

Lhermitte-Duclos Disease (Dysplastic Cerebellar Gangliocytoma) in a Young Patient

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Abstract

Lhermitte - Duclos disease also called dysplastic gangliocytoma of cerebellum is an extremely rare cerebellar neoplasm. It usually presents with raised intracranial pressure along with cerebellar signs. We report a rare case of Lhermitte - Duclos disease of a 20 years male who presented with signs & symptoms of raised intracranial tension. CT features were suggestive of Lhermitte-Duclos disease. Subtotal excision of the mass followed by histopathological evaluation confirmed the diagnosis. This case has been reported to emphasize clinical presentation and radiological features of this rare entity.

Key words: Lhermitte -Duclos disease, dysplastic cerebellar gangliocytoma, imaging features.

Introduction

Lhermitte-Duclos disease or dysplastic cerebellar gangliocytoma, is a rare neoplasm of posterior cranial fossa arising from the cerebellar cortex. Lhermitte and Duclos in 1920, were first to describes this rare entity [1]. Dysplastic cerebellar gangliocytoma is seen most frequently in young adults i.e. third or fourth decade of life with an average age of 34 years [1,2]. Less frequently, it occurs in pediatric patients [2]. There is no sex predilection. We report a rare case of Lhermitte-Duclos disease in a young male patient.

Case Report

A 20 year young male presented with complains of intermittent occipital headache and diplopia for three months. There was no associated history of vomiting and seizures. Neurological examination revealed an unsteady tandem gait, with tendency to fall on right. Fundoscopy showed no papilloedema. Laboratory investigations were within normal limits. Non-contrast followed by contrast enhanced computed tomography (CT) of head was done. CT scan revealed a well defined hypodense mass of size 4.5×3×3 cm in the left cerebellar hemisphere with prominent and thickened cerebellar folia. There was no evidence of calcification within the mass. The mass was causing compression and distortion of the fourth ventricle

with resultant dilatation of third and bilateral lateral ventricles, suggestive of hydrocephalus. Patient underwent suboccipital craniotomy with subtotal excision of mass. Histopathological features were suggestive of dysplastic cerebellar gangliocytoma. (Figure 1 and 2a, and 2b)

Discussion

Lhermitte-Duclos disease is a rare neoplasm of posterior fossa with incidence of about 5 per million per year [3]. It is also known as granular cell hypertrophy, diffuse hypertrophy of the cerebellar cortex, Purkinjeoma, cerebellar hamartoma, gangliomatosis of the cerebellum, ganglioneuroma, neurocystic blastoma, and hamartomoblastoma [2]. Cerebellar granule neuron is the cell of origin of Lhermitte-Duclos disease. Combination of its aberrant migration & hypertrophy is responsible for its development [4]. Whether this disease is hamartomatous or neoplastic, it is still not clear [5]. If neoplastic, it corresponds histologically to WHO grade I. Malformative histopathological features, absent proliferative activity and absence of progression, favour hamartomatous origin.

Clinically, patients may be asymptomatic. The presenting clinical symptoms are due to mass effect in

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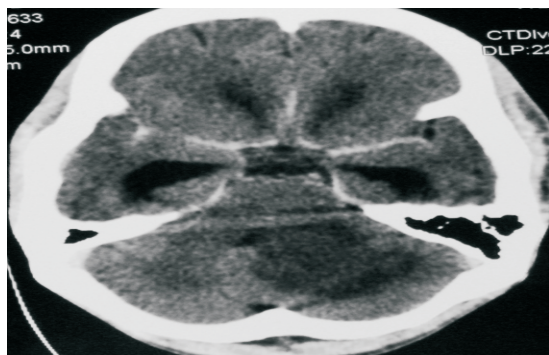


Figure 1. Axial CT image showing hypodense mass in the left cerebellar hemisphere compressing the fourth ventricle resulting in hydrocephalus.

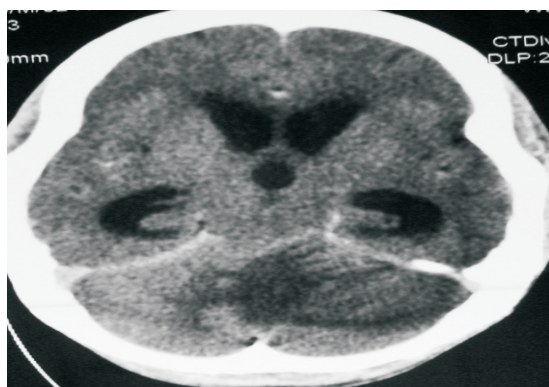


Figure 2 a



Figure 2 b

Figure 2a and 2b. Axial CT image showing hypodense mass in the left cerebellar hemisphere with thickened and prominent cerebellar folia along with hydrocephalus.

the posterior cranial fossa cerebellar dysfunction, hydrocephalus and signs of increased intracranial pressure [2]. Raised intracranial pressure and the long-standing progressive nature of this patient's symptoms argued against acute infarction or cerebellar encephalitis. Mental retardation may be present. This disease is commonly associated with other congenital malformations such as megalencephaly, polydactyly, multiple hemangiomas and skull abnormalities [2,6]. The association of dysplastic gangliocytoma with Cowdens disease is recently been recognized. Cowdens syndrome is an autosomal dominant disease characterized multiple hamartomas and neoplasms affecting the skin, thyroid, breast and genitourinary and gastrointestinal tracts. The skin lesions are present in 90% of patients. The mucocutaneous manifestations form the pathognomonic basis for diagnosing Cowden's disease and consist of features like trichilemmomas, acral keratoses, mucinous fibromas and oral papillomas [7,8].

Grossly the lesion appears poorly circumscribed mass showing thickened, enlarged, firm gyri in contrast to the adjacent normal appearing folia. On histopathology, it is characterized by regional enlargement of the cerebellar stratum granulosum, absence of Purkinje cell layer and progressive hypertrophy of the granular cell neurons with increased myelination [9,10].

The differential diagnoses are ganglion cell tumor, gangliocytoma & ganglioglioma. The classical radiological features along with geographic confinement of ganglion cells to internal granular layer favor the diagnosis of Lhermitte-Duclos disease. Other differentials are cerebellar astrocytoma and hemangioblastoma, medulloblastoma, metastases, etc. Lack of enhancement on the contrast enhanced images helped us rule out leptomeningeal metastases and inflammatory diseases. The "striated appearance" are typical of Lhermitte-Duclos disease [2]. Cerebellar astrocytoma and hemangioblastoma manifest as cystic tumors with an enhancing nodule. In addition, cerebellar astrocytoma and medulloblastoma usually manifests in pediatric patients. Metastases are common in older patients (from the 5th decades onward), tend to be multiple, and may be hemorrhagic [2].

On CT scan, Lhermitte-Duclos disease appears as hypodense non-enhancing mass in cerebellar hemisphere with enhancing and thickened folia. The mass may extend to the midline, distorting the fourth ventricle resulting in supratentorial hydrocephalus. Peripheral calcifications have also been mentioned. Angiography shows an avascular tumour [11]. Magnetic resonance imaging (MRI) is the modality of choice. MRI reveals a non-enhancing cerebellar mass with a typical "Striated or Tiger striped" folial pattern due to alternate hyperintense & isointense bands on T2W images (2,4,14), corresponding to the thickened cerebellar folia [11,12,13]. The mass appears hypointense on T1-weighted sequences. MRI is excellent in defining the limits of the lesion to accomplish the most radical excision during surgery. Although MRI provides accurate morphological examination, Positron emission test (PET) could provide a better primary diagnosis of brain tumors, identify a molecular target for therapy and assess the response to therapy [15].

Despite the benign nature, surgical excision is the treatment of choice, although some centres may prefer conservative management in asymptomatic cases. Recurrence of up to 20 years has been reported after surgical resection [2,13]. Total resection may not be possible in all cases due to absence of a cleavage plane between lesion & surrounding normal brain parenchyma. It may be the cause for recurrence noted in some cases [2,13,16]. Radiation therapy is ineffective. Long-term follow-up is indicated.

Conclusion

This case has been reported to emphasize that Lhermitte-Duclos disease or dysplastic gangliocytoma is a rare tumour of posterior cranial fossa. Due to its classical radiological appearance on CT and MRI, the diagnosis can be easily made. MRI provides valuable information regarding location and extent of the lesion. Surgical removal is the only effective treatment.

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