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Case Report

LHERMITTE-DUCLOS DISEASE

***Prateek S. Gehlot[1] and Mohit Agarwal[2]**
[1]Department of Radio-Diagnosis, R.D. Gardi Medical College, Ujjain (MP)
[2]Department of Radio-Diagnosis, R.D. Gardi Medical College, Ujjain (MP)
**Author for Correspondence*

ABSTRACT
Lhermitte-Duclos disease is a rare dysplastic gangliocytoma of cerebellum. It is usually manifested with features of raised intracranial pressure or discovered incidentally during imaging for unrelated conditions. The natural history of disease is not well established. Magnetic Resonance Imaging is the diagnostic modality of choice, demonstrating characteristic thickened cerebellar folia giving a laminated or striated appearance. Surgical treatment of dysplastic gangliocytoma provides a good prognosis in most cases, although local recurrence of disease has been described. In this paper we report a case of Lhermitte-Duclos disease diagnosed at age of 24 years, presenting with progressive occipital headache.

Key Words*: Gangliocytoma, Lhermitte-Duclos, Cowden's Disease, Hydrocephalus*

INTRODUCTION
Lhermitte-Duclos Disease (LDD), also termed as diffuse hypertrophy of the cerebellar cortex and dysplastic cerebellar gangliocytoma was first reported by Lhermitte and Duclos in 1920. Typically, LDD presents in young adults with features of raised intracranial pressure while cerebellar signs being much less prominent. Neuroimaging with Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) demonstrates abnormal laminated patterns of cortical architecture. Although alternating isodense and hypodense layers are discernible on CT scan, the architecture of the lesion is more clearly evident on MRI. LDD may exist in isolation or it may be associated with megalencephaly, heterotopia, microgyria, polydactyly, partial gigantism, macroglossia, multiple visceral hamartomas and neoplasm.

CASE REPORT
A 24-year-old man visited our out patient Department of Medicine with a 3 year history of progressive occipital headache occasionally associated with nausea, vomiting and blurring of vision. CT scan was prescribed and it showed an ill-defined hypodense lesion of the left cerebellum with significant mass effect leading to compression of fourth ventricle and aqueduct of Sylvius resulting in obstructive hydrocephalus (Fig 1). Since the CT findings were inconclusive MRI scan was prescribed which revealed a hyperintense expansile lesion on T2 weighted images with hyperintense and isointense laminations (Fig 2) and hypointense on T1 weighted images with no significant enhancement on Gadolinium enhanced T1W images (Fig 3). The lesion appeared faintly hyperintense on diffusion weighted image (DWI) and apparent diffusion coefficient (ADC) map (Fig 4) suggestive of no restricted diffusion. The lesion was causing compression of pons and right cerebellar hemisphere with hydrocephalus as seen on CT scan. There was minimal cerebrospinal fluid (CSF) ooze with effacement of sylvian fissures and sulci of both cerebral hemispheres.

DISCUSSION
Lhermitte-Duclos disease is a rare cerebellar dysplasia that is characterized by focally indolent growth of cerebellar cortex in which folia enlarge due to profusion of dysplastic cortical neurons that effaces sulci and asymmetrically expand cerebellar hemisphere. LDD can occur in isolation or may be associated with neurocutaneous syndrome, Cowden's disease (multiple hamartoma syndrome). Other associations include megalencephaly, polydactyly, local gigantism, heterotopias and cutaneous hemangiomata.

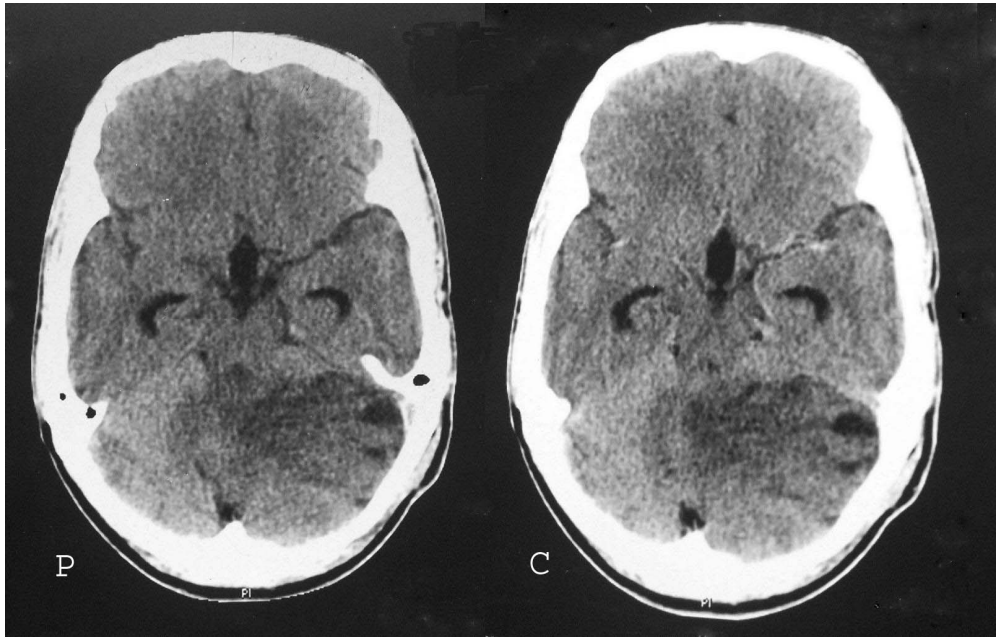

Figure 1: Plain and contrast enhanced axial CT images showing hypodense non enhancing mass in left cerebellar hemisphere.

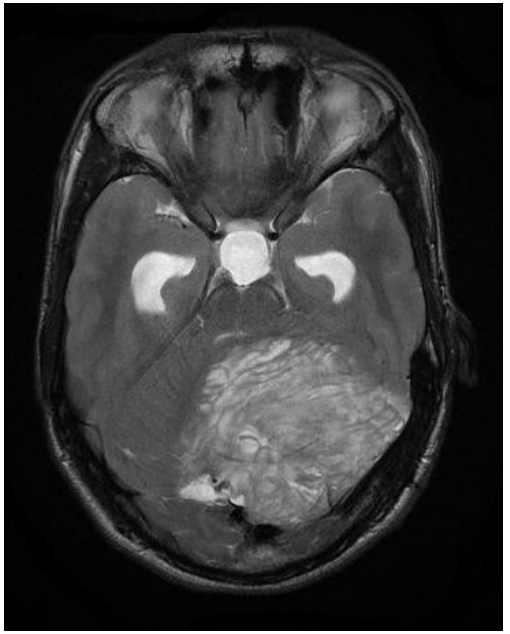

Figure 2: T2W axial images reveal mass in left cerebellar hemisphere with hyperintense and isointense striations causing compression of pons and 4th ventricle resulting in hydrocephalus.

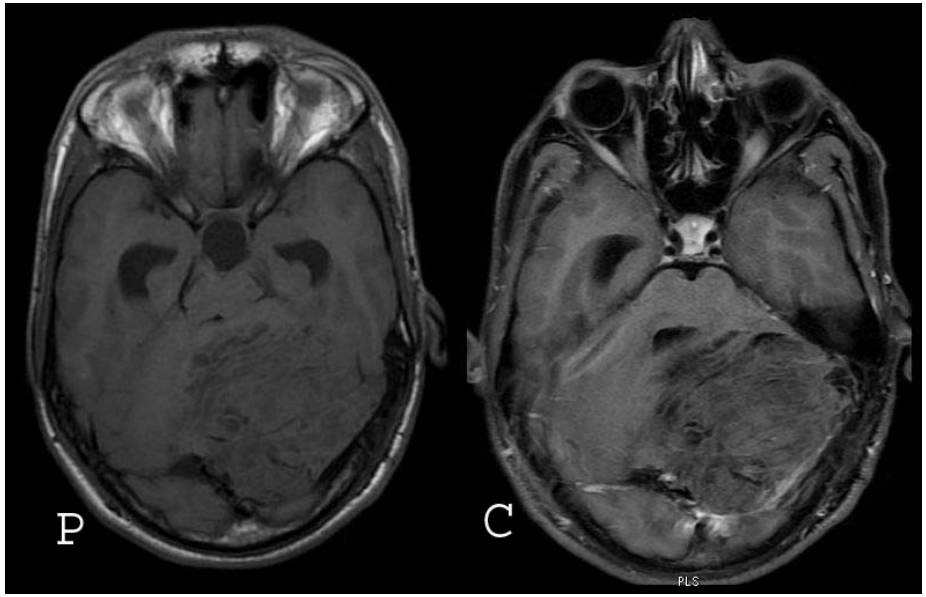

Fig 3: Plain (P) and contrast enhanced (C) T1W axial sequences revealing heterogeneously hypointense mass with no enhancement.

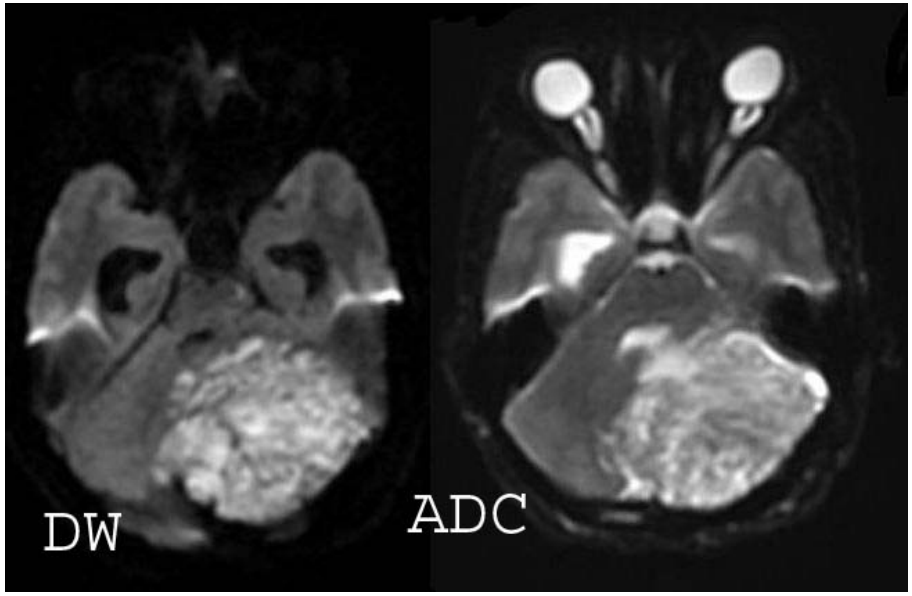

Figure 4: Lesion in left cerebellar hemisphere appearing hyperintense on DW images and ADC map suggesting no diffusion restriction and is due to T2 shine-through effect.

The exact etiology of LDD is controversial; it is probably a brain malformation and not a true neoplasm [Kulkantrakorn 1997], [Osborn 2009]. Imaging plays an indispensable role and skull radiographs may reveal thinning of the skull in the occipital region suggestive of an adjacent long-standing space occupying lesion. CT shows the presence of a nonspecific poorly delineated hypodense or isodense cerebellar mass which can mimic posterior fossa neoplasm [Osborn 2009]. The inner portion of the folia consisting of the white matter, the abnormal granular cell layer and deep molecular layer was hypodense on CT and hypointense on T1 weighted and hyperintense on T2 weighted images. The outer portion of the folia consisting of the outer molecular layer and leptomeninges within effaced sulci was isodense/isointense on CT and MRI. Rarely, vascular proliferation in the pia and adjacent outer cortex may be associated with calcifications or contrast enhancement [Kulkantrakorn 1997].

MRI is the imaging modality of choice, as it is for any posterior fossa abnormality. MRI reveals a cerebellar mass with a typical striated, corduroy or tiger-striped folial pattern that consists of alternating bands on both T1 weighted and T2 weighted images. Calcification is an uncommon finding, but it has been reported and is better appreciated on CT scan. Most dysplastic gangliocytomas do not enhance; however, enhancement has been reported and is probably due to the presence of anomalous veins [Shinagare 2009]. If contrast enhancement is present, other diagnoses, such as hemangioblastoma and medulloblastoma might be suggested. Hemangioblastomas commonly have enlarged vessels, solid and cystic components and no striations [Klisch 2001]. In adults, medulloblastoma usually have heterogeneous contrast enhancement with restricted diffusion, where as in LDD the lesion appears bright on DW images due to T2 shine-through effect (rather than restricted diffusion) as reflected by lack of hypointensity on ADC mapping [Mittal 2009].

Making the preoperative diagnosis of LDD through MRI obviates the need for biopsy. In symptomatic patients, decompression of the ventricular system is the immediate goal of therapy. A ventricular shunt is placed initially followed by partial/complete resection of mass. Long-term follow-up is advised as some cases of recurrence are reported [Shinagare 2009].

To summarize, a nonenhancing mass in the posterior fossa with unilateral hemispheric expansion, linear striations and characteristic signal intensity pattern on MRI should be considered specific for LDD. A screening for Cowden's disease should be done in every patient. Surgical decompression is the treatment of choice and patient should be kept on follow up as long term recurrence can occur.

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