

Case Report:**Cerebellar liponeurocytoma: a case-report**

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ABSTRACT

Cerebellar liponeurocytoma is a rare cerebellar neoplasm of adults with advanced neuronal / neurocytic and focal lipomatous differentiation, a low proliferative potential and a favorable clinical prognosis corresponding to World Health Organization grade I or II. Only a few cases have been described in the literature (approximately 20 cases) by different names. A 48-years old female, presented with history of headache and dizziness associated with neck pain; restricted neck movements, drop attacks and occasional regurgitation of food since one year. Magnetic resonance imaging disclosed a right cerebellar mass lesion. Gross total resection of the tumour was accomplished through a suboccipital craniotomy. The excised tissue was diagnosed as cerebellar liponeurocytoma, a rare entity, based on histopathological examination and immunohistochemistry. The morphological appearance of this neoplasm can be confused with that of oligodendroglioma, neurocytoma, ependymoma, medulloblastoma, solid hemangioblastoma and metastatic carcinomas etc., with unpredictable prognosis, which require postoperative radiotherapy, hence the importance of accurately diagnosing this rare neoplasm. This tumour should be added to the differential diagnosis of mass lesions of the posterior fossa.

Key words: *Liponeurocytoma, Cerebellum, Posterior cranial fossa*

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INTRODUCTION

Cerebellar liponeurocytoma (ICD-O Code - 9506/1) is a rare tumour of the posterior fossa that has been reported fewer than 20 times in the literature. This uncommon tumour was first described by Bechtel et al.¹ in 1978 in a 44-year old man. The terms neurocytoma /lipoma (neurolipocytoma),² lipomatous medulloblastoma,³ lipidized medulloblastoma,⁴ medullocytoma,⁵ lipomatous glioneurocytoma⁶ and lipidized mature neuroectodermal tumour of the cerebellum⁷ have been proposed, so as to emphasize its similarity to central neurocytoma and the prognostic difference from the ordinary medulloblastoma. It has been suggested in the available literature that these tumours have a favorable prognosis.^{3,5} Cerebellar liponeurocytomas were recognized in the revised WHO classification of tumours of the central nervous system 2000, as a distinct clinicopathological entity.⁸ Here, we report a case of cerebellar

liponeurocytoma with clinical, histological, and radiological studies.

CASE REPORT

A 48-year old female patient presented with head ache and dizziness of one year duration associated with neck pain, restricted neck movements, drop attacks and occasional regurgitation of food. On physical examination there was mild motor deficit in the right upper limb, loss of pain and touch sensations by 20% of normal, right upper motor neuron paralysis of VII nerve and positive cerebellar signs on ipsilateral side.

Investigations revealed neutrophilic leukocytosis with elevated erythrocyte sedimentation rate (ESR) (38 mm at the end of 1st hour) and normal biochemical parameters. Magnetic resonance imaging (MRI) showed heterogeneously enhancing mass lesion in the right cerebellar hemisphere

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Involving vermis with compressed and effaced fourth ventricle and transtentorial herniation. With the clinical and radiological suspicion of either high grade glioma or medulloblastoma, the patient underwent suboccipital craniotomy and excision of the tumour. The excised specimen was grey-reddish colored fragmented tissue *in toto* measuring 5.5×5.0×1.2 cm. The entire tissue was subjected for histopathological examination followed by immunohistochemistry (IHC) for further confirmation. The postoperative course was uneventful and the patient was discharged on the tenth postoperative day.

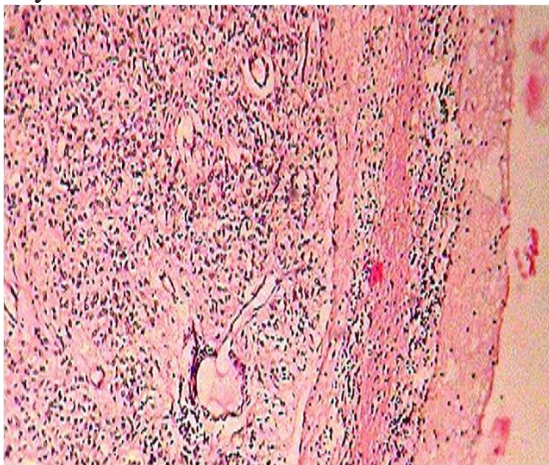


Figure 1: Photomicrograph showing cerebellar tissue with tumour component (Haematoxylin and eosin × 400)

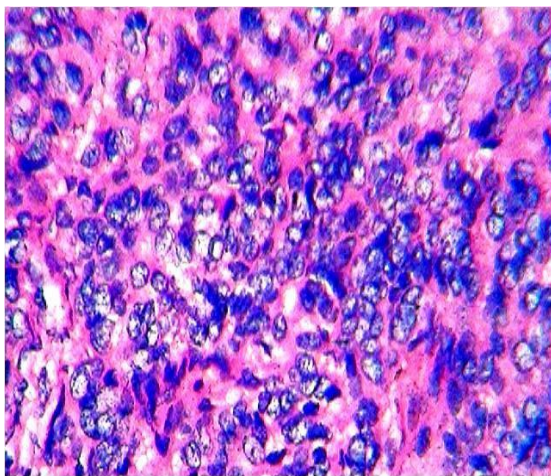


Figure 3: Densely populated highly pleomorphic tumour cells with indiscernible cytoplasm and atypical mitotic figures (Haematoxylin and eosin × 400)

Histopathological examination revealed cerebellar tissue with the tumour component exhibiting biphasic pattern comprising of isomorphic small neuronal cells with round to oval nuclei and clear cytoplasm. These cells are seen as sheets and at areas densely populated with indiscernible cytoplasm, highly pleomorphic nuclei with atypical mitotic figures. The tumour is significantly vascular (plexiform capillary arcade). Fine fibrillar intervening matrix, psammoma body like spherules of calcifications, foci of

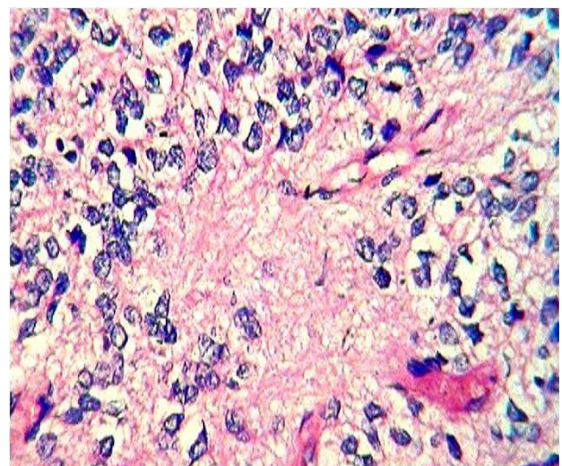


Figure 2: Photomicrograph showing pleomorphic tumour cells, mildly acidophilic to clear cytoplasm with intervening nucleus-free areas of neuropil (Haematoxylin and eosin × 400)

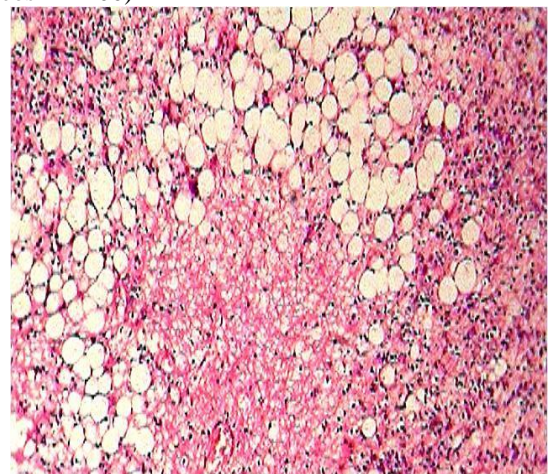


Figure 4: Photomicrograph showing tumour component exhibiting lipomatous differentiation and foci of microcystic degeneration (Haematoxylin and eosin × 400)

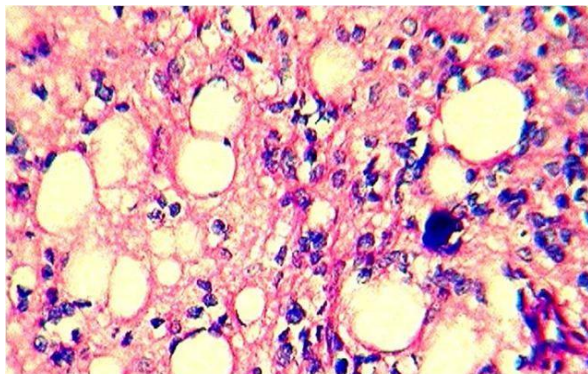


Figure 5: Photomicrograph showing tumour component with intervening lipidized cells and psammoma body like calcific spherule (Haematoxylin and eosin $\times 400$)

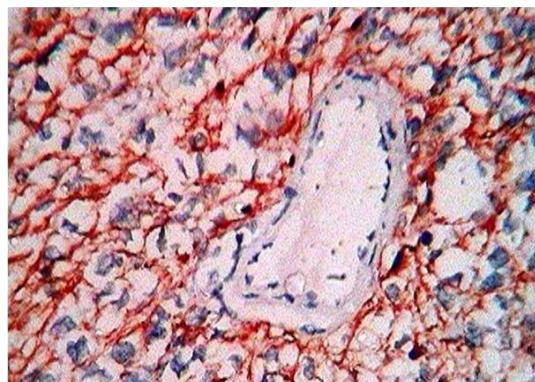


Figure 6: Photomicrograph showing tumour component with immunoreactivity for GFAP (Horse radish peroxidase $\times 400$)

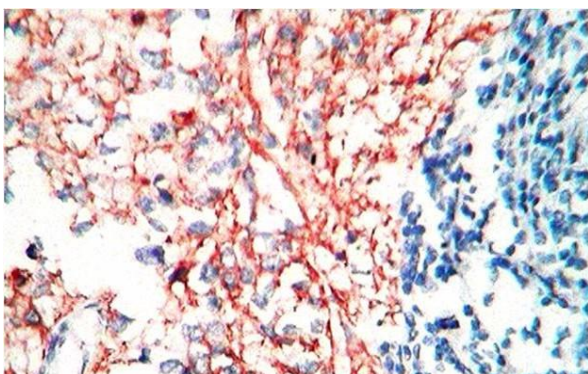


Figure 7: Photomicrograph showing tumour component with immunoreactivity for NSE. (Horse radish peroxidase $\times 400$)

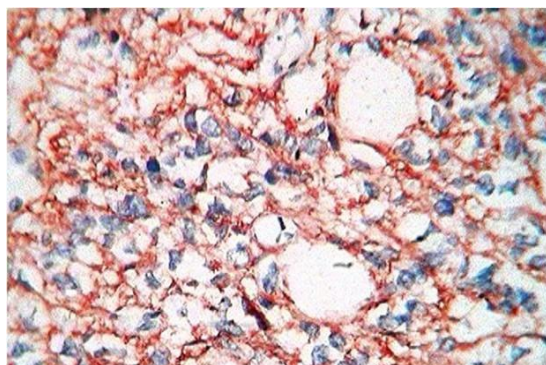


Figure 8: Photomicrograph showing tumour component with immunoreactivity for synaptophysin (Horse radish peroxidase $\times 400$).

lipoid cell change and microcystic degeneration present (Figures 1, 2, 3, 4 and 5). Immunohistochemically these neoplastic cells were reactive for glial fibrillary acidic protein (GFAP), neuron specific enolase (NSE) and synaptophysin (Figures 6, 7 and 8). Basing on these findings a diagnosis of cerebellar liponeurocytoma with focal aggressive areas [World Health Organization (WHO) Grade II] was made.

DISCUSSION

Cerebellar liponeurocytomas are neuroectodermal tumours consisting of both neuronal and glial elements. Immunohistochemistry for GFAP, synaptophysin and NSE are usually positive indicating the mixed glial and neuronal elements. Histogenetically immunoreactivity to neuronal antigens and GFAP includes cell bodies embracing fat globules.

This suggests that the fat containing cells result from lipomatous differentiation of tumour cells, rather than admixture of non-neoplastic adipocytes⁹. Since 2000, the WHO Central Nervous System tumours classification includes a subset called cerebellar liponeurocytoma.¹⁰ These tumours tend to occur in older patients ranging from 36 to 77 years of age, with a mean age of approximately 53 years.²⁻⁵ Cerebellar liponeurocytoma has a relatively benign clinical course and a recurrence may appear after a long period of time.^{2,4-6,8} Reviews published in the literature show a 5 year survival rate of 81%¹¹ but Jenkinson¹² reported a patient who developed a recurrence 12 months after a subtotal resection. His case demonstrated an atypical clinical course of a highly aggressive and radiation

-resistant tumour, despite the consistent absence of aggressive histological features. Radiological diagnosis is difficult due to the rarity of the tumour and a variable imaging appearance.¹³ The most challenging differential diagnosis of these tumours is to distinguish them from oligodendrogliomas, medulloblastomas, ependymomas, solid hemangioblastomas, astrocytomas and metastases. MRI appearance is variable and may be related to the distribution and proportion of lipidized tissue.⁴ Surgery to establish the diagnosis and remove the lesion should be the initial therapeutic manoeuvre. The aim of the surgery is gross total resection of the tumour. In most of cases there is a reasonable border between the tumour and surrounding tissue^{11,13} and gross total removal of the tumour is possible. The recent 2000 WHO classification of tumours of the nervous system describes a new entity, cerebellar liponeurocytoma.¹⁰ This rare neoplasm manifests with cerebellar symptoms in adults and is characterized histopathologically by the presence of the tumour with neuronal, astrocytic and focal lipomatous differentiation having low proliferative potential in an otherwise typical primitive neuroectodermal neoplasm. The present case looks aggressive which warrants close follow up. Most of the information available from previous case reports indicate that this tumour has a favorable prognosis, but this is yet to be confirmed in a larger series of cases.

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