tumor Pathol 2017;34:28-35.

How to cite this article: Hadelsberg UP, Kahanov L, Vargas A. Cerebellar liponeurocytoma with an unusual metastatic CSF spinal seeding. Surg Neurol Int 2022;13:45.