Central liponeurocytoma: Case report and review of literature

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ABSTRACT

Cerebellar liponeurocytoma consists of well-differentiated neurons with the cytology of neurocytes in addition to a population of lipidized cells. Hence it is biphasic in appearance and has been included in the category of glioneuronal tumors of the central nervous system by the WHO working group on the Classification of Tumors of the Nervous System. However, liponeurocytoma is not exclusive to the cerebellar or fourth ventricular location. Since its inclusion in the central nervous system tumor classification, nine cases with similar histological and immunohistochemical features have also been described in the lateral ventricles. We describe here such a lateral ventricular tumour in a 30-year-old woman, characteristically showing divergent glio-neuronal differentiation and lipidized neoplastic cells. Therefore, we suggest that future WHO tumor classification should consider that liponeurocytomas are not entirely restricted to the cerebellum and henceforth change of nomenclature might be considered, as also pointed out by other authors.

Key words: Central liponeurocytoma, cerebellar liponeurocytoma, glioneural tumors

INTRODUCTION

Cerebellar liponeurocytoma has been included in the category of glioneuronal tumors of the central nervous system by the WHO working group on the "Classification of Tumors of the Nervous System.[1] This tumor consists of well-differentiated neurons with the cytology of neurocytes in addition to a population of lipidized cells resembling mature adipose tissue, and hence is biphasic in appearance. Such tumors occur in older adults and have a relatively good long-term prognosis and were earlier considered to be a variant of medulloblastomas. Their relatively benign nature and different morphological features led to this change of nomenclature.[2] Cerebellar liponeurocytoma is the most frequent neuroepithelial CNS tumor with adipose-like cells. Liponeurocytoma is not exclusive to the cerebellar or fourth ventricular location. Since its inclusion in the central nervous system tumor classification, nine cases with similar histological and immunohistochemical features have also been described in the lateral ventricles.[3] Therefore, linking these type of tumors only to cerebellum obscures the existence of similar neoplasms at other sites, such as among classic central neurocytomas of the lateral and third ventricles.

We describe here such a lateral ventricular tumor in a 30-year-old woman, characteristically showing divergent glio-neuronal differentiation and lipidized neoplastic cells.

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CASE REPORT

We recently had a 30-year-female patient who presented to us with chief complaints of holocranial headache associated with off and on vomiting and progressive painless diminution of vision in both eyes for two months. Patient also had progressively worsening gait during this period. Patient was conscious, alert with normal higher mental functions. Patient's visual examination revealed visual acuity of 6/9 in both eyes with no field cuts. Fundus examination showed grade 3 papilloedema in both eyes. Rest of the cranial nerves was normal. There was no associated limb weakness or sensory loss. MRI showed a large irregular mass in left lateral ventricle pushing the septum pellucidum to right side and also extending across it. Lesion was iso to hypointense on T1W and heterogenously hyperintense on T2W images and showed heterogenous enhancement on contrast injection [Figure 1]. Radiological impression was of a primary lateral ventricular tumor and neurocytoma was considered to be the most probable diagnosis as per the

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radiological features, tumor location and patient's age. Patient underwent a left frontoparietal quadrangular flap craniotomy and near total excision of the intraventricular tumor. Tumor was reddish grey, highly vascular, soft and suckable. Intra operative frozen section was inconclusive regarding the tumor type. Final histology showed sheets of small round cells separated by thin vascular channels [Figure 2a, H and E, ×100]. Higher magnification showed acellular fibrillary areas (arrow) and cellular areas with clearing of cytoplasm resembling oligodendroglial cells [Figure 2b and c, H and E, ×200]. Areas of lipomatous differentiation were seen [Figure 2d, H and E, ×100]. On immunohistochemistry, diffuse cytoplasmic positivity for synaptophysin and nulearpositivity for Neu-N [Figure 3a and b, ×200] was seen. Neu N nuclear positivity is also seen in the cells showing lipomatous change [Figure 3c, ×200]. MIB 1 labeling index was less than 1% [Figure 3d, ×100]. A final diagnosis of central liponeurocytoma was made based on these findings.

DISCUSSION

The origin of adipocytic elements in these tumors is still disputed. Some researchers suggest that the presence of such cells indicates that the neoplasm has evolved in dysgenetic areas where distinct cellular elements intimately co-exist. Others feel that there is a progressive accumulation of lipid vacuoles in the cell's cytoplasm, probably due to some degenerative or metabolic changes. These cells, therefore, appear similar to adipocytes on microscopy though on immunohistochemistry their neuroglial origin becomes clear.[4,5] Our case showed glial, neuronal and lipomatous differentiation in a lesion which was located in lateral ventricle. On reviewing the literature, we noticed that these tumors characteristically affect adults and show a much more benign evolution.[6] However, there have been case reports of recurrence and malignant progression in such tumors. Recurrence has been observed in 20-32% of patients, on an average of 10 years after surgery (range 8-12 years) in cerebellar subtype.[7,8] As a result, the benign nature of the tumor is being questioned.[9,10] Total resection is considered as the optimal treatment and the role of radiotherapy is controversial.[7,8,11] There is still no consensus regarding post operative adjuvant therapy. In most of the cases reported till now, adjuvant radiotherapy was offered when residual tumor was present.[7,12] Only long-term follow up of a large number of such patients can give us an insight into the biological behavior of this rare tumor and will help to establish whether there is any prognostic difference between the relatively commoner cerebellar liponeurocytomas and the uncommon supratentorial location variants, as the only case where early recurrence and malignant transformation have been reported are the cerebellar liponeurocytomas. Therefore we suggest that future WHO tumor classification should consider that liponeurocytomas are not entirely restricted to the cerebellum and henceforth change of nomenclature might be considered, as also pointed out by other authors.

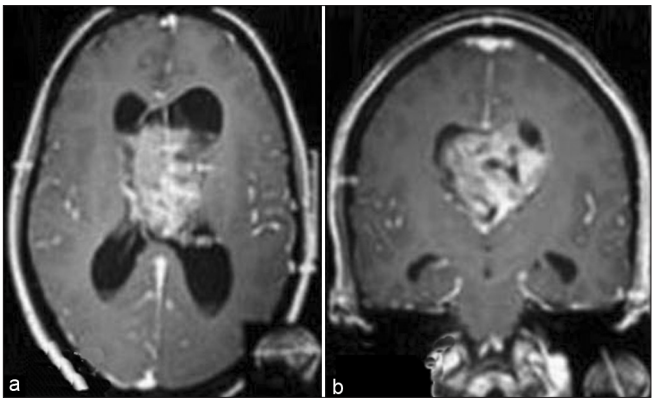

Figure 1: Contrast MRI axial (a) and coronal (b) section showing heterogenous, enhancing lesion primarily in left lateral ventricle pushing the septum pellucidum to right and also extending to the other side

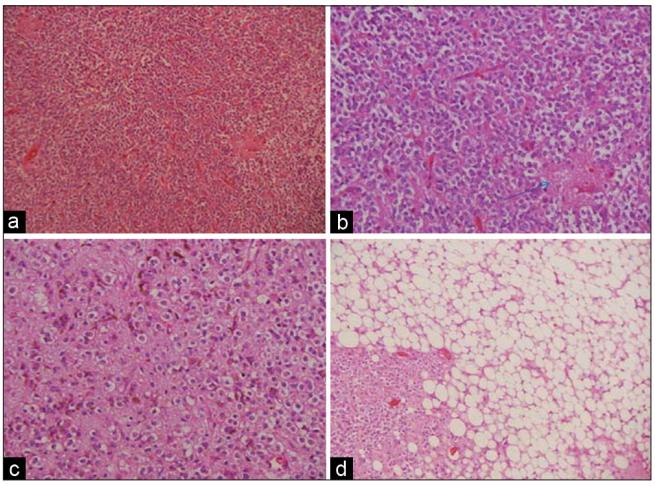

Figure 2: Histology slides (H and E) showing biphasic cellular architecture

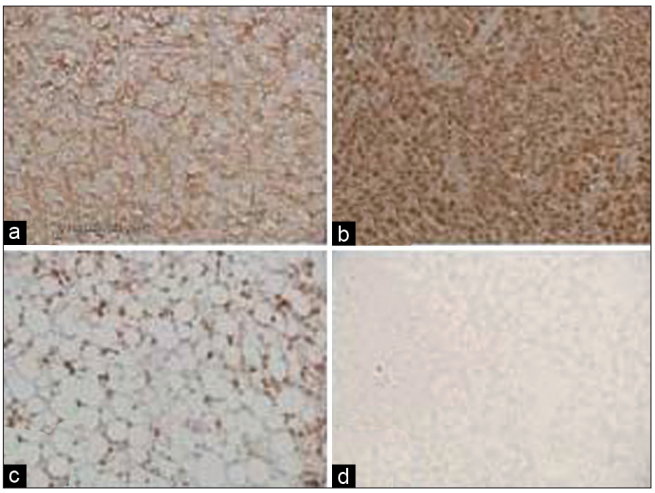

Figure 3: Immunohistochemistry confirming the diagnosis of liponeurocytoma

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