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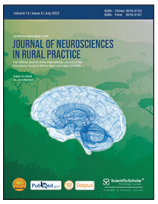

Letter to Editor

Multifocal cerebellar liponeurocytoma: How to deal with it?

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Dear Editor,

Liponeurocytoma is a rare grade II glioneuronal tumor of the central nervous system that mostly occurs in middle-aged adults and is located in the cerebellar hemisphere.[1] Multifocal cerebellar liponeurocytoma is extremely rare and their management is still the subject of controversy. To the best of our knowledge, only 5 cases have been reported in the literature to date. Herein, we present our experience in the management of two cases.

Case 1. A 59-year-old female with a history of diabetes mellitus presented with progressive headaches and gait disturbance evolving for 6 months. Her neurological examination revealed a normal consciousness, a severe cerebellar ataxia, and a left-sided dysmetria. A brain computed tomography (CT) scan was

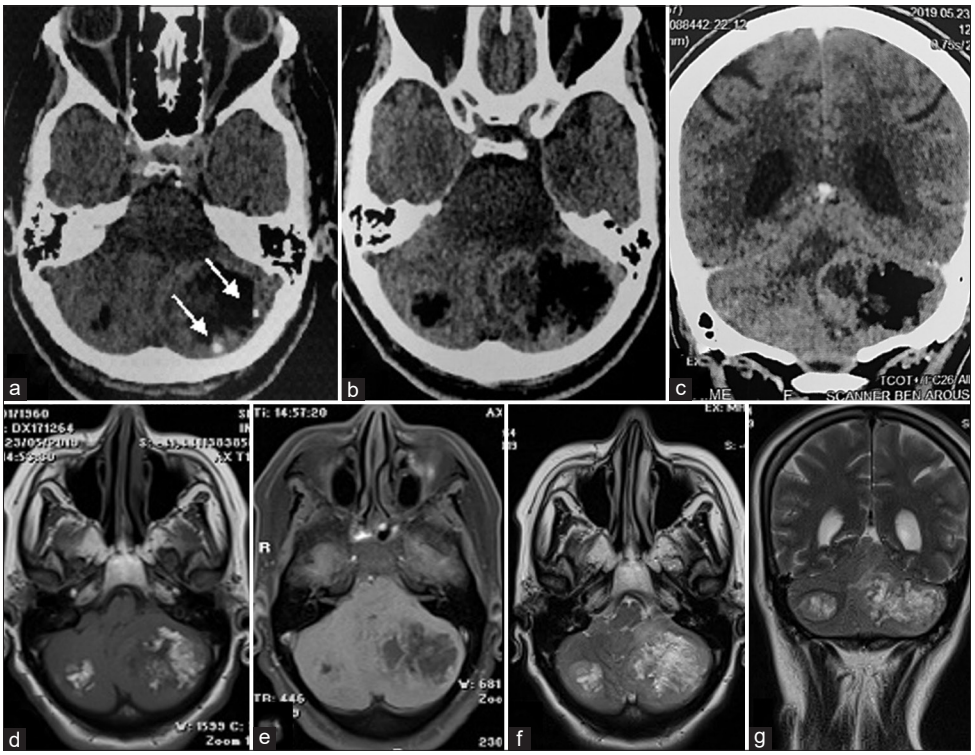

Figure 1: (a and b) Computed tomography scan on axial and (c) coronal sections showing two cerebellar bilateral lesions having the same appearance but the one in the left hemisphere is much larger. The tumor presents an irregular hypodense center which corresponds to a fat component and a peripheral solid isodense part, containing small calcifications (arrows). (d) The fat component is hyperintense on magnetic resonance images T1-weighted images, (e) gets inverted in fat-suppressed images, and (f and g) is hyperintense on T2-weighted images.

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performed, revealing two cerebellar masses: the largest one was located in the left hemisphere, measuring 50 × 39 mm, while a smaller lesion was located in the right cerebellar hemisphere, measuring 23 × 18 mm. Both lesions were formed by a large hypodense fatty center and a peripheral solid compartment with a heterogeneous contrast enhancement. Peripheral calcifications were noticed in the left lesion. On magnetic resonance images (MRI), the fat component was hyperintense on T1- and T2-weighted images and gets inverted in fat-suppressed images [Figure 1]. The patient was operated on through a left-sided suboccipital craniotomy to remove the largest lesion since it was the symptomatic and life-threatening one. During surgery, the lesion was yellowish, soft, mildly vascularized, and well-circumscribed [Figure 2]. A gross total resection was achieved. The post-operative course was uneventful. Histopathological analyses showed neurocytic neoplastic cells containing focal areas of lipomatous differentiation. The Ki-67 labeling index was <1% [Figure 3]. No adjuvant therapy was undertaken. Brain and spine MRI scan performed 1 year after surgery showed no evidence of recurrence of the resected lesion nor the appearance of new lesions and the right cerebellar tumor remained unchanged [Figure 4]. The last routine post-operative follow-up was at 25 months.

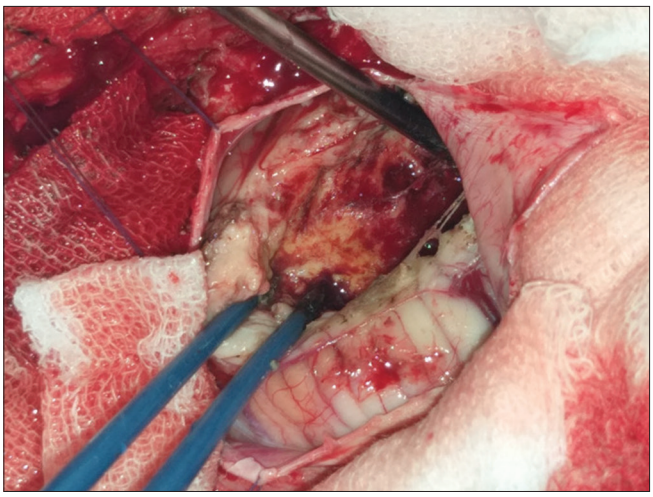

Figure 2: Perioperative microphotograph representing the yellowish fat tumoral tissue during resection through a left cerebellar cortectomy.

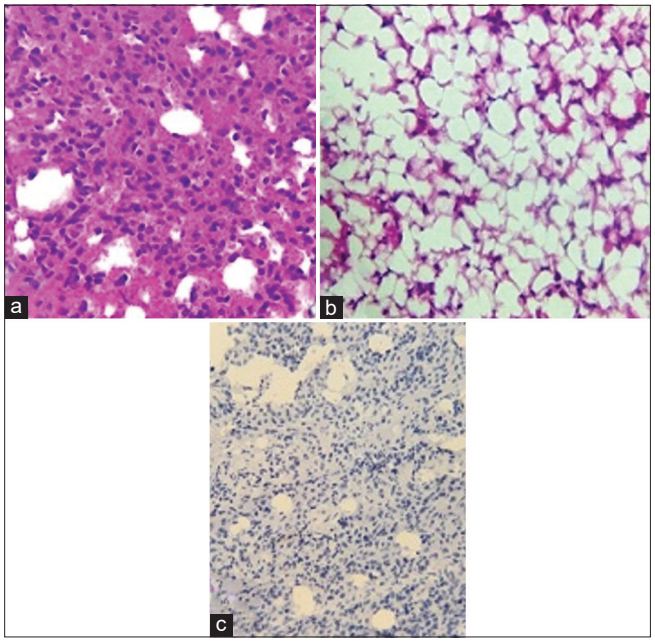

Figure 3: Histopathological examination. (a) Hematoxylin and Eosin (H and E) × 20: Uniform population of small round neurocytic cells arranged in sheets. (b) (H and E) × 20: Lipomatous differentiation with accumulation of adipocytic cells. (c) Ki-67 immunohistochemical staining showing a low proliferative index.

Case 2. A 68-year-old female complained of a 5 months' history of dizziness and gait disturbance. In her medical history, she had a transient ischemic stroke 10 years ago during which two cerebellar masses have been incidentally discovered. Physical examination revealed only cerebellar ataxia. A brain MRI was performed, revealing a large vermian solid cystic tumor measuring 54 mm in largest diameter, and a second small tumor located in the left cerebellar hemisphere. This latter remained grossly identic comparing to the first

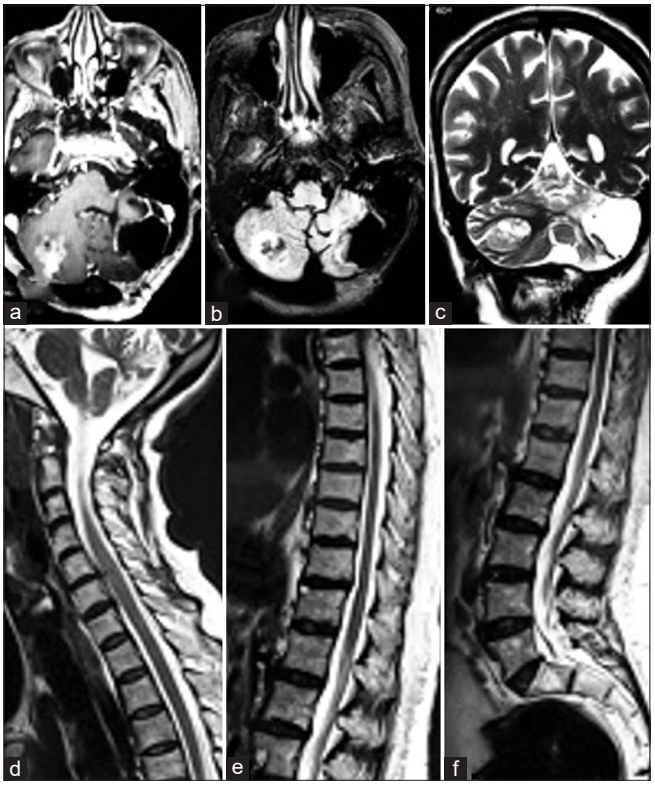

Figure 4: Brain magnetic resonance images (MRI) performed 1 year after surgery in axial T1 with (a) Gadolinium sequence, (b) FLAIR, and (c) coronal T2 image showing no recurrence nor evolution of the non-resected lesion. (d-f) Whole spinal MRI showed no evidence of metastatic tumor in the neuraxis.

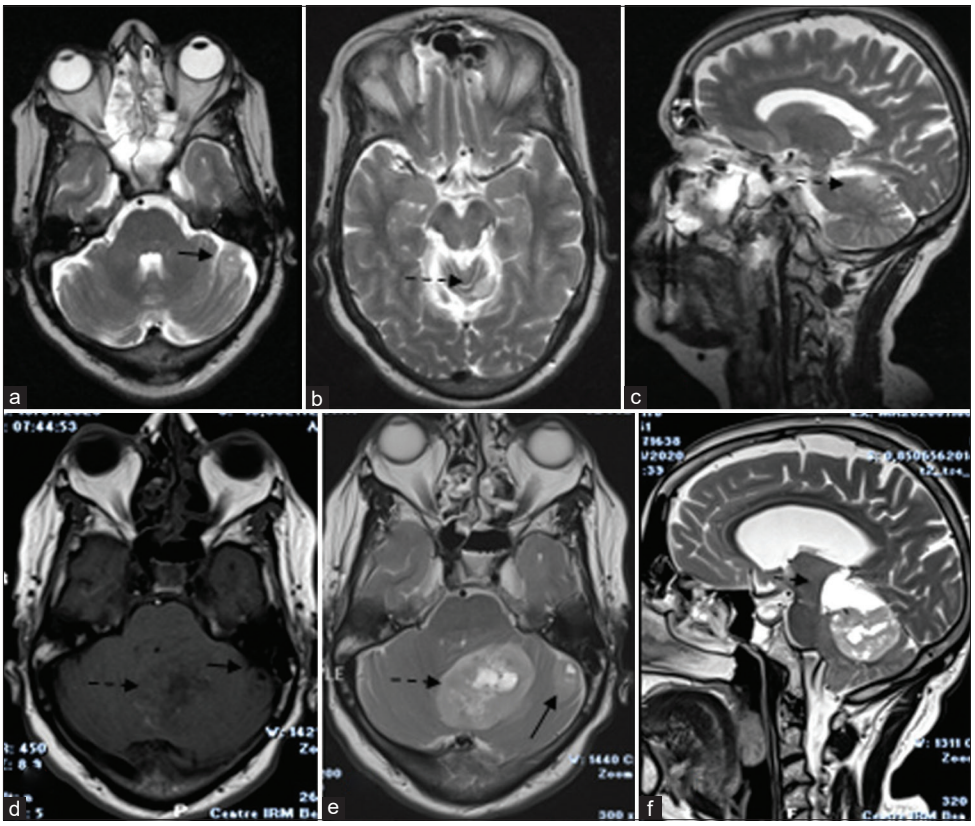

Figure 5: Brain magnetic resonance images (MRI) of the second case. (a and b) T2-weighted images in axial and (c) sagittal section of the first scan 10 years ago performed for a transient ischemic stroke, showing an incidental finding of two cerebellar masses, one in the upper vermis (dash arrow) and the second in the left hemisphere (arrow). Pre-operative MRI: (d) Axial T1 (e) axial T2 and (f) sagittal T2 images showed important enlargement of the vermian mass (dash arrow), while the left hemispheric one did not significantly grow (arrow).

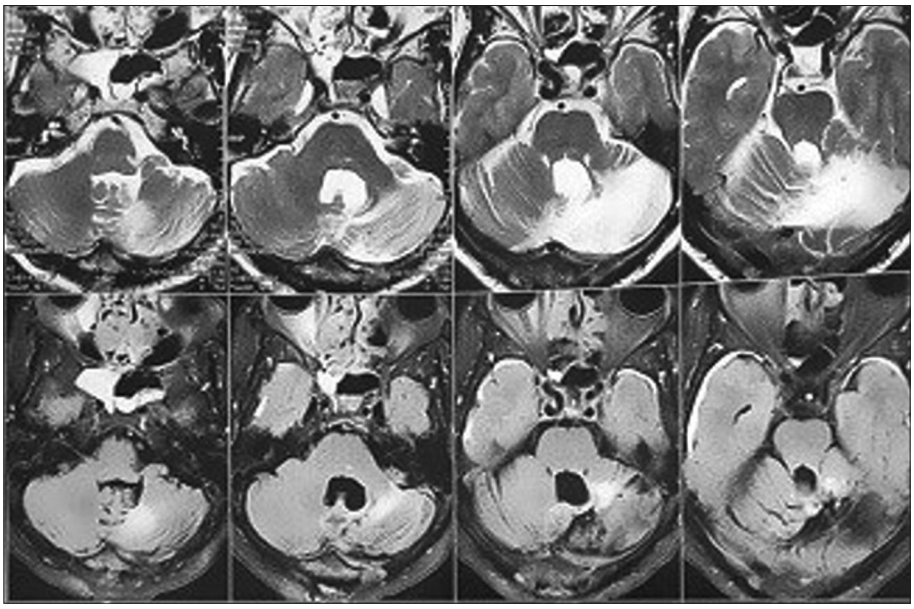

Figure 6: Post-operative brain magnetic resonance images of case 2. Axial T2-weighted images (top row) and FLAIR images (bottom row) show no recurrence nor residual tumor.

Table 1: Cases of multifocal cerebellar liponeurocytomas reported in the literature.

Author/year	Age(year)/ Sex	Location	Treatment-adjuvant therapy	Proliferative index	Follow-up/recurrence
Pelz *et al.*[11] 2013	54/F	- The largest lesion in the fourth ventricle extending to C1 level - The second lesion in the right cerebellar hemisphere laterally	Subtotal resection of the tumor through a suboccipital craniotomy	Ki-67 low	• MRI at 2 years after surgery showed enlargement of a residual nodule: surgical removal • 2 years later, a second recurrence and a new left cerebellar nodule: radiation proposed but declined by the patient
Dhar *et al.*[12] 2015	49/F	- The largest lesion in the left CPA - The second lesion in right cerebellar hemisphere	Excision of the left-sided lesion through a left retro mastoid suboccipital craniectomy	<1%	Not reported
Scoppetta *et al.*[13] 2015	42/M	- The largest lesion in the left CPA - The second lesion in the right cerebellar hemisphere	Biopsy Conservative	Not reported	Not reported
Sivaraju *et al.*[6] 2017	37/M	- The largest lesion in the vermis with extension to the left hemisphere - Multiple other lesions in both cerebellar hemispheres	Near total excision of the largest lesion, Radiotherapy 60 Gy	MIB 6%	MRI at 2 years after surgery showed no recurrence, the remaining lesions remained the same, no additional lesions
Khatri *et al.*[7] 2018	36/F	- The largest lesion in the left cerebellar hemisphere - The second lesion in the right cerebellar hemisphere	Gross total resection of the largest tumor through a left paramedian sub-occipital craniectomy	Ki-67 low	MRI at 8 months postoperatively: No residual tumor nor recurrence. The other lesion did not change. A new tumor appeared in the right cerebellar hemisphere: radiotherapy
Present first case	59/F	- The largest lesion in the left cerebellum - The second lesion in the right cerebellum	Total resection of the largest tumor through a left paramedian sub-occipital craniectomy	Ki-67 <1%	MRI at 1 year after surgery: No recurrence of the resected large lesion, no changes in the contralateral small lesion
Present second case	68/F	- The largest lesion in the vermis and the left cerebellar hemisphere - The second lesion in the left cerebellar hemisphere laterally	Total excision of both lesions through a left paramedian suboccipital craniotomy	Ki-67 <1%	CT scan at 3 months after surgery: no residual tumor

CPA: Cerebellopontine angle, MRI: Magnetic resonance images, CT: Computed tomography

MRI undertaken 10 years ago [Figure 5]. The spinal MRI was normal. The patient underwent a surgical removal of both lesions through a large left suboccipital craniotomy. The solid part of the large lesion was soft, yellowish, slightly bleeding, and had an ill-defined cleavage plane. A gross total resection was achieved. The post-operative course was uneventful. Histopathology analyses were consistent with a bifocal cerebellar liponeurocytoma with Ki-67 labeling index <1%. Radiotherapy was not indicated on the neuro-oncological board. A follow-up MRI was performed 1 year after surgery, showing no evidence of tumor recurrence [Figure 6]. The last post-operative follow-up was at 14 months.

Liponeurocytoma is a grade II glioneuronal tumor.[2] At first reports, liponeurocytoma appeared to be specifically located in the posterior fossa[3] and was described predominantly in cerebellar hemispheres.[1,4,5] In 2018, Gembruch *et al.*[1] reviewed all reported cases of cerebellar liponeurocytoma from 1978 to 2018 and identified only 73 cases. The posterior fossa was the affected region in 80.8% of cases, while supratentorial localizations accounted for only 19.2% of cases. Tumor occurs mostly in adults between the fourth and the fifth decades with a slight female predominance (40 females and 33 males). Diagnosis of cerebellar liponeurocytoma is suspected on brain imaging when evidence of fat tissue is

found: areas that are hypodense on CT scan and hyperintense on T1-weighted images on MRI. Histopathologically, liponeurocytoma is characterized by a mixed neuronal and astrocytic differentiation with foci of lipomatous differentiation.[1] Immunohistochemical staining reveals both neural and glial differentiation.[1] Mitosis, vascular hyperplasia, or necrosis are rarely found.[5] Proliferative index Ki-67 is usually below 5%.[4-6] Hence, liponeurocytoma is considered as a benign and slow-growing tumor.

Surgical resection represents the most effective treatment of cerebellar liponeurocytoma.[1,3,7] Total resection of the tumor is associated with a good outcome. Residual tumor and a high Ki-67 index are considered as risk factors for recurrence.[3] Systematic post-operative radiotherapy is not recommended following a gross total resection and should be reserved only for recurrences and tumors expressing a high Ki-67 index.[5,8,9] Recurrence occurs after a mean interval of 8.5 years;[3] thus, follow-up periods should be prolongated.[8] Multifocality is exceedingly rare in liponeurocytoma. To the best of our knowledge, only 5 cases have been previously reported. In all cases, tumors were located in the posterior fossa, bilateral, or in the same cerebellar hemisphere [Table 1]. Whenever possible, total surgical removal should be the gold standard treatment in case of multifocal cerebellar liponeurocytoma. Even in multifocal lesions, we consider that post-operative radiation should be reserved only for cases already operated on with a recurrent lesion and a high proliferative index. Spinal MRI is recommended in patients with liponeurocytoma, as lumbar localization has been described in the case of Anghileri *et al*.,[10] occurring 11 years after posterior fossa tumor resection.

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Declaration of patient consent

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Conflicts of interest

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Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The author(s) confirms that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using the AI.

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