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Case report

Cerebellar liponeurocytoma with extracranial extension: Case report

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ARTICLE INFO

Article history:
Received 12 July 2014
Received in revised form
30 August 2014
Accepted 20 October 2014
Available online 28 October 2014

Keywords:
Liponeurocytoma
Cervico-occipital junction
Magnetic resonance imaging
Surgery

ABSTRACT

Cerebellar liponeurocytoma is a newly recognized, rare clinicopathological entity commonly described in the cerebellar hemispheres or the vermis.

We present a rare case of cerebellar liponeurocytoma arising from the left cerebellar amygdala with extracranial extension. Such a condition has never been previously reported.

1. Introduction

Cerebellar liponeurocytoma is a rare neoplasm with distinctive morphologic features, involving typically the cerebellar hemispheres of middle-aged to older adults and composed of densely packed neuronal cells mixed to foci of well-differentiated adipocyte-like cells, amazingly resembling to mature adipose tissue [1,2].

We present the first case of cerebellar liponeurocytoma arising from the cerebellar amygdala with extra cranial extension. In addition, we discuss the diagnostic, radiological and clinical features associated with this rare tumor.

2. Case description

A 59-year-old, previously healthy, woman was referred to our institution in June 2013 for a 9 months history of occipital pain. She did not present with nausea, vomiting or cerebellar/brainstem dysfunction signs. General physical examination was unremarkable, but neurological examination revealed exaggerated deep tendon reflexes in both lower and upper

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Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging.

http://dx.doi.org/10.1016/j.pjnns.2014.10.005

limbs with extensor plantar response in both sides. Fundoscopic examination disclosed a grade I papilledema.

Computed tomography (CT) scan revealed a poorly enhancing cerebellar lesion of the midline with central area exhibiting the attenuation values of fatty tissue. The lesion develops mainly in the posterior fossa but extends through the foramen magnum reaching the C2 level (Fig. 1). Magnetic resonance imaging (MRI) delineated better the tumor with its extent. The 4×4.5 well demarcated posterior fossa mass developed through the foramen magnum and compressed the fourth ventricle without obstructive hydrocephalus (Fig. 2). Spontaneously hyperintense area on T1-weighted sequences (Fig. 3) correlated with the CT hypo dense area, confirming the presence of fat within the tumor. Post contrast images revealed no significant tumor enhancement.

A posterior fossa craniotomy with C1 laminectomy was performed in the prone position. During surgery, the tumor was purple colored, moderately hemorrhagic and arising from the left cerebellar amygdala. It has a good cleavage plane with the rest of the cerebellum and the fourth ventricle and gross total resection was achieved.

Pathological examination revealed a highly cellular neoplasm composed of small monomorphic neuronal cells with oval to carrot-shaped hyper chromatic nuclei surrounded by scanty cytoplasm. Focal areas of lipomatous differentiation were distributed throughout the tumor but were predominant in the central lipidized area (Fig. 4). Immunohistochemistry was positive for synaptophysin and GFAP. These pathologic features were consistent with a liponeurocytoma. The Ki-67 proliferation marker stained positively in approximately 3% of the tumor cells.

Post-operative course was uneventful and the patient recovered quickly. Her pain completely disappeared and post-operative MRI scan performed three days after the surgery demonstrated no residual tumor. MRI of the whole spine was also performed to assess the presence of spinal drop metastases. No such lesions were identified. No adjuvant radiation therapy was administered and at most recent follow-up examination, 18 months after surgery, the patient is totally asymptomatic with no clinical or radiological evidence of recurrence.

3. Discussion

Cerebellar liponeurocytoma is a rare, recently defined, neoplasm composed of densely packed neuronal cells mixed to foci of well-differentiated adipocyte-like cells. Since its first description by Bechtel et al. [3] in 1978, this tumor has been variably reported as lipomatous medulloblastoma, lipidized medulloblastoma, medullocytoma, among others based on the fact that histopathological examination essentially revealed a medulloblastoma with heavily lipidized cells [1]. Over the years, several other cases have been reported and it became evident that the tumor clearly distinguishes itself from common medulloblastoma due to its different clinical behavior and age at onset. It was then introduced in the 2000 World Health Organization (WHO) classification of central nervous system tumors [4,5] and the 2007 revision reclassified this tumor as a grade II neuronal and mixed neuronal–glial neoplasm [6].

More than 40 cases of cerebellar liponeurocytomas have been reported in the English literature [7]. This rare tumor was described predominantly in the cerebellar hemispheres but may be located in the paramedian region and may extend to the cerebellopontine angle or the fourth ventricle [8]. A case of recurrent cerebellar liponeurocytoma with supratentorial extension has also been described [9] and one cerebellar liponeurocytoma extended from the left cerebellar hemisphere to the cerebellopontine angle and C1 space [10]. In the present case, the tumor was depending on the left cerebellar amygdala and developed mainly in the posterior fossa with extension through the foramen magnum reaching the C2 level, such a finding has never been reported.

Symptomatic period prior to presentation is often long [2]. Patients usually present between the ages of 58 and 60 years [11] with no sex predilection. Obstructive hydrocephalus is common and may be the reason for presentation [10]. Clinically, headache and other signs of raised increased intracranial pressure are the most common presenting symptoms [2]. Other manifestations include dizziness, unsteadiness, gait disturbance, frequent falls, visual symptoms, and signs of cerebellar or brainstem dysfunction depending on the tumor location [12].

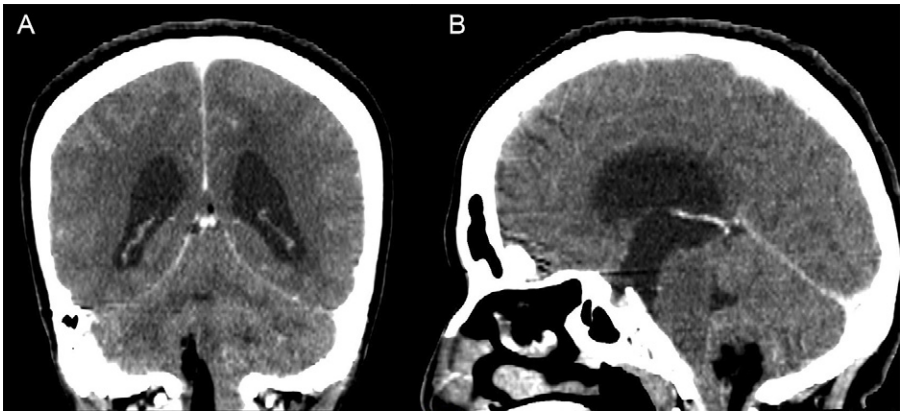

Fig. 1 – *Post contrast CT scan* in coronal (A) and sagittal (B) views, demonstrating a poorly enhanced cerebellar mass with a central area of marked hypo attenuation corresponding to fat density passing through the foramen magnum.

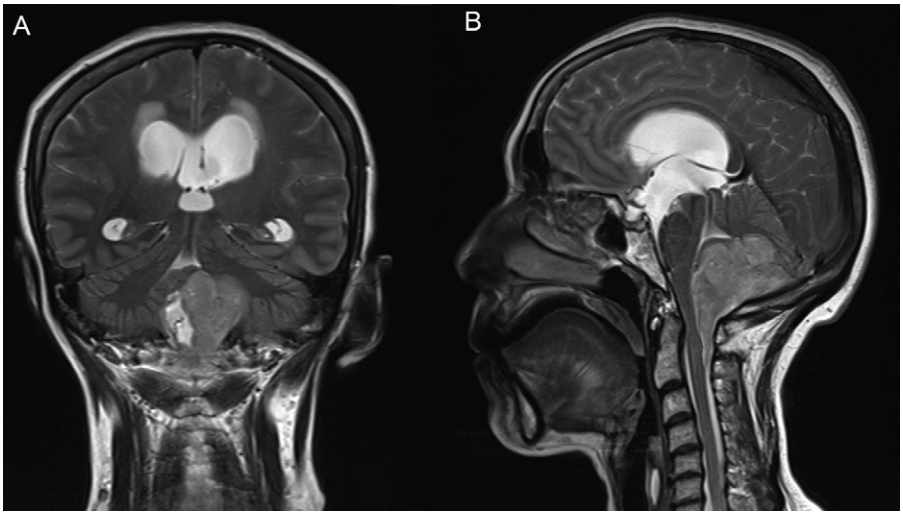

Fig. 2 – *Preoperative T2-weighted MR images* in coronal (A) and sagittal (B) views, showing a well circumscribed slightly hyperintense cerebellar mass; compressing the fourth ventricle and extending through the foramen magnum. Note the hyperintense streak corresponding to adipose content (A).

In sharp contrast, our patient presented with occipital pain and clinical signs of high cervical compression which is more suggestive of a cervico-occipital lesion than a posterior fossa tumor.

On CT scans, the tumor is variably isodense or hypodense relative to brain parenchyma with focal areas of marked hypo attenuation corresponding to fat density [10]. The lesion may or may not be well demarcated [2]. On T1-weighted MRI images, the tumor appears isointense to hypointense, with patchy areas of hyper intensity corresponding to regions of high lipid content. Contrast enhancement is often heterogeneous and may be minimal. Peritumoral edema is typically absent or minimal. Fat-suppressed images may be helpful in supporting the preoperative diagnosis of a liponeurocytoma [10]. In the present case, radiological findings were suggestive of a liponeurocytoma. Central lipidized zone passing through the foramen magnum is certainly uncommon in posterior fossa lesions but is of capital importance in assessing the differential diagnosis.

Surgery should be the initial therapeutic maneuver and whenever feasible gross total resection should be the goal [2]. Macroscopically the tumor is usually well demarcated with good cleavage plane between the lesion and the surrounding tissue.

Microscopically, it consists of small, round to ovoid cells with eosinophilic, scanty cytoplasm [2] morphologically similar to the prototypical neurocytic tumor of the central nervous system: central neurocytoma. Characteristically, a variable proportion of the tumor cells display cytoplasmic vacuoles

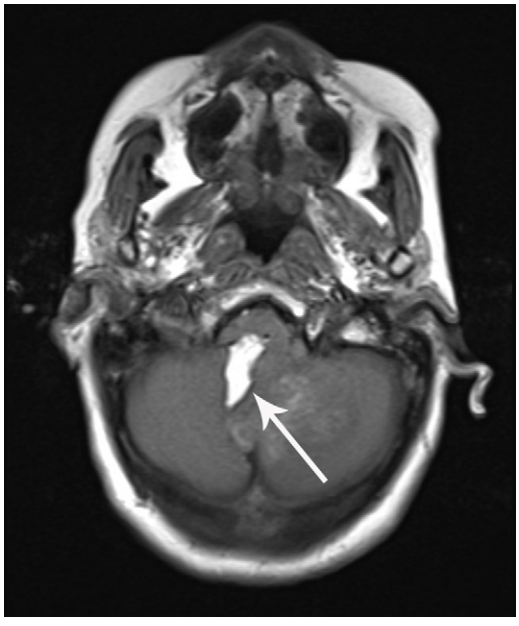

Fig. 3 – *Preoperative T1-weighted MR image* in axial view showing a central hyperintense area corresponding to adipose content (Arrow).

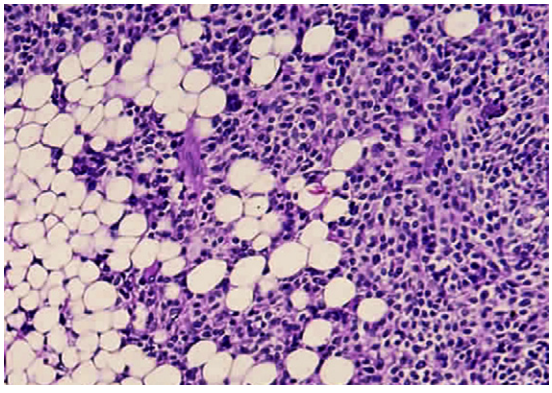

Fig. 4 – *Histological section of the tumor specimen* showing typical features of cerebellar liponeurocytoma with a highly cellular neoplasm composed of small monomorphic neuronal cells associated to focal areas of lipomatous differentiation (Hematoxylin & Eosin; original magnification ×100).

that are typically single and large. In those areas of lipidization, the tumor nuclei are displaced and compressed to the periphery of the cell, causing the tumor cells to resemble mature adipocytes [13]. Mitotic activity is typically low. Necrosis and vascular endothelial proliferation are rare but have been associated with more aggressive biologic behavior. One reported case [14] showed striated muscle differentiation in addition to lipidization, whereas another reported case [15] included a prominent glial component having pilocytic features. The immunostaining profile of previously published cases of cerebellar liponeurocytoma document uniform positivity for neural markers like synaptophysin, neuron-specific enolase, and microtubule-associated protein 2 (MAP-2) [4]. Expression of glial fibrillary acidic protein (GFAP) has been reported in most cases, although staining is typically focal and of limited intensity [16,17]. The MIB-1 (Ki-67) labeling index is typically between 1% and 3% but may be higher [18,19]. The histological differential diagnosis, therefore, includes variants of more common tumors featuring lipidized cells like ependymoma, glioblastoma, meningioma, as well as tumors showing morphologic features similar to cerebellar liponeurocytoma. First among that group is medulloblastoma.

The 5 year survival rate of cerebellar liponeurocytomas is 48%, but this should be interpreted with caution because of the rarity of this tumor and the lack of systematic follow-up [4]. In one collective series, reported by Horstmann et al. in 2004 [19], 6 of 15 patients (40%) with available follow-up information had recurrence during a period ranging from 1 to 12 years. Recurrent tumors may display increased mitotic activity, increased proliferative activity as assessed by MIB-1 staining, vascular proliferation, and necrosis [20–22]. However, such findings are tempered by other reports of recurrence in the absence of such atypical histopathological features [9,23] and it remains to be seen whether these atypical features are truly predictive of a more aggressive biologic behavior or not.

Radiation therapy may still have a role given the potential for local recurrence; and when administered, it is generally limited to the posterior fossa [11], as no cases of spinal drop metastasis from cerebellar liponeurocytoma have been reported to date. Due to the paucity of long-term follow-up data, it is unclear whether radiation therapy should be given in the immediate postoperative period or be reserved for recurrent tumor [18,19] or in case of high proliferation index [1]. In the present case no adjunctive radiation therapy was administered.

4. Conclusion

Cerebellar liponeurocytoma may extend through the foramen magnum and manifest like a cervico-occipital junction lesion.

The diagnosis must be suggested when facing a cerebellar lesion with adipose content and whenever feasible; gross total removal should be the goal.

As the characteristics of this rare tumor and its optimal management guidelines still need to be defined, we believe that radiation therapy should be reserved to cases of recurrences or in tumors with high proliferative activity.

Long term follow-up is mandatory to spot recurrences early.

Conflict of interest

None declared.

Acknowledgement and financial support

None declared.

Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.pjnns.2014.10.005.

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