

## Clinical Image

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# Lhermitte-Duclos disease: A case report

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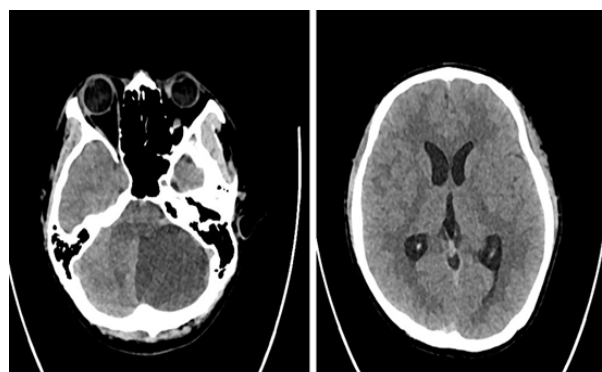
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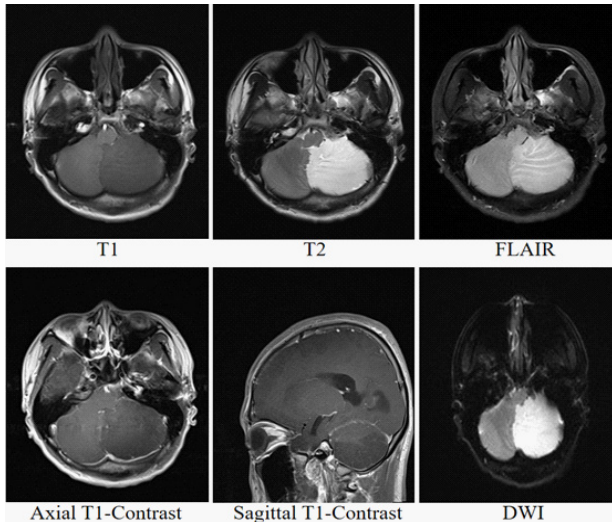
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## Description

A 25-year-old young man came to the emergency department of our hospital with sudden dizziness and vomiting for one day. The neurologist performed a physical examination of the patient, found no positive signs, and ordered the patient to perform a head Computed Tomography (CT) examination, which showed a left cerebellar hemisphere mass with obstructive hydrocephalus (Figure 1). After admission, the brain Magnetic Resonance Imaging (MRI) was performed, and the results showed that the left cerebellar hemisphere showed low signal in T1, hyperintense signal in T2, FLAIR and DWI, and no significant enhancement of cerebellar lesion on T1-contrast scan (Figure 2). The pathological results of postoperative cerebellar specimens showed focal hyperplasia of neuronal cells in the cerebellar granule cell layer, Syn (synapsin) (+), NF (neurofilament) (+), GFAP (glial fibrillary acidic protein) (-), Ki67 (<1%). Pathology considered dysplastic cerebellar gangliocytoma.



**Figure 1:** The CT scan of the patient. CT shows a low-density mass in the left cerebellar hemisphere and a squeeze of the fourth ventricle. The lateral and third ventricles are dilated because of obstruction.



**Figure 2:** The MRI of brain. MRI shows characteristic “tiger stripes” because of high signal of cerebrospinal fluid during loose gap. The lesion is without enhancement.

### Discussion

Lhermitte-Duclos Disease (LDD), also called dysplastic cerebellar gangliocytoma, is a slow-progressing, rare benign cerebellar mass with features of hamartomas and true tumors, belonging to neuronal and mixed neuron-glioma tumors [1]. LDD is closely related to Cowden syndrome, an autosomal dominant disorder caused by the PTEN (Phosphatase and tensin homolog) gene germline [2]. The cerebellar granule cell layer contains densely stained granule cells and Golgi cells. In the pathological state, the granule cell layer and the Purkinje cell layer are replaced by large abnormal neurons, and the molecular layer is myelinated and widened, and abnormally parallel myelinated fibers appear [3]. Pathologically, neuronal related immunological indicators such as Syn, NSE (neuron-specific enolase) and NF are positive due to the presence of neuronal cells in the cerebellar granule cell layer. LDD has low proliferative activity, so Ki67 is negative [4]. LDD occurs more often in young people, and its symptoms progress slowly, mainly manifested as cerebellar dys-

function, such as dizziness, vomiting and unsteady walking [5]. A characteristic “tiger stripes” may appear on MRI, because the atrophy of cerebellar central white matter forms a loose gap that makes the cerebrospinal fluid signal appear streaked. Because of the poor blood supply, the lesion is not significantly enhanced on contrast scanning [6]. LDD is mainly treated surgically, and the prognosis is generally acceptable [7].

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