Liponeurocytoma of Cerebellum: rare entity, case based study

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Abstract: Cerebellar liponeurocytoma (LNC) is a very rare neuroepithelial tumor. About thirty eight cases have been reported, in literature till date, mostly in the form of isolated case reports due to rarity of the occurrence. Authors report interesting case in 27-year old male presented with headache and cerebellar signs. MRI imaging revealed posterior fossa mass. He underwent sub occipital ctraniectomy with gross total resection of lesion. Histopathological report of specimen was consistent with liponeurocytoma with range of MIb index less than three. He is under regular follow-up. However he was not advised any adjuvant therapy. Diagnosis of LNC requires high index of suspicion and neuroimaging with typical MRI findings may be help to confirm the diagnosis. The tumor needs to be differentiated from medulloblastoma, which is far more common and aggressive and requires post-operative chemo-radiation and carries poor prognosis Management and pertinent literature will be reviewed.

Key words: liponeurocytoma, imaging, surgical management

Introduction

Liponeurocytoma is a rare neuroepithelial neoplasm involving cerebellum usually seen in adult population. These tumors are slow-growing, runs indolent course. Anatomically cerebellar hemispheres are commonly involved, but may have epicentre in the paramedian region or even vermis (1). It is typically well circumscribed and does not infiltrate surrounding neural tissue, however, may show pressure effect on adjoining anatomical structure, with progress in the size of lesion cause pressure and distortion over fourth ventricle leading to blockage of CSF pathway with development of obstructive hydrocephalus. The patient initially presents with cerebellar signs and as lesion progresses features of raised intracranial pressure also appears. It runs favourable course and surgical management is treatment of choice. It needs to be differentiated from medulloblastoma, where surgical treatments followed by adjuvant chemotherapy constitute an integral part of management due to aggressive

biological behaviour. Microscopically it consists of neurocytic cells with focal areas of differentiated cells resembling mature adipose tissue (2).

Case report

A 27-year old male presented with complaints progressive continuous headache associated intermittent vomiting for four months. He also noticed imbalance while walking two months back prior to admission. For 3 months and gait imbalance for 2 months. Patient did not give any history of systemic complaints on examination. There was no history of any co-morbidity. Visual acuity was normal funfi examination showed bilateral papilloedema, had bilateral cerebellar signs more prominent on left side, and rest of the neurological examination were within normal limit. Routine hematological and biochemical investigations were within normal limits.

Non contrast computed tomogram (NCCT) scan of head revealed hypodense lesion in midline posteror to fourth ventricle with obstructive hydrocephalus (Figure 1). Magnetic resonance imaging (MRI) of brain revealed heterogeneous space-occupying lesion without perilesional edema in vermis and adjoining cerebellar hemisphere more towards left side, with a dimension of 4.4 x 4.3 x 3.9 cm sized causing compression of fourth ventricle. Which was hyperintense on T1W I (Fig- 2a), hypointense on T2W images (Figures 2b&2c) and showed mild patchy enhancement after injection of intravenous gadolinium (Figure 3). In view of feature of raised intracranial pressure and cerebellar signs and atypical radiological findings, patient was planned for surgery.

He underwent midline sub occipital ctraniectomy in the prone position, posterior arch of C1 removed. After dural opening, CSF was released. The tumor was a grayish-yellow, soft, intra-axial lesion, arising from the vermis. It was encapsulated and moderately vascular and CUSA suckable, after securing hemostasis dura was closed primarily and wounds closed in layers. Gross total excision of the tumor was done. The frozen section showed features suggestive of liponeurocytoma.

He had an uneventful postoperative course and was discharged on fifth postoperative day. Post-op stay in the hospital was uneventful and his headache and vomiting subsided. Postoperative computerized tomography (CT) brain did not show any evidence of residual tumor. (Figure 4) Patient was not advised postoperative chemotherapy and radiotherapy and was doing well at last follow-up visit six months after surgery. Histopathological examination of the excised specimen showed a cellular lesion with extensive areas of lipomatous differentiation. The tumor was composed of round to polygonal cells with distinct cellular outline and moderate amount o cytoplasm. No mitosis, necrosis or endothelial proliferation was seen. The tumor cells were immunopositive for synaptophysin, NSE, MAP, and negative for p53. The MIB I labeling index was approximately 3%. The final impression was liponeurocytoma of WHO grade II.

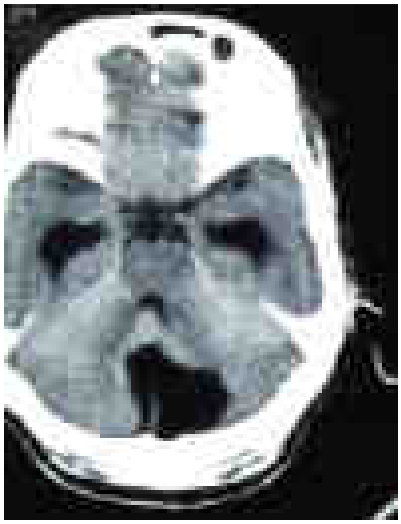
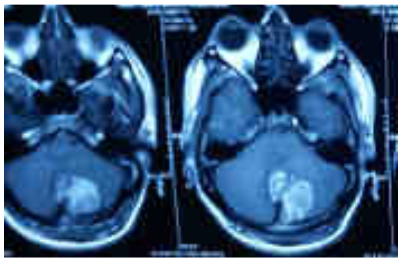


Figure 1 - Non-contrast computed tomography head showing hypodense lesion in midline in the posterior fossa causing obstructive hydrocephalus



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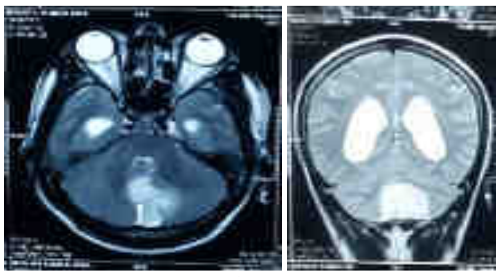

2b 2c

Figure 2 - Magnetic Resonance images showing posterior fossa mass, 2a: T1-Weighted axial scan axial section showing hyperintense, 2b. T2-Weighted axial scan and 2c. T2-Weighted coronal scan showing hyperintense mass

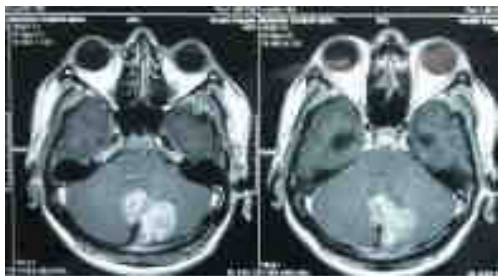

Figure 3 - Contrast-enhanced Magnetic resonance images showing mass in the posterior fossa with heterogeneous contrast enhancement

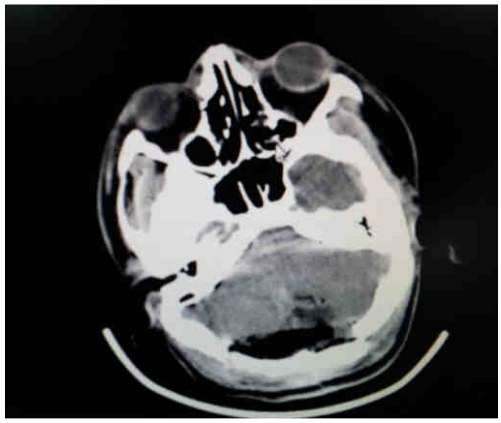

Figure 4 - Post-operative computed tomography scan showing complete excision of lesion

Discussion

Cerebellar liponeurocytoma is an uncommon tumor occurring in adult age group in the cerebellum; it was first described in 1978 by Bechtel (3). It was recognize as a distinct clinical entity and placed as grade II tumor in year 2000, revised WHO classification of CNS tumors in (10). It is a rare tumor occurring in the posteror fossa usually involving cerebellar hemisphere. In a review carried out by de Araújo et al in 2011 reported a total of 32 cases including his one case report (6). Different synonyms has been used in the

literature to denote such tumor lipomatous medulloblastoma(4, 9), loipidized medulloblastoma (5), and others include medullocytoma, lipomatous glioneurocytoma (2) to emphasize similarity to central neurocytoma and differentiate the prognosis from medulloblastoma.

These tumors mostly affect adult population (5) and there is no gender predisposition (2). Patients usually presents with feature of cerebellar signs with raised intracranial pressure as pressure on fourth ventricle lead to obstructive hydrocephalus. Clinical symptoms may range s few months. Neurological evaluations usually show presence of cerebellar symptoms. Pathoanatomically commonly occurs in the cerebellar hemispheres but may have epicentre in the paramedian region or even vermis (1). It is typically well circumscribed but may show pressure effect on adjoining anatomical structure. With advent of blockage of CSF pathway can have obstructive hydrocephalus.

On computed tomography imaging, it may appear heterogeneous isodense or hypodense with respect to brain parenchyma with focal areas of marked hypodensity like to fatty tissue (5). MRI is investigation of choice and appearance is heterogeneous caused by distribution and proportion of loipidized content, which may vary from case to case and even in the same lesion On T1-weighted image is usually heterogeneous iso to hyperintense (1, 5). On contrast administration, it shows usually irregular and heterogeneous minimal enhancement. On T2-weighted slightly hyperintense to the surrounding brain, with focal areas of more pronounced hyperintensity. Peritumoral oedema is may be typically absent or minimal (1, 5).

The optimum mode of treatment of these lesions is gross total surgical excision to aid in diagnosis of lesion and ameliorating feature of raised intracranial pressure. As surgery provides immediate relief in headache and vomiting, however, controversies prevail regarding the ideal adjuvant therapy.

LNC is a neuroectodermal tumor and consists of both glial and neural components. Immunohistochemistry for GFAP, synaptophysin and NSE are usually positive showing origin of mixed glial and neuronal elements. Microscopically, the tumor consists of small, ovoid cells with areas of mature adipocytes4 resembling loipidized cells (2). Mitotic activity is usually absent and MIB-1 labeling index usually lies in the range of 1%-3%. Most of the available pertinent literature shows case reports only and there is no consensus yet on the optimum line of management of this rare entity (1, 2).

LNC has a relatively benign clinical course following surgery (7, 11). Usually surgery is the treatment of choice and radiotherapy or chemotherapy is usually not advocated, when gross total excision has been carried out during primary surgery. In the literature 5-year survival has been reported to be 81%; however recurrence were encountered in 20-32% patients on an average follow-up of 10-years following surgical treatment (11). However, Jenkinson et al reported case which recurred 12 months following subtotal resection with atypical highly aggressive course unresponsive to radiation (8).

Kleihues advocated avoidance of adjuvant therapy is if the MIB-1 index is within the range of 1% to 3% and no residual lesion following surgical decompression and such case carry favorable prognosis (9). There is no report of spinal drop metastasis and hence spinal radiation is not advisable (2). Diagnosis of LNC requires high index of suspicion and neuroimaging with typical MRI findings may be help to confirm the diagnosis. The tumor needs to be differentiated from medulloblastoma, which is far more common and aggressive and requires post-operative chemo-radiation and carries poor prognosis (6).

Conclusion

Cerebellar neurocytoma is a newly acknowledged clinicopathological entity, very rare tumor. Usually surgical excision is the initial mode of treatment. In view of favourable prognosis with probable benign nature of the lesion mandates follow up without any adjuvant chemotherapy or radiotherapy following gross total surgical excision and, post operative scan showing no residual lesion and the MIB-labeling index is less than three percent. Preoperative awareness on basis of neuroimaging can lead to total excision and can obviate need for chemo-radiation adjuvant therapy. Neuroscientist should differentiate it from medulloblastoma, which is far more common in occurrence, runs much aggressive clinical course with repeated recurrence even after post-operative chemo-radiation adjuvant therapy following micro-neurosurgical excision.

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