

Multiple Cerebellar Liponeurocytoma mimicking Left Cerebellopontine Angle and Right Cerebellar Dermoid - A Case Report

Anil Dhar^{1*}, Manish Vaish¹, Rahul Gupta¹, Kapil Jain¹, Mary Abraham², Manish Marda², Shiv Pratap², Vivek Patel¹, Abhay¹ and Arvind Pal Singh¹

¹Department of Neurosurgery Fortis Hospital, Uttar Pradesh, India

²Department of Neuroanaesthesia Fortis Hospital, Uttar Pradesh, India

*Corresponding Author: Anil Dhar, Department of Neurosurgery Fortis Hospital, Uttar Pradesh, India.

Received: July 21, 2015; Published: September 08, 2015

Abstract

Cerebellar liponeurocytoma is a rare WHO grade I or II well differentiated neurocytic tumor of the cerebellum with focal lipomatous differentiation. Mainly reported in adulthood, it is thought to be a posterior fossa benign tumor. We report a case of multiple cerebellar liponeurocytoma which is very rare in a 45 year old female who presented with signs and symptoms of raised intra cranial pressure. We operated the case with pre operative diagnosis of a left CP angle dermoid but histopathology diagnosis was cerebellar liponeurocytoma. The patient was discharged on 4th day with improvement in pre-operative symptoms. The patient is on our follow up.

Keywords: Liponeurocytoma; CP angle; Dermoid; Intracranial pressure

Introduction

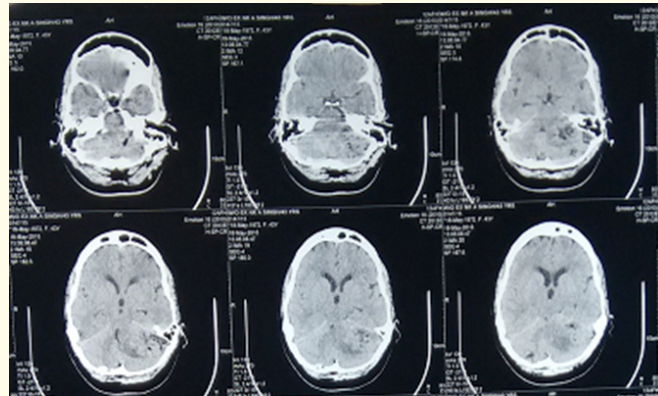
First described by Bechtel and collaborators in 1978, cerebellar liponeurocytomas are posterior fossa lesions composed of densely packed neuronal cells admixed with foci of well differentiated adipocyte like cells. Similar tumors have also been referred to as lipomatous medulloblastoma, lipidized medulloblastoma, medulloctoma, neurolipocytoma, lipomatous glioneurocytoma, or lipidized mature neuroectodermal tumor of the cerebellum [2]. These different appellations reflect the histopathological similarities between liponeurocytomas, medulloblastoma as and central neurocytomas. However, cerebellar liponeurocytoma is genetically distinct from medulloblastoma and central neurocytoma [9]. It is now recognized as a separate entity by the World Health Organization (WHO) and the 2000 classification discouraged use of the term lipomatous medulloblastoma in light of this tumor's advanced neurocytic differentiation and favorable prognosis, particularly in comparison to classic medulloblastoma. Cerebellar liponeurocytoma is currently in vogue because the term most accurately reflects this tumor's pathologic and biologic features. It is classified with the neuronal and mixed neuronal-glial neoplasms in the most recent revision (2007) of the WHO classification of central nervous system tumors [10].

Case Report

We present a 49 year old female who was admitted in our department with one month history of headache accompanied by recurrent episodes of vomiting. Neurological examination did not reveal any other deficit, there was no papilledema on ophthalmological examination. Non contrast and contrast CT head was done which showed well defined heterogeneous mass lesion predominantly hypoattenuating involving left cerebellopontine angle and cerebellar hemisphere measuring 42 x 44 x 32 mm in size with heterogenous enhancement, multiple pockets of fat seen. Another lesion extra axial, well defined, hypo dense 20x18x16 mm is seen in posterior fossa on right side with few pockets of fat attenuating areas within it. This lesion is causing focal compression displacing cerebellum anteriorly. These features were suggestive of ruptured dermoid in left cerebellum and left CP angle and dermoid in right cerebellum (Figure 1a and 1b).

Citation: Anil Dhar, et al. "Multiple Cerebellar Liponeurocytoma mimicking Left Cerebellopontine Angle and Right Cerebellar Dermoid a Case Report". *EC Neurology* 2.1 (2015): 55-60.

1A



1B

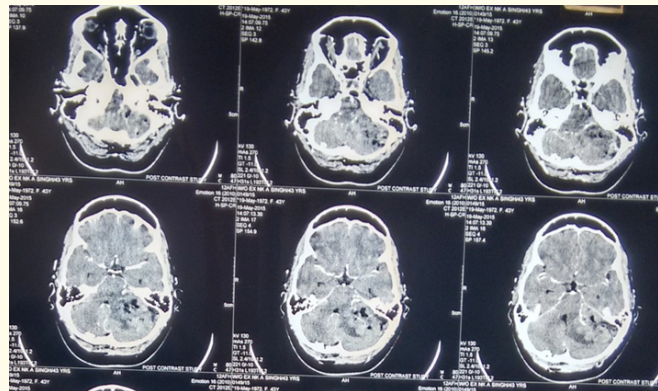


Figure 1A & 1B: NCCT head and contrast CT head was done which showed well defined heterogeneous mass lesion predominantly hypoattenuating involving left cerebellopontine angle and cerebellar hemisphere measuring 42 x 44 x 32 mm in size with heterogenous enhancement, multiple pockets of fat seen. Another lesion extra axial, well defined, hypodense 20 x 18 x 16 mm is seen in posterior fossa on right side with few pockets of fat attenuating areas within it.

MRI Brain Contrast showed evidence of large irregular, well defined extra axial mass involving left CP angle cistern and extending to tentorium and inferiorly up to foramen magnum with mild compression over brain stem, cerebellum and fourth ventricle. Lesion displayed heterogeneous hyperintense signal on T2W images and hypo intense signal on T1W images with heterogenous enhancement on contrast (Figure 2a and 2b). Another heterogenous enhancing lesion was seen in posterior part of right cerebellar hemisphere. An impression of left CP angle dermoid and right cerebellar dermoid was made. After pre anesthetic check up and work up, left retro-mastoid sub occipital craniotomy with excision of tumor was done. Intra operative findings revealed that the tumor was suck able, encapsulated, very vascular extending up to brain stem and tentorium in left CP angle region.

Histopathological features showed a biphasic tumor predominantly comprising of small round cells with round to oval nuclei with foci displaying clear cytoplasm and lipidized cells resembling mature adipose tissue. Intervening thin vascular network seen and there is no evidence of mitosis or necrosis. IHC profile showed synaptophysin positive and GFAP positive cells with ki 67 labelling index of less than 1%, so a diagnosis of left CP angle liponeurocytoma was made (Figure 3a and 3b).

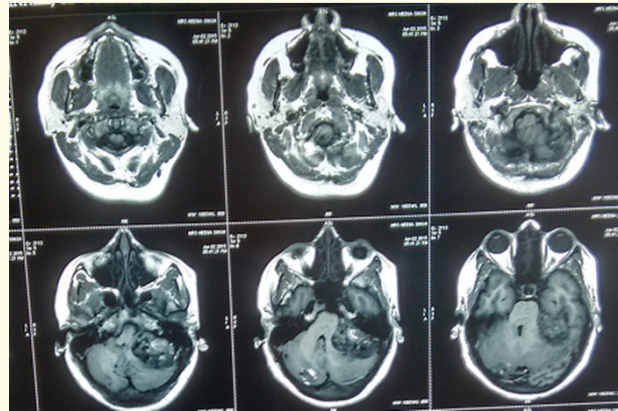


Figure 2A: T1 weighted axial image showing heterogenous signal intensities in left CP angle and posterior part of right cerebellum.

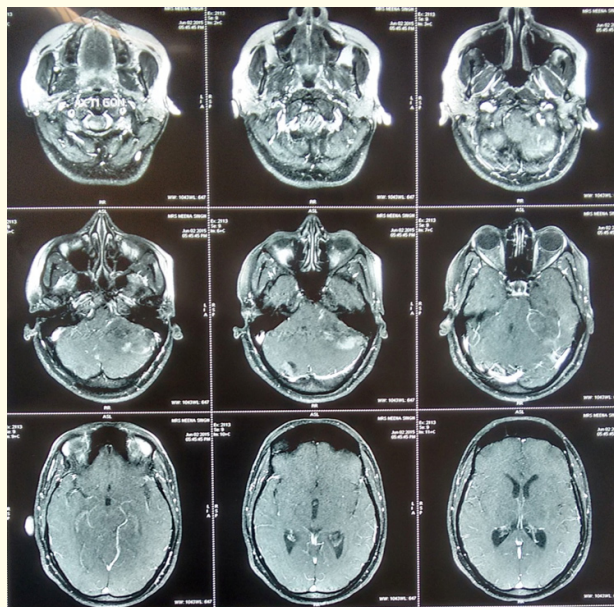


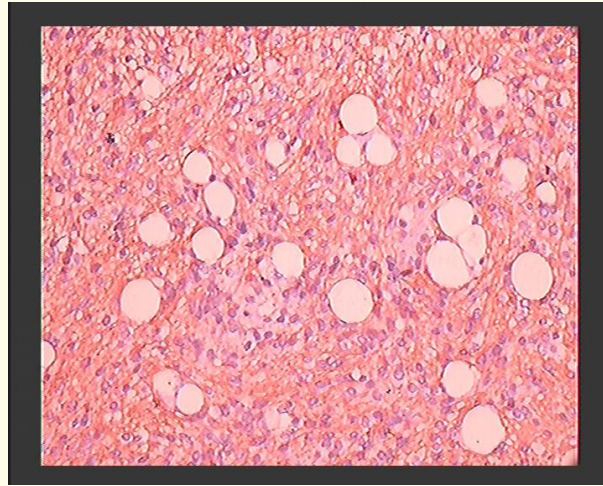
Figure 2B: Contrast axial images showing heterogeneously enhancing lesions in left CP angle and posterior part of right cerebellum

Discussion

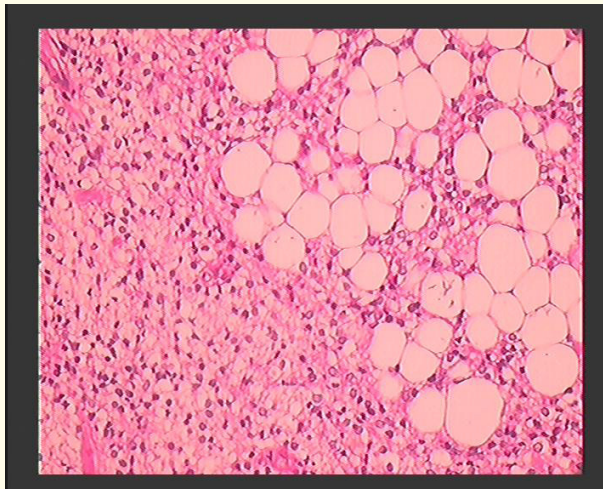
Cerebellar liponeurocytoma is a rare neoplasm with distinctive morphologic features. It typically involves the cerebellar hemispheres of middle-aged to older adults. The tumor is composed of a uniform population of neurocytic cells possessing round to oval nuclei and pale to clear cytoplasm. A variable degree of lipidization of the tumor cells is present, lending a resemblance to mature adipose tissue. Immuno histochemistry serves to confirm the neurocytic differentiation of the tumor cells. In the 2007 revision of the WHO classification of central nervous system tumors, cerebellar liponeurocytoma was reclassified as a grade II neoplasm to reflect a higher recurrence rate than was previously appreciated [11].

Citation: Anil Dhar, *et al.* "Multiple Cerebellar Liponeurocytoma mimicking Left Cerebellopontine Angle and Right Cerebellar Dermoid a Case Report". *EC Neurology* 2.1 (2015): 55-60.

3A



3B



Clinical Features

More than 40 cases of cerebellar liponeurocytomas have been reported in literature [12]. Patients usually present between the ages of 58 to 60 years [13]. The mean age is approximately 53 years. That is in contrast to classic medulloblastoma, which typically affects children and young adults. Men and women are affected equally. Presenting signs and symptoms are most often related to increased intracranial pressure, including headache, vomiting and altered consciousness. Other manifestations include dizziness, unsteadiness, gait disturbance, frequent falls, visual symptoms, and other signs of cerebellar or brainstem dysfunction [12,13].

Radiographic features

Cerebellar liponeurocytoma most commonly involves the cerebellar hemispheres but may be located in the paramedian region or vermis and may extend to the cerebellopontine angle or fourth ventricle. Cerebellar liponeurocytoma is typically well circumscribed but may show mass effects on adjacent structures (Eg., fourth ventricle). On computed tomography scans, the tumor is variably iso-dense or hypo-dense relative to brain parenchyma with focal areas of marked hypo attenuation corresponding to fat density [14]. On T1-weighted

magnetic resonance imaging images, the tumor appears iso-intense to hypo intense, with patchy areas of hyper intensity corresponding to regions of high lipid content.

Contrast enhancement is often heterogeneous and may be minimal. On T2-weighted magnetic resonance images, the tumor appears slightly hyper-intense to the surrounding brain, with focal areas of more pronounced hyper intensity. Peri-tumoral edema is typically absent or minimal. Fat-suppressed images may be helpful in supporting a preoperative diagnosis of liponeurocytoma [14].

Treatment

Whenever feasible, the standard treatment of cerebellar liponeurocytoma is complete surgical resection. Patients may have prolonged recurrence-free survival following surgical resection alone. In light of that, the WHO 2000 working group sought to distinguish liponeurocytoma from medulloblastoma to avoid aggressive and potentially unnecessary adjuvant therapy.[15] Radiation therapy may still have a role, given the potential for local recurrence. Due to the paucity of long-term follow-up data, it is unclear whether radiation should be given in the immediate postoperative period or be reserved for recurrent tumor, which may occur many years following initial surgery [16,17].

When administered, radiotherapy is generally limited to the posterior fossa [18]. No cases of spinal drop metastasis from cerebellar liponeurocytoma have been reported to date. Recurrences are not uncommon, up to 40% in one study [19].

We have presented a case of cerebellar liponeurocytoma with characteristic radiological and histopathological features with one exception that in our case we had a separate satellite lesion in right cerebellum. This type of multiple liponeurocytomas are very rare and only one case has been reported in literature till now [20].

Conclusion

CP angle Liponeurocytoma is a rare tumor in posterior fossa seen commonly in adulthood. Radical resection is the treatment of choice.

Bibliography

1. Bechtel JT, *et al.* "Mixed mesenchymal and neuro-ectodermal tumor of the cerebellum". *Acta Neuropathologica* 15.41 (1978): 261-263.
2. Aker FV, *et al.* "Cerebellar liponeurocytoma/lipidized medulloblastoma". *Journal of Neuro-Oncology* 71.1 (2005): 53-59.
3. Gonzalez Campora R and Weller RO. "Lipidized mature neuroectodermaltumour of the cerebellum with myoid differentiation". *Neuropathology and Applied Neurobiology* 24.5 (1998): 397-402.
4. Orlandi A, *et al.* "Lipomatous medulloblastoma". *Clinical Neuropathology* 16.4 (1997): 175-179.
5. Soylemezoglu F, *et al.* "Lipomatous medulloblastoma in adults. A distinct clinical pathological entity". *The American Journal of Surgical Pathology* 20.4 (1996): 413-418.
6. Giangaspero F, *et al.* "Medullocytoma (lipidized medulloblastoma) A cerebellar neoplasm of adults with favorable prognosis". *The American Journal of Surgical Pathology* 20.6 (1996): 656-664.
7. Davis DG, *et al.* "Lipidized medulloblastoma in adults". *Human Pathology* 24.9 (1993): 990-995.
8. Chimelli L, *et al.* "Lipomatous differentiation in a medulloblastoma". *Acta Neuropathologica* 81 (1991): 471-473.
9. Horstmann S, *et al.* "Genetic and expression profiles of cerebellar liponeurocytomas". *Brain Pathology* 14.3 (2004): 281-289.
10. Kleihues P, *et al.* "The WHO classification of tumors of the nervous system". *Journal of Neuropathology & Experimental Neurology* 61.3 (2002): 215-225; discussion 226-229.
11. Todd Nishimoto MD and Brock Kaya MD. "Cerebellar Liponeurocytoma". *Archives of Pathology & Laboratory Medicine* 136 (2012): 965-969.
12. Patel N, *et al.* "Cerebellar liponeurocytoma". *Canadian Journal of Surgery* 52.4 (2009): E117-E119.

13. Jackson TR., *et al.* "Cerebellar liponeurocytoma: case report and review of literature". *Journal of Neurosurgery* 95.4 (2001): 700-703.
14. Alkahdi H., *et al.* "Neuroimaging of cerebellar liponeurocytoma: case report". *Journal of Neurosurgery* 95.2 (2001): 324-331.
15. Kleihues P., *et al.* "The WHO classification of tumors of the nervous system". *Journal of Neuropathology and Experimental Neurology* 61.3(2002): 215-225.
16. Horstmann S., *et al.* "Genetic and expression profiles of cerebellar liponeurocytomas". *Brain Pathology* 14.3 (2004): 281-289.
17. Aker FV., *et al.* "Cerebellar liponeurocytoma/lipidized medulloblastoma: case report and review of the literature". *Journal of Neuro-Oncology* 71.1 (2005): 53-59.
18. Jackson TR., *et al.* "Cerebellar liponeurocytoma: case report and review of literature". *J Neurosurg.* 2001;95 (4):700-703.
19. Kleihues P., *et al.* "Cerebellar liponeurocytoma. In: Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, editors. World Health Organization classification of tumours of the central nervous system, fourth edition". Lyon: International Agency for Research on Cancer 56 (2007): 110-112.
20. Pelz D., *et al.* "Multifocal Cerebellar Liponeurocytoma". *Canadian Journal of Neurological Sciences* 40.6 (2013): 870-872.

Volume 2 Issue 1 September 2015

© All rights are reserved by Anil Dhar., *et al.*