

## Case Report

# Cerebellar Lipo Neurocytoma: Case Report and Review of the literature

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### Abstract

*Background:* Cerebellar liponeurocytoma is a rare tumor of the central nervous system occurring mainly in the posterior fossa, which shows divergent glioneuronal differentiation and lipidized neoplastic cells and has been considered as a distinct clinicopathological entity.

*Case:* Herein, we describe a 19-year-old male patient who presented to the hospital complaining of headache, nausea with balance difficulty. MRI revealed a contrast enhancing midline lesion of posterior fossa. After tumor resection, histological examination and immunohistochemistry were done and the diagnosis of cerebellar liponeurocytoma was confirmed.

*Conclusion:* Liponeurocytoma is a rare benign tumor with cerebellum is the typical site for it. The therapy of choice is surgery. Postoperative radiotherapy has to be discussed individually.

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### Introduction:

Cerebellar lip neurocytoma is a rare tumor entity mainly located in the posterior cranial fossa<sup>1</sup>. Cerebellar lip neurocytoma is most commonly found in adults<sup>2</sup>. These tumors have a very similar appearance to medulloblastomas on light microscopy, with the additional element of well-developed fat cells<sup>3</sup>. The term "lip neurocytoma" was first introduced in the 2000 World Health Organization as "a distinct clinic -

pathological entity in the group of glioneuronal tumors". Now, it is defined as a WHO grade II tumor with neuronal consistent, variable astrocytic and focal lipomatous differentiation and with infiltrative nature, low proliferation index, but high likelihood of recurrence<sup>4</sup>. Clinical symptoms include the symptoms of the cerebellar dysfunction, such as gait disturbance and uncoordinated movements, which are slowly progressive and generally not detected in the early

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stages of the disease. Vomiting and progressive visual symptoms begin to present, as a result of increased cranial pressure in the later stages of the disease. The present study reports a case located in the cerebellum, and also provides a review of the literature<sup>2</sup>.

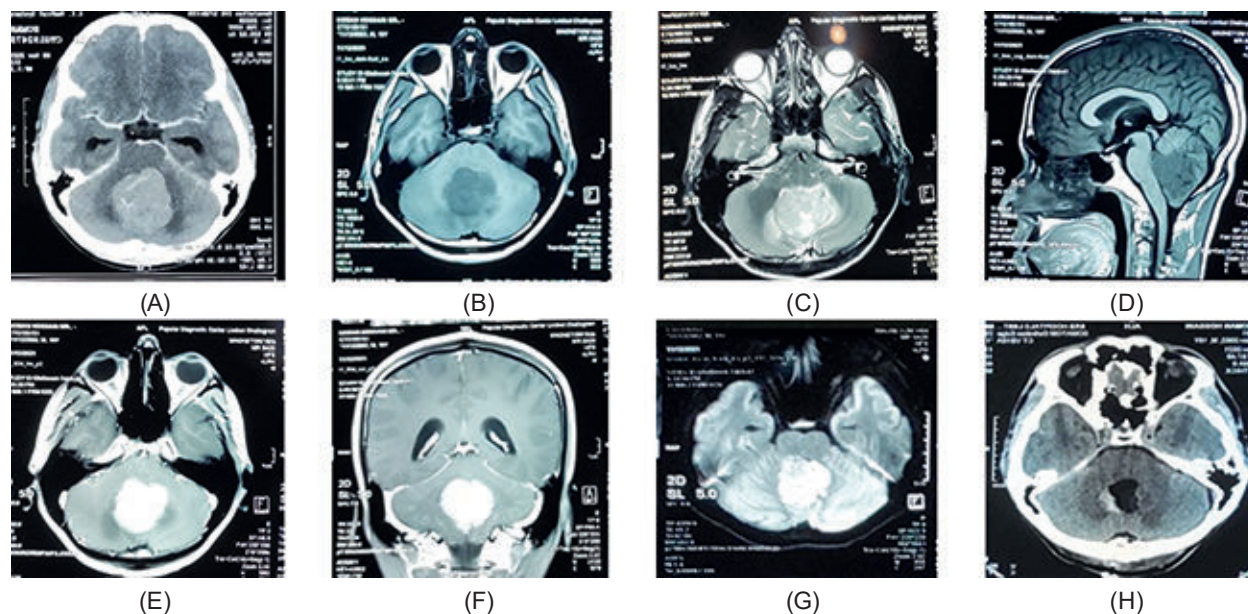
### Case report:

A 19-year-old righthanded, male, Muslim, unmarried patient presented to our department suffering from headache, nausea and mild balance difficulty. He did not suffer from any other co-morbidities; had no history of significant past medical or surgical illness, not used to any regular medicine or substance abuse history, none of his family member suffered from such kind of illness. Neurological examination revealed no cranial nerve deficits or cerebellar sign, normal response of his reflexes, and normal standing and walking abilities without any unstableness. Blood pressure, pulse, temperature, and laboratory findings (that is, complete blood count, liver function, renal function, and) were within normal range. The patient subsequently underwent

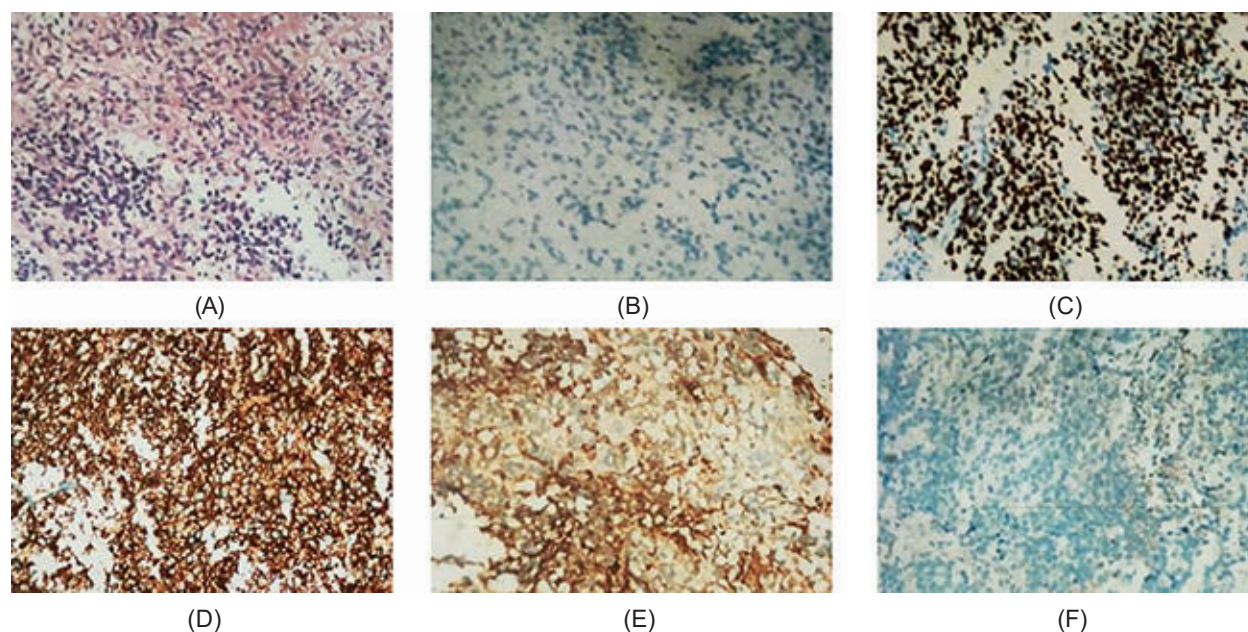
Computed tomography (CT) and magnetic resonance imaging (MRI) of the brain, CT scan of brain with contrast revealed well defined contrast enhancing hyperdense mass is seen in the posterior fossa at almost midline location compressing the 4<sup>th</sup> ventricle

with mild dilatation of others ventricles. With and without contrast enhancement MRI revealed a lesion in the midline of posterior fossa measuring about 3.6 x 3.6 x 4.7 cm (anterior-posterior x transverse x cranial-caudal) which is hypointense on T1 weighted images, mildly hyperintense on T2 weighted and FLAIR images and intensely enhance with diffusion restriction. There is moderate perilesional edema, 4<sup>th</sup> ventricle is almost completely effaced with mild dilatation of both lateral ventricles. After taking informed written consent he underwent suboccipital craniotomy under general anesthesia. After Dural opening, we observed a gray - reddish tumor areas that was on the posterior aspect, it was well demarcated from the surrounding cerebellar cortex and slightly adhering to the tentorium and lateral wall of the fourth ventricle. It was; however, easily cleavable and gross total removal was achieved by microsurgical technique. The wound was closed in layers. After surgery he was kept overnight in neurosurgical intensive care unit for close observation and was transferred to a general ward the following day without neurological deficiency. The postoperative course was uneventful and the patient was discharged on 4<sup>th</sup> post-operative day. On histopathological examination,

hematoxylin and eosin-stained paraffin sections showed a tumor made of biphasic with admixture of



**Fig.-1:** Well defined contrast enhancing hyperdense mass is seen in the posterior fossa at almost mid-line compressing the 4<sup>th</sup> ventricle with mild dilatation of others ventricles (Fig A). Intensely enhancing lesion in the posterior fossa which is hyperintense on T1WI, mildly hyperintense on T2WI and FLAIR image and diffusion restricted. 4<sup>th</sup> ventricle completely effaced with dilatation of lateral ventricle (Fig B to G). Post-operative CT scan (Fig H).



**Fig.-2: Histopathological slide**

neurocytes and lipidized cells. Neurocyte are arranged in densely cellular sheets of monotonous cells with scanty and often clear cytoplasm, round to oval nuclei, salt and pepper chromatin. The lipidized cell are typically clustered and resemble mature adipocyte. In immunohistochemical analysis tumor cells were positive with NSE, GFAP, Synaptophysin and Ki67. Tumor cells were negative with P<sup>53</sup> protein, which compatible with liponeurocytoma. He receiving no adjuvant treatment.

#### **Discussion:**

The first case of cerebellar liponeurocytoma, in a 44-year-old man, was described in 1978 by Bechtel et al. Findings from computed tomography (CT) and magnetic resonance imaging (MRI), and pathological and immunohistochemical characteristics have been reported in around 70 cases<sup>1</sup>. The patients' ages ranged from 4 to 69 years, with a median of 49 years. No gender predominance has been reported<sup>5</sup>. Cerebellar liponeurocytoma has been known by many names, including lipomatous medulloblastoma, lipidized medulloblastoma, medulloctoma, neurolipocytoma, lipomatous glioneurocytoma and lipidized mature neuroectodermal tumor of the cerebellum<sup>3</sup>. Differential diagnoses include oligodendrogliomas, clear cell ependymomas, or high-grade tumors like medulloblastomas. Oligodendrogliomas may show similar pathological findings demonstrating neuronal differentiation with

glial and neuronal characteristics. Oligodendrogliomas show highly positive results of Olig2 immunohistochemistry in contrast to liponeurocytomas<sup>1</sup>. Despite its rarity, it is mandatory to distinguish it from medulloblastomas. This entity usually has a more favorable prognosis and does not require chemotherapy or radiotherapy<sup>6</sup>. Medulloblastoma with lipidized cells and lipomatous ependymomas can be differentiated using immunohistochemical panels. Medulloblastomas show hyperdense areas with calcification in around 20% of cases on CT scans and a homogenous contrast enhancement can be observed on CT scans with contrast enhancement. On MRI, a hypointense signal on T1-weighted images and an isointense to hyperintense signal on T2-weighted images can be observed for medulloblastomas. A hyperintense FLAIR image can be detected. Diffusion-weighted images of medulloblastomas are hyperintense and on T1-weighted images with contrast enhancement medulloblastomas normally present a heterogeneous strong contrast enhancement<sup>1</sup>. Morphologically, the most distinctive feature is the presence of cells indistinguishable from mature adipocytes. The mature adipocyte-like cells generally are thought to result from lipomatous differentiation of tumor cells rather than an upset ion cellular metabolism or an admixture of non-neoplastic mesenchymal cells<sup>7</sup>. Areas of neuronal differentiation are positive for markers such as synaptophysin and MAP-2.8 Reactivity for GFAP

is usually present but focal. Fat-containing cells also typically express neuronal markers and GFAP, consistent with aberrant differentiation of tumor cells rather than them being true adipocytes. There is usually no histological evidence of anaplasia or rapid growth, such as mitosis, necrosis, or vascular hyperplasia. The Ki 67/MIB-1 index is usually low (< 5%); however, patients with a relatively high proliferation rate and cytopathologic atypia have been reported to have a greater risk of recurrence and, therefore, a poorer prognosis<sup>8</sup>. The present case shows the classical morphology and immunohistochemical traits of cerebellar liponeurocytoma, although neuronal and glial differentiation is less evident. There is currently no established standard of care, but maximal safe resection followed by postoperative imaging surveillance for recurrence seems prudent. The roles of adjuvant chemotherapy and radiation have not been established<sup>8</sup>. Based on literature reviews, recurrence has been observed in 20-32% of patients, on an average, 10 years after surgery (range 8-12 years). Total resection is considered the optimal treatment, while additional EBRT is controversial<sup>9</sup>.

**Conclusion:** The small number of patients with reported cerebellar liponeurocytomas limits our understanding of the tumor's natural history. In daily practice, cerebellar liponeurocytomas must be remembered as a differential diagnosis of cerebellar masses, and must be distinguished from medulloblastomas, to avoid complementary therapies, thus expecting a better outcome. Most of the information available from case reports indicates that this tumor is less aggressive than typical medulloblastomas and is similar in prognosis to central neurocytomas.

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