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Case Report
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Cerebellar Liponeurocytoma: Case Report

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Abstract

Background: Cerebellar liponeurocytoma is a rare tumour of the posterior fossa included in 2000 by the WHO in the classification of brain tumors. It is characterized by divergent glioneuronal differentiation and lipidized neoplastic cells and has been considered as a distinct clinicopathological entity.

Case: In this article, the authors report a case of this rare tumour in a 45-year-old female patient clinically presenting with signs of increased intracranial pressure and cerebellar dysfunction. CT scan and MRI showed a heterogeneous mass in the left cerebellar hemisphere with hydrocephalus. A VP shunt was placed, and then gross total resection was performed, without adjuvant therapy. The postoperative period was uneventful. There was no recurrence after 2 years follow-up.

Conclusion: Long survival after surgical resection is often reported in this rare tumor but recurrence has been reported on an average of 10 years. The role of adjuvant therapy is still a matter of debate.

Keywords: Liponeurocytoma; Lipidized medulloblastoma; Adipose tissue; Treatment and recurrence

Introduction

Cerebellar Liponeurocytoma (CL) is a rare and slow-growing tumour displaying neural, astrocytic and focal lipomatous differentiation. Initially termed "cerebellar medulloblastoma", the 2000 WHO classification of tumors of the CNS, proposed the term of "cerebellar liponeurocytoma" as a growing body of evidence suggested that this entity is a distinct clinicopathological entity, different from medulloblastoma [1]. We report a case of this tumour in a 45-year-old female patient and highlight its particularities regarding its neuropathological features and treatment options.

Case Presentation

A 45-year-old woman presented at the Neurosurgical Department with a 3 month history of occipital headaches worsening, vomiting and unsteadiness of gait. Upon physical examination, she was found to have papilloedema. The rest of the neurological examination was normal. Computed tomography and MRI showed a contrast enhancing heterogeneous mass in the left cerebellar hemisphere, displacing the forth ventricle and causing moderate mass effect and supratentorial hydrocephalus (Figure 1 and 2). The patient underwent a VP shunt followed by a left suboccipital craniotomy in the prone position; a dural incision was made, then a gross total resection was performed without complications. Microscopic examination revealed a heterogeneous tumor, composed of a population of small round neurocytic cells, accompanied by a focal lipomatous differentiation composed of cells resembling mature adipose tissue. Tumoral cells appeared blank, uniform with very few mitotic figures. Microvascular proliferation and necrosis were absent. Postoperative CT scan showed no evidence of residual contrast enhancement in the cerebellum (Figure 3). The post-treatment course was uneventful and the patient was discharged 5 days after the procedure. The patient received no adjuvant treatment and there is no evidence of tumour recurrence after 2 years follow-up.

Discussion

Cerebellar liponeurocytoma is a rare neoplasm showing neuronal, glial neoplastic and focal lipomatous differentiation [2,3]. Since 1978, when first described by Bechtel et al. [4], there have been more than 40 reported cases [5]. It has many morphological similarities to medulloblastoma and neurocytoma, and this tumour is reported under different names like "lipomatous medulloblastoma", "lipidized medulloblastoma", "neurolipocytoma", "medullocytoma" or "lipomatous glioneurocytoma" [2,6,7]. The WHO Working Group proposed the term

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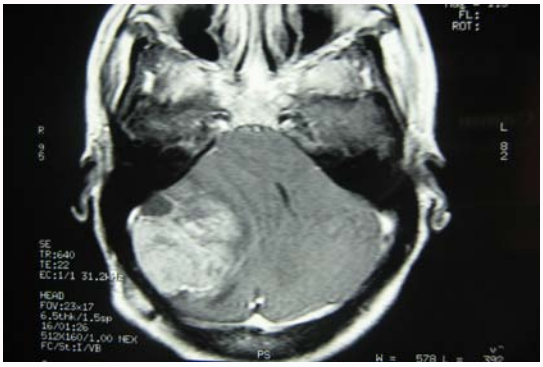

Figure 1: Radiological appearance of the lesion, Axial T1W MRI with gadolinium showing a large posterior fossa lesion involving the right cerebellar hemisphere, with local mass effect on the fourth ventricle, the lesion is heterogeneous.

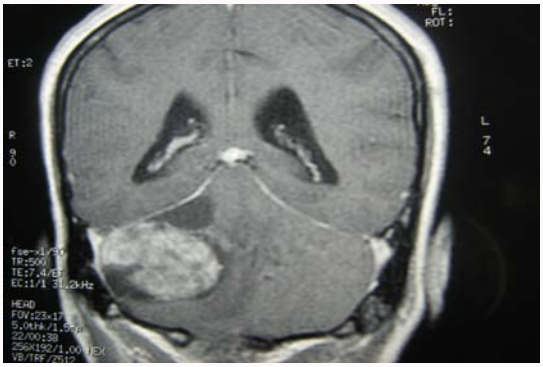

Figure 2: Coronal T1W MRI obtained after gadolinium administration revealing an irregular enhancing lesion in the right cerebellar hemisphere.

"cerebellar liponeurocytoma", to emphasize its similarities with central neurocytoma and to avoid any possible confusion with medulloblastoma resulting in an unjustified aggressive adjuvant therapy [1]. The tumor was first assigned the WHO grade I for its blank histological feature and the good prognosis. However, recurrences proved later to be frequent in this tumor; more than in 50% of cases, in the absence of any morphological hallmark of malignancy, leading the WHO Working Group in the classification of the central nervous tumors to upgrade the tumour to grade II in the 2007 version of the classification. Cerebellar liponeurocytoma is currently included in the group of neuronal and mixed neuronal-glial tumors. The pathogenesis of lipid accumulation in these tumors is uncertain but has been ascribed a variety of mechanisms including metaplasia, divergent differentiation and endocytosis of external lipids. Immunohistochemistry data suggest that the adipose-like cells result from lipomatous differentiation of tumour cells, as they express GFAP and neuronal markers and should derive from a pluripotent precursor cell able to differentiate into several ways (astrocytic, neuronal, mesenchymal) but with a preferential shift to neuronal differentiation. Microscopically, liponeurocytoma has small, round to ovoid cells with scanty eosinophilic cytoplasm [5]. No capsule is seen, mitoses and necrosis are generally absent [2]. Immunohistochemistry shows astrocytic and neuronal differentiation by revealing the expression of glial GFAP, and neuronal proteins (synaptophysin and NSE and MAP2) [1,2]. Synaptophysin and GFAP expression in fat-containing cells indicates true lipomatous differentiation instead of some entrapped adipocytes. The presence of adipose cells in

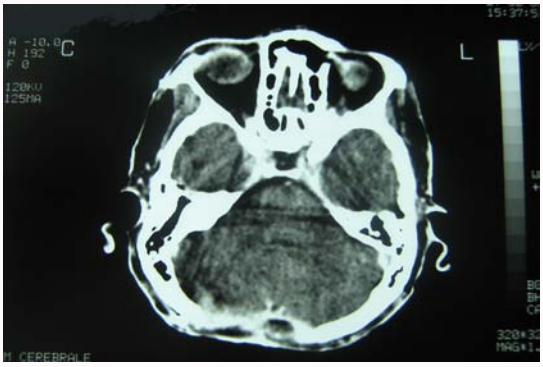

Figure 3: Postoperative axial CT scan showing no residual tumour and no contrast enhancement.

neuroglial neoplasms has been exceptionally related in the literature not being restricted to cerebellum, neither to medulloblastoma. It has also been described in spinal cord neoplasms and in supratentorial ependymomas [7]. MIB-1 labeling index is usually in the range of 1% to 3%. All of the reported cases of cerebellar LPN have been found in adults, and the tumour classically manifests between the fourth and sixth decades and the age of the patient ranges between 36 to 67 years [3]. There seems to be no sex predominance [8]. The lesion may be situated within the cerebellar vermis or the hemispheres [9]. Preoperative diagnosis is difficult because of non-specific radiological features. Computerized tomography scan demonstrates heterogeneous hyperdense or isodense lesion sometimes associated with parenchymal cysts or cerebellar hemorrhage [9]. Magnetic resonance imaging demonstrate the areas of fat density, in T1-weighted sequence, the tumour is hypointense with scattered foci of hyperintense signal and displays moderate contrast enhancement, peritumoral edema is not usually seen.

Because of the rarity of the tumors, there is no consensus regarding the treatment strategy [7]. Patel suggested that gross total removed with long-term follow-up period is sufficient and avoids exposing the patients to the risks and side-effects of radiotherapy, without any evidence to support its usefulness in preventing recurrence [5]. On the other hand with longer clinical follow-up periods it has become clear that this tumour has a rate of recurrence that is higher than previously thought. Recently, recurrence has been observed after a long period of time (around 10 years) in 20% to 32% of patients [10,11]. Pasquale [11] reported early recurrence after 3 years following a gross total removal of CL without adjuvant therapy. Jenkinson reported an unusual case of a cerebellar liponeurocytoma that presented a first recurrence 12 months after subtotal removal followed by radiotherapy of the first lesion, and a second recurrence 3 months after a second surgical removal [12]. In 2009, Limaiem reported 9 cases of recurrent CL published in the literature, all were confined in the posterior fossa and one with supratentorial extension, and concluded that there is not always a correlation between histological features and biological behavior of this neoplasm [13]. Cacciola stated that these tumors recur with histology more aggressive than the original tumour; however he suggested that complete resection seems to be the best appropriate treatment [10]. If tumour recurrence does occur, then additional surgery, followed by fractionated radiotherapy, may be necessary. Jackson, et al. reviewed the finding for 20 reported cases of CL and noted late recurrences in 50% of patients who survived surgical resection and who did not receive adjuvant therapy, he concluded that resection and adjuvant

radiotherapy to the posterior fossa seemed to be the best approach to reduce the local recurrence rate [8]. Châtillon suggested that adjuvant radiotherapy should be considered after complete resection of tumors with a high proliferation index [14]. However, longer follow-up and analysis of similar cases are obviously necessary to confirm if the histological appearance can be predictive of an unfavorable clinical course [15].

Conclusion

We report our first case of cerebellar liponeurocytoma occurring in 45-year-old woman, who underwent a gross total resection without adjuvant treatment. A 2-year follow-up period showed no evidence of recurrence. However, cerebellar liponeurocytoma proved too frequently recur and an extensive follow-up is then necessary. Whether an adjuvant therapy is needed, in conjunction with surgery, is still a matter of debate.

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