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Case Report

Cerebellar liponeurocytoma: Rare posterior fossa tumor ☆

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ABSTRACT

Cerebellar liponeurocytoma is a rare WHO grade II glioneuronal tumor, characterized by a benign course and a more favorable prognosis. In this paper, we report a 52-year-old man, hospitalized in emergency because of clinical signs of increased intracranial pressure, and gait disturbances. The MRI revealed a lateral well shaped solid lesion within the left cerebellar hemisphere, having a low signal intensity compared to the cerebrospinal fluid, and causing a triventicular hydrocephalus and tonsillar herniation. The patient underwent a direct approach of the tumor through a left suboccipital craniectomy. Total removal of the lesion was achieved, and the histological diagnosis was cerebellar liponeurocytoma. The tumor cell proliferation index was < 6 %, therefore, radiotherapy and chemotherapy were not implemented after surgery. After 5 years of close follow-up there were no clinical or radiological signs of recurrence.

Our objective in reporting another case of this unusual tumor is to discuss clinical profile, radiologic and histologic features as well as treatment and prognosis of this rare posterior fossa tumor.

Introduction

Cerebellar liponeurocytoma is a rare glioneuronal tumor first described in 1978. Since then, some cases were published under different names as lipomatous medulloblastoma, medullocytoma, neurolipocytoma, lipomatous glioneurocytoma, and lipidized mature neuroectodermal tumor of the cerebellum [1]. These different names are in fact due to the histological similarities between liponeurocytoma and medulloblastoma. However, there is a genetic difference between the two tumors which has led The World Health Organization (WHO) to recognize this rare tumor as a distinctive grade I entity according to the 2000 WHO classification

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of Central Nervous System (CNS) tumors. In the 2016 WHO classification, the tumor was upgraded to grade II because the long-term follow-up studies have shown a higher recurrence rate estimated to 60%, with even aggressive behavior for some cases [2,3].

This report highlights the clinical aspects, radiological and histological features as well as therapeutic strategy of this rare posterior fossa tumor.

Case report

Signed written consent has been obtained from the patient to report the case in a scientific journal, which is available on request.

A 52-year-old right-handed man, with unremarkable past medical history, presented to the emergency department complaining of progressive severe headaches, nausea, vomiting, and gait disturbances of four months duration. Two weeks prior to this visit, he began to notice marked ataxia. However, he didn't report any dysphonia, change swallowing, weakness, or sensory disturbance problems to exclude damage to the brain stem or cranial nerves.

Neurological examination showed a conscious patient having kinetic cerebellar syndrome associated with a slight bilateral horizontal nystagmus. The ophthalmological examination revealed a reduced visual acuity to 7/10 in the right eye and 6/10 in the left eye with a bilateral stage II papillar oedema at funduscopy.

Cerebral CT scan revealed well defined hypoattentuating mass involving left cerebellar hemisphere, enhancing heterogeneously with contrast. MRI showed a lateral well shaped solid lesion within the left cerebellar hemisphere, which was isointense on T1- weighted imaging (WI) (Fig. 1A), slightly hyperintense on T2-WI compared with the cortex (Fig. 1B), with focal areas of hyperintesity on both T1 and T2-WI. After administration of contrast agent, the tumor showed heterogeneous and minimal enhancement (Fig. 1C). Moderate mass effect was evident, resulting in tonsillar herniation into the foramen magnum and compression of the fourth ventricle with active triventricular hydrocephalus, this aspect suggested a liponeurocytoma or a medulloblastoma.

The patient underwent surgery in the prone position through a left suboccipital craniectomy. After dural opening and corticectomy, we discovered a cerebellar grey-reddish hemorrhagic tumor with bright yellow areas. Thanks to the existence of a plane of cleavage between the tumor and the surrounding cerebellar parenchyma, total removal of the lesion was achieved. The postoperative course was uneventful and the patient was discharged four days later.

Histological study revealed that the tumor was composed of closely packed round to oval cells having irregular nuclei with clumps of heterochromatin. Lipid accumulation was a prominent feature of neoplastic cells pointing out the diagnosis of neurolipocytoma (Fig. 2). Immunohistochemistry was positive for synaptophysin, GFAP (glial fibrillary acidic protein), and desmin.

The tumor cell proliferation index (MIB-1) was < 5 %, The proliferative activity of tumors can be assessed using the MIB1 antibody to detect the Ki-67 nuclear antigen, which is associated with cell proliferation. Therefore, the patient didn't receive adjuvant radiotherapy. He has been monitored radiologically by MRI every 03 months for two years and every 6 months for 03 years with no signs of recurrence.

Discussion

Liponeurocytoma is defined by the WHO as a rare, well-differentiated neurocytic tumor of the cerebellum with local or regional lipomatous differentiation. It is distinct from medulloblastoma due to its low growth potential and more favorable prognosis. [4].

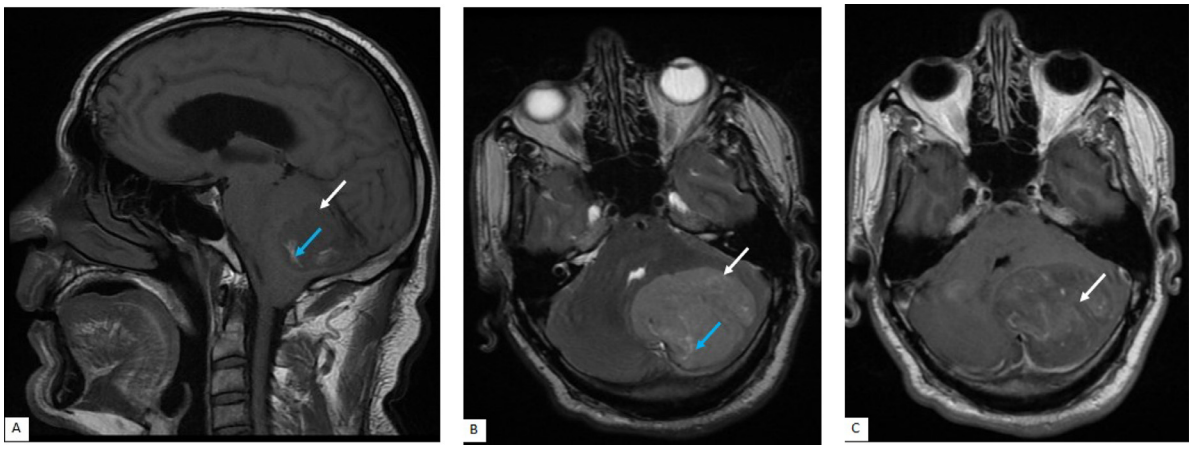

Fig. 1 – Brain MR images: Sagittal T1-WI (A), Axial T2-WI (B), Axial post contrast T1-WI, revealing a lateral well-shaped solid lesion within the left cerebellar hemisphere without apparent edema, isointense on T1-WI (A), slightly hyperintense on T2-WI (B) (white arrows), with focal areas of hyperintesity on both T1 and T2-WI (blue arrows). After administration of contrast agent (C), the tumor showed heterogeneous and minimal enhancement (white arrow). Moderate mass effect was evident, resulting in tonsillar herniation into the foramen magnum and compression of the fourth ventricle with active triventricular hydrocephalus.

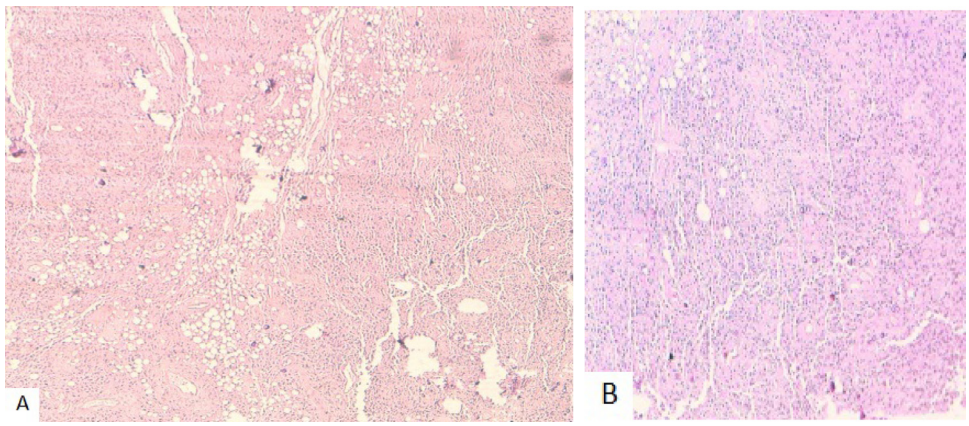

Fig. 2 – Histological examination by hematoxylin and eosin (HE) staining Shows closely packed round to oval cells having irregular nuclei with a high nuclear to cytoplasmic ratio (A), and prominent areas of lipidisation (B).

Topography of liponeurocytoma is controversial, although the majority of cases reported in the literature have been located in the cerebellar zone, some recent studies have suggested that the tumor can be localized supratentorially [5], particularly in the lateral ventricle [6].

All reported cases of liponeurocytoma have occurred in middle-aged adults (fourth and fifth decades),with mean age of 51 years (range from 30 to77 years) [7,8]. That is in contrast to classic medulloblastoma, which typically affects children and young adults. Men and women are affected equally, according to the study by Patel et al., in which 42 cases of liponeurocytoma from the literature were reviewed in 2009 [8].

The symptomatic period prior to presentation is generally long and the tumor often grows to over 4 cm in size before diagnosis. Clinical symptoms are often related to raised intracranial pressure and include headache, vomiting, nausea, visual symptoms and altered consciousness [9], our patient fit this clinical profile. Other manifestations may be seen including dizziness, ataxia with unsteadiness and frequent falls, and other signs of cerebellar or brainstem dysfunction.

Radiologically, intracranial liponeurocytoma most often presented as a well circumscribed solid mass. On CT scans, it may appear isodense or hypodense with focal areas of marked hypodensity corresponding to fat density and a minimal perilesional oedema but a clear mass effect to the adjacent structures [9–11]. Parenchymal cysts and/or associated cerebellar hemorrhage were also described [9]. This tumor is usually unique, rarely multicentric [5].

MRI is the neuroimaging exam of choice. On non contrast T1-weighted sequences, the tumor is generally iso-intense to hypo intense, with patchy areas of hyper intensity corresponding to regions of high lipid content which may be very suggestive [8,11].

On T2-weighted magnetic resonance images, the tumor appears slightly hyper-intense to the surrounding brain, with focal areas of more pronounced hyper intensity corresponding to fat. Some cystic components may be present, but usually peri-tumoral oedema is absent or minimal. After administration of contrast agent, the tumor shows usually heterogeneous and minimal enhancement [8,11].

In our patient, this was the case, and this imaging characteristic might be considered reliable, although there is no specific radiological behavior.

The histopathological origin of this tumor is still controversial. Some authors suggest that neoplasm evolved by progressive lipidization of tumor cells, probably due to degenerative or metabolic changes. Others advocate a metaplastic transformation of neuroectodermal cells into fat cells [8].

On the histological study, cerebellar LNC is a glioneuronal tumor with lipidized neoplastic cells, which are scattered throughout the tumor. Areas of neuronal differentiation express neuronal markers such as Syn, NSE, and MAP-2. Reactivity for GFAP was also described and supports the theory of neuroectodermal origin [12,13]. In this case, histological study revealed round to oval cells having with irregular nuclei. Lipid accumulation was a prominent feature of neoplastic cells. Immunohistochemistry was positive for synaptophysin, GFAP (glial fibrillary acidic protein), and desmin which is consistent with the literature data concerning histological features of cerebellar liponeurocytoma.

The absence of mitoses, necrosis, vascular hyperplasia, and nucleo-cytoplasmic atypia, and the low proliferation index (MIB-1 $<$ 5%) in most published cases, are histological arguments in favor of benign tumor. However, many recent publications report cases with relatively high levels of Ki-67 with cytologic atypia and which were associated with high recurrence rates and more aggressive behavior [13,14].

Regarding therapeutic management, surgery with total resection represents the cornerstone of treatment. The tumor demonstrate a favorable course. Without adjunctive therapy, long-term survival is expected [5]. However, a close follow up is mandatory because the tumor has a recurrent nature, although this occurs in the long term, with an average recurrence time of around 8.5 years from 20% to 32% of the patients as it was reported by Limaiem et al. [15].

There is no evidence to support the use of adjuvant radiation in the prevention of recurrence and because of the side effects of radiation, the majority of studies in the literature recommended gross, quality-of-life total resection and long-term monitoring plan [1,16].

On the other hand, there is no consensus to date concerning the usefulness of adjuvant radiation in the prevention of recurrence [1,6,17], that is why the majority of studies in the literature recommended only a gross total resection, with a long–term follow–up plan (7–9). When recurrence was observed, the standard of care to treat it involves repeat surgery and adjuvant radiation, because the pathology of recurrence was always more aggressive than the primary tumor.

The treatment plan for the present patient was surgerytotal resection without adjuvant radiotherapy, close follow-up was maintained during the subsequent 5 years; no clinical or radiological signs of recurrence were found. This therapeutic management was consistent with those of the literature.

Conclusion

Cerebellar neurolipocytoma is an adult well-differentiated neurocytic tumor of the cerebellum with local or regional lipomatous differentiation and which is characterized by a particular favorable outcome. Total resection is the basis of treatment. Adjuvant radiotherapy seems to be Adjuvant radiotherapy seems to be useful in cases of tumor recurrence. However, other studies are needed to better define the therapeutic approach and have more data on the evolutionary profile of this rare entity.

Patient consent

The consent has been obtained by the patient for publication in a scientific journal, and is available in the patient's file.

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