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RARE PRESENTATION OF LHERMITTE-DUCLOS DISEASE

Radio Diagnosis

Dr. Aarthi Parthasarathy* MD, Department of Radio Diagnosis, Santhi Social Services , Coimbatore , TamilNadu. *Corresponding Author

Dr. Muhil Kannan MD, Department of Radio Diagnosis, Santhi Social Services , Coimbatore , TamilNadu.

ABSTRACT

Lhermitte-Duclos disease (LDD) arises from an uncommon benign lesion which alters the normal cerebellar laminar pattern. This case report shows the imaging findings of this condition in 39 year-old male, presented with history of headache and dizziness for duration of ten days. Intial assessment included a non-contrast CT which showed a mass in left cerebellar hemisphere with surrounding mass effect. MRI multiplanar sequences showed a well defined T2 hyperintense lesion with a tigroid appearance.The lesion showed a mild bright signal on diffusion-weighted images, whereas MR Spectroscopy shows reduced choline levels. Differential diagnosis in terms of imaging include subacute infarct, cerebellitis, glial tumor,encephalitis but these do not give the classical tigroid appearance on MRI. Lack of contrast enhancement , significant diffusion and cellularity of the lesions can help in differentiating among them.

KEYWORDS

MRI , Lhermitte duclos syndrome , neuroradiology

Clinical Findings:
39 year-old male, presented with history of headache and dizziness for a duration of ten days . There was no prior history of migraines and review of systems as well as laboratory data were noncontributory. He had no symptoms of raised intracranial tension including blurred vision, headache and vomiting.

Imaging Findings:
Intial assessment included a non contrast CT which showed a mass in left cerebellar hemisphere with surrounding mass effect. MRI multiplanar sequences showed a well defined T2 hyperintense lesion measuring about 74 x 56 x 64mm with widened cerebellar folia , thickened cortex, minimal restricted diffusion is noted in left cerebellar hemisphere giving striated/ tigroid appearance. It crosses the midline and involves the right cerebellar hemisphere.It causes significant mass effect in the form of compression of fourth ventricle ,displacement of rotation of brainstem towards right. Superiorly the lesion is extending into supratentorial region into the ambient and quadregeminal cistern. The lesionalso extends into the left internal auditory meatus. No significant post contrast enhancement is seen .MR Spectroscopy shows reduced choline levels.

DISCUSSION:
There has been considerable discussion regarding the origin of the lesion as a hamartomatous lesion or true neoplasm. This lesion is currently classified as World Health Organization grade one tumor that is slowly progressive with possibility for recurrence.

Multiple studies have tried to find an association of Lhermitte-Duclos disease with Cowden syndrome, a familial phakamatosis characterized by autosomal dominant inheritance pattern and with varied neuroectodermal manifestations. [1] Histopathogical findings include absence of the Purkinje cell layer, hypertrophy of the granular cell layer, widening of the molecular layer of the cerebellar cortex and replacement by abnormal ganglion cells. [1]

Lhermitte–Duclos disease (LDD) is a rare entity also referred to as dysplastic cerebellar gangliocytoma initially described by two French physicians in 1920 Lhermitte and Duclos [2].

It is commonly seen around 3rd to 4th decade and can be associated with other congenital anomalies. This lesion is commonly seen in the posterior fossa with a nonspecific clinical presentation usually related to cerebellar dysfunction, raised intracranial pressure, and cranial nerve deficits. The clinical presentation for this presentation is varied, ranging from months to more than 10 years. Cerebellar symptoms are not seen consistently .Computedtomographic (CT) in these patients demonstrate a non enhancing posterior fossa lesion, which can be partially calcified ,iso or hypodense [2]

This lesion is commonly diagnosed by magnetic resonance imaging with pathognomic imaging findings [2] Tigroid or folial cerebellar appearance of alternating superficial bands of hyper and hypointensity on T1 and T2 weighted imaging is noted with enlarged cerebellar folia . Diffusion restriction and enhancement are not typically seen. Spectroscopy demonstrates nonspecific findings of decreased NAA and increased lactate, though unlike glial malignancies there is evidence of increased myo-inositol and choline to creatine ratio levels[2]

Differential diagnosis in terms of imaging include subacute infarct, cerebellitis, glial tumor, encephalitis but these do not give the classical tigroid appearance on MRI. Lack of contrast enhancement , significant diffusion and cellularity of the lesions can help in differentiating among them. PNET tumors usually show elevated Cho/Cr ratio on spectroscopy [2]
This benign entity is commonly unreported and frequently associated malignancies can be missed especially in young patients. Symptomatic patients require immediate decompression of the ventricular system with a shunt in order to decrease the mass effect and raised intracranial pressure Tumour resection is likely to follow although complete resection is difficult because of ill-defined margins. Due to possibility of recurrence, long term follow up is indicated. [3]

Figure Origin for all figures were taken from: MRI PACS in Department of Radiology , Santhi Social Services , Coimbatore ,Tamil Nadu

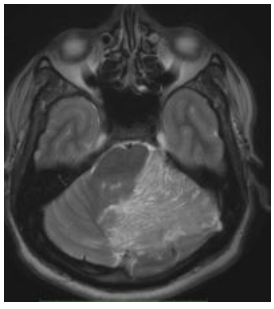

Figure 1 – MRI in Department of Radiology , Santhi Social Services , Coimbatore ,Tamil Nadu

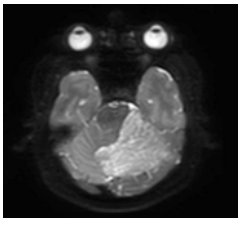

Figure 2 – MRI in Department of Radiology , Santhi Social Services , Coimbatore ,Tamil Nadu

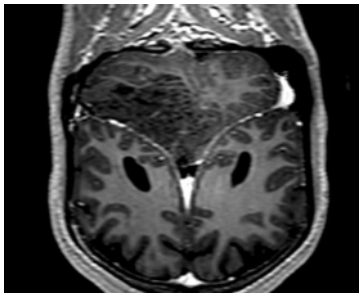

Figure 3 – MRI in Department of Radiology , Santhi Social Services , Coimbatore ,Tamil Nadu

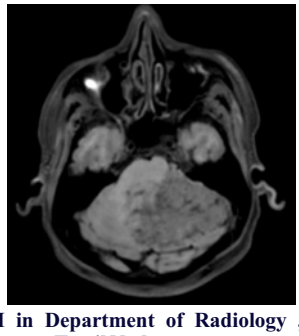

Figure 4 - MRI in Department of Radiology , Santhi Social Services , Coimbatore ,Tamil Nadu