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Review Article

UNUSUAL PRESENTATION OF AN UNUSUAL CEREBELLAR MASS: DYSPLASTIC CEREBELLAR GANGLIOCYTOMA/LHERMITTE DUCLOS DISEASE-REVIEW OF LITERATURE AND REPORT OF 2 CASES

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ABSTRACT

Lhermitte-Duclos disease or dysplastic gangliocytoma of the cerebellum is a rare benign unilateral mass of the cerebellar cortex having unique diagnostic MRI features characterized by classic 'tiger striped' appearance. Correct recognition of the disease is particularly important given the possible correlation with Cowden syndrome and therefore the need to identify concurrent malignancies. MRI is a considered diagnostic and is helpful in distinguishing LDD due to its unique imaging features. Two biopsy proven cases of this unusual disease were reported with atypical presentations. Atypical presentations include- presence of diffusion restriction in the dysplastic left cerebellar hemisphere in one case and involvement of bilateral cerebellar hemisphere and vermis in the second one.

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INTRODUCTION

Dysplastic gangliocytoma/LDD is a rare benign condition of uncertain nature involving the cerebellum and first described by Lhermitte and Duclos in 1920¹. Approximately 220 cases of this disease was reported till 2006². Later many more additions were made. It is an unusual hamartomatous lesion characterized by slowly enlarging mass within the cerebellar cortex. The lesion may be part of a syndrome and associated with congenital malformations. Microscopically the tumour is composed of granule, Purkinje and glial cells and the histological architecture of the cerebellar cortex is reproduced within the lesion in disorganized fashion². It is characterized by overgrowth of cerebellar ganglion cells which replace granular cells and Purkinje cells resulting in gross thickening of cerebellar folia. Correct diagnosis of this entity is important as it can be a part of familial Cowden disease. The possibility of preneoplastic states in Cowden syndrome stresses the importance of diagnosis. MR imaging is the modality of choice, as it is for any posterior fossa abnormality. MR imaging reveals a cerebellar mass with a typical striated, corduroy, or tiger-striped folial pattern that consists of alternating bands on both T1- and T2-weighted images. Diffusion MR shows a hyperintense area at the level of the lesion, due to residual T2 contrast. In the Apparent

diffusion coefficient map, the mass appears isointense to the cerebellar parenchyma since the percentage of free water does not vary. T2 hyperintensity is due to morphological alterations of the cells making up the internal molecular layer associated with atrophy of the cerebellar white matter, thus confirming the dysplastic and malformative origin of LDD³. We hereby add 2 further cases of this rare entity with atypical imaging findings.

METHODS

The study was conducted in department of radiodiagnosis at a tertiary care hospital over a period of 6 months from June 2016 to December 2016. Ethical approval for this study was obtained from local ethics committee. The patients included in the study were those who were referred for evaluation of suspected intracranial mass lesion. Review of literature was done and 2 cases were studied using conventional MRI sequences with diffusion imaging using 1.5 T machine. Histopathological correlation was done. Patients were followed up for an year post surgery.

RESULTS

Cases

1st Case- the first patient is a 9 yr old child who complained of left sided weakness, occipital headache, blurring of vision

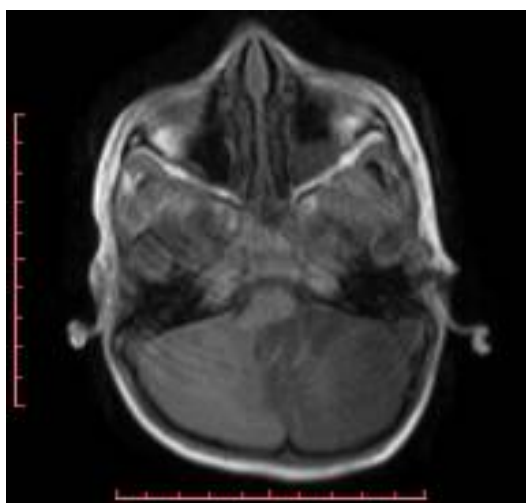
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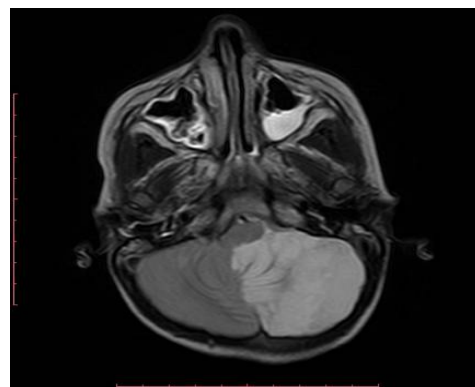
and progressive respiratory distress. On examination, signs of increased intracranial pressure and abnormal gait were observed. On suspicion of intracranial lesion, the patient was referred to our department for further evaluation. Her previous history was unremarkable. A CT scan was done which showed triventricular hydrocephalus with periventricular ooze due to compression of fourth ventricle by a large left cerebellar non enhancing mass. The MRI (1.5 T) revealed left cerebellar expansion with altered signal intensity-T1 predominantly hypointense with prominent iso - hyperintense striations. T2 hyperintense with preserved isointense striations. On FLAIR, striations were getting suppressed in the background of hyperintensity differentiating it from cerebellitis. Possibility of subacute infarct was ruled out as gyral pattern was maintained and lesion did not follow any characteristic arterial distribution. DWI shows restricted diffusion with no significant post contrast enhancement. The lesion was showing mass effect in the form of compression of left medulla and fourth ventricle with upstream dilatation of lateral and third ventricles and periventricular subependymal ooze. Patient died within a week of radiological diagnosis. Autopsy was carried out and histological examination of the specimen showed classic changes of LDD.

2ND CASE - 2nd patient was a 45 yr old middle aged man who presented to the OPD with complaints of giddiness and ataxia. On examination, cerebellar signs were demonstrated. MRI showed an expansile lesion in the vermis and left cerebellar hemisphere that was typically hypointense on T1 and hyperintense on T2 weighted sequences. The mass was characterized by parallel hyperintense striations also called as tiger-striped with vermis and bilateral hemispheric expansion (predominantly left side) and preserved gyral pattern. No enhancement noted on post contrast images. Bilaterality of the lesion was unusual finding as LDD is typically a unilateral lesion⁵. Within a week of imaging, surgical resection of cerebellar dysplastic vermis, left cerebellar hemisphere and part of right cerebellar hemisphere was performed. Postoperative course was uneventful.

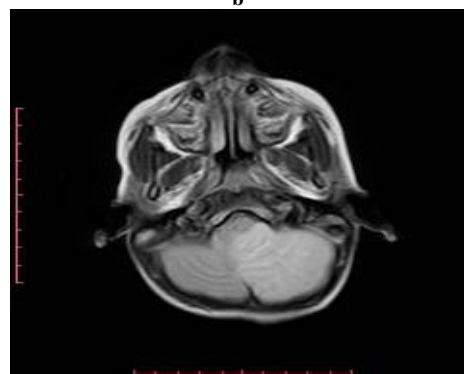
Case 1



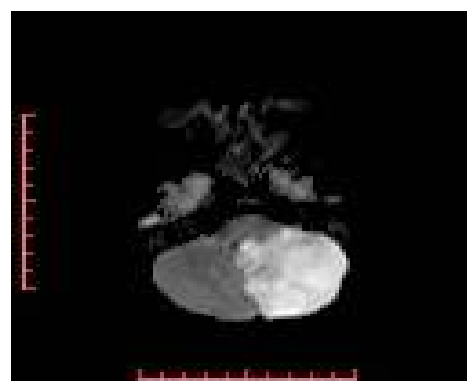
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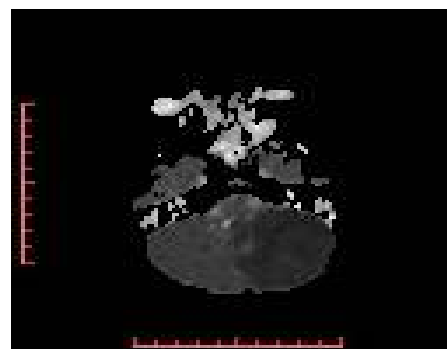
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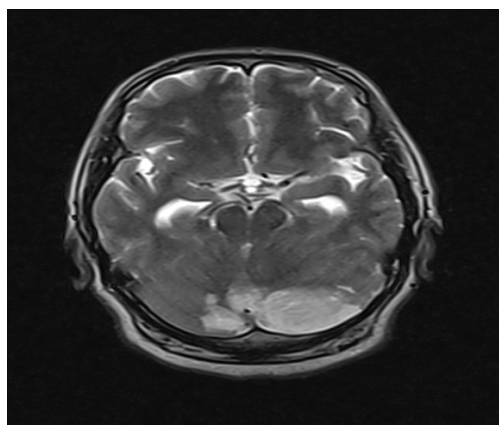


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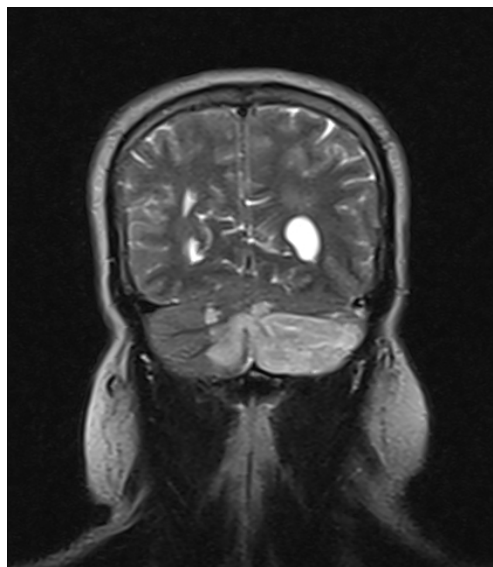
Fig 1

a) T1 weighted axial MR image. The mass is predominantly hypointense with prominent hyperintense striations located in the left cerebellar hemisphere causing mass effect on the left medulla. 4th ventricle is compressed hence not adequately depicted. Triventricular obstructive hydrocephalus was present.
 b) T2 Weighted axial image. The mass is predominantly hyperintense lesion with typical alternate high and normal signal intensity bands.
 c) Axial Fluid attenuated inversion recovery sequence shows that the mass is not suppressed whereas the striations are getting suppressed
 d,e) DWI and ADC images shows increased signal intensity on DWI with corresponding decreased ADC values hence causing true restriction. (*atypical)

Case 2



a



b

Fig 2

a, b) T2 weighted axial and coronal MR image shows hyperintense mass with parallel striations giving tigroid appearance. Mass lesion is present in left cerebellar hemisphere, vermis and part of right cerebellar hemisphere confirming bilateral involvement (*atypical) with compression of 4th ventricle and upstream dilatation of bilateral lateral ventricle and 3rd ventricle

DISCUSSION

Dysplastic cerebellar gangliocytomas are seen most frequently in young adults aged 30 - 50 years (average age, 34 years). Less frequently, it occurs in pediatric patients. May occur in infants and adults over 60 years. There is no sex predilection. There is considerable controversy over the cause of this disease: it may have a hamartomatous, neoplastic, or congenital malformative origin. The condition is often associated with different types of malformation such as macrocephaly, megalencephaly, syringomyelia, polydactyly, multiple haemangiomas and mucocutaneous lesions as well as breast, thyroid, genitourinary and gastrointestinal malignancies. There has been suggested a genetic correlation between LDD and Cowden's syndrome, an autosomal dominant syndrome characterized by a genetic aberration. Eighty percent of patients with Cowden's syndrome have germline mutations in the PTEN gene at locus 10q23.2, which has been identified as the major susceptibility gene for Cowden's syndrome. Most LDD patients appear to

have a germline loss of the PTEN allele and go on to some point, thereby allowing abnormal growth of the granule cells.

Clinically, patients may be asymptomatic, or they may present with symptoms of ataxia, headache, cranial nerve palsy, paroxysm of vertigo, psychic deterioration and, in severe cases, signs and symptoms of intracranial hypertension secondary to hydrocephalus. Usually, patients have long - standing symptoms that have been present for years, indicating the slowly progressive nature of this disease.

Imaging plays an important role in the diagnostic process. The dysplastic gangliocytoma is hypoattenuated on unenhanced computed tomographic (CT) images. In such cases, the only diagnostic clue may be the mass effect, which manifests as compression of the fourth ventricle, effacement of the cerebellopontine angle cistern and hydrocephalus. No appreciable enhancement is in contrast enhanced CT images. However, CT remains of limited value because of beam hardening artifacts caused by petrous temporal bone that obscure the details.

MR imaging is the modality of choice. MR reveals a cerebellar mass alternating bands on both T1- and T2-weighted images. The bands are hyper- isointense relative to gray matter on T2 - weighted images and iso - hypointense on T1 - weighted images. Calcification is an uncommon finding, but it has been reported. Most dysplastic gangliocytomas do not enhance; however, enhancement has been reported and is probably due to the presence of anomalous veins. Mass effect is common and causes compression of the fourth ventricle and occlusive hydrocephalus. Syringohydromyelia may be present. MR spectroscopy was not performed in these patients; however, this modality reveals reduced N-acetylaspartate-choline and N-acetylaspartate-creatine ratios compared with those of normal cerebellar tissue. Lactate peak may also be present.

It has been suggested that neurologic imaging is sufficient for diagnosis of this condition. Although a case of medulloblastoma mimicking dysplastic gangliocytoma on neurologic imaging has been reported in a pediatric patient, diagnostic confusion with medulloblastoma is unlikely in most patients because of differences in age group, medical history, and usual imaging features.^{4,6-10}

Decompression of the ventricular system is the immediate goal of therapy in virtually all symptomatic patients. A ventricular shunt is placed initially and followed by tumor resection. However, complete resection may be difficult because of poorly defined margins. Most patients have an uneventful postoperative period, although recurrence is possible. Long-term follow-up is indicated.³

CONCLUSION

Our cases confirm the widely accepted view that conventional MRI with diffusion imaging is the most useful technique in the study of Lhermitte Duclos disease. They are able to demonstrate pathognomonic features such as thickened cerebellar folia suppressing on FLAIR sequence thus differentiating it from cerebellitis. Secondly, the mass lesion has maintained folial pattern (as they expanded and thickened but not lost) and does not follow arterial distribution thus differentiating it from subacute

infarct . Futhur more, our cases showed some unusual features of this unusual disease like, diffusion restriction and bilateral cerebellar hemisphere involvement both of which are rare presentations as LDD is typically a unilateral lesion of cerebellum showing no diffusion restriction on DWI images.

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