Lhermitte-Duclos disease (LDD), or dysplastic gangliocytoma of the cerebellum, is a rare hamartomatous lesion of the cerebellar cortex which may be associated with Cowden's syndrome and the PTEN germline mutation. In this report, we describe a case of pathology proven LDD. The clinical presentation, radiological features and histopathologic findings are described, and the relevant literature is reviewed. Axial T1-weighted MR image; the mass lesion in the left cerebellar hemisphere was predominantly hypointense and had typical prominent iso-hypointense striations on T1-weighted images.