Imaging in Lhermitte: Duclos disease

Dear Editor,

Lhermitte–Duclos disease (LDD) is a benign brain tumor of the third and fourth decade.[1] 221 cases of this disease have been reported in the medical literature[2] so far. The disease is characterized by abnormal development of the cerebellum, with diffuse hypertrophy of the stratum granulosum likely to be caused by mutations of the *PTEN* gene.[3] It is often associated with *Cowden syndrome* where it is pathognomonic for the syndrome.

Our patient, a 63-year-old male, known hypertensive on medication, presented with severe headache, acute neck pain and vomiting was referred for imaging. The basis to diagnose LDD, keeping the remote options of differential diagnoses[1] on Magnetic Resonance Imaging (MRI), was the nonenhancing cerebellar mass with unilateral hemispheric expansion, hypo intense on T1-weighted images (T1WI) and hyper intense on T2-weighted images (T2WI), with parallel linear striations or 'tiger- striped' appearance [Figure 1a and b] on the surface of the lesion.[1] The atrophic white matter, the adjacent layers of abnormal ganglionic neurons and the inner most part of the molecular layers had prolonged signal (hypo intense on T1WI and hyper intense on T2WI and FLAIR). There was indentation of brain stem by the mass, effacement of fourth ventricle, and minimal tonsillar herniation.

The morphologic features of the LDD may be demonstrated most clearly with the short TI inversion recovery (STIR) sequence and the turbo inversion-recovery magnitude (TIRM) sequence. Areas of calcifications, if present, will be depicted on susceptibility-weighted imaging (SWI). Apparent diffusion coefficient (ADC) mapping may show no disturbance of water diffusion since the signal on diffusion-weighted images (DWI) depends on cell density, low extra cellular water content, and enhancement of the tumor. As compared with the TIRM sequence, the ADC mapping is helpful in postoperative control, because it delineates tumor from surgical resection margins, whereas on T2WI, differentiation between both components could be difficult. Contrast enhancement is not a typical sign of LDD.[4,5] If contrast enhancement is present, other diagnosis, such as hemangioblastoma, may be suggested, which has enlarged vessels, heterogeneous components within, and no striations. Magnetic resonance spectroscopy (MRS) in LDD shows characteristics of tumor [Figure 2], such as decreased N-acetyl-aspartate (NAA) and increased lactate, but no increase in level of lipids. In contrast, decreased Cho/Cr (choline to creatine ratio) (and myo-inositol/Cr) ratios are in strict contrast to observations in cerebral tumors, where increase of Cho (and myo-inositol) levels are associated with enhanced membrane turn over and demyelination.[1,6]

The increased 18-fludeoxyglucose (18-FDG) uptake with positron emission tomography (PET) and 201 – Tl single-photon emission computed tomography (SPECT) may simply reflect the focally increased cell density and/or a different glucose metabolism of these dysplastic cells. The mass appeared as hypo attenuating on computed tomography (CT) scan. Calcification within the lesion seen in this case [Figure 3] is not a common association.

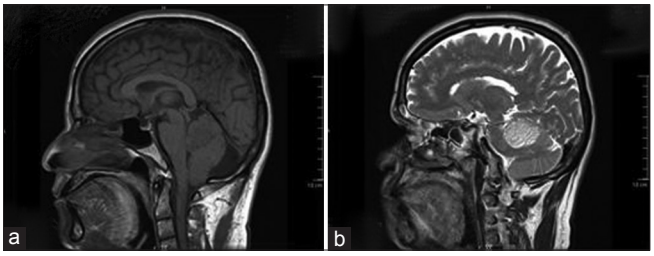

Figure 1: (a) T1WI para sagittal section demonstrates indentation of brain stem by the mass, effacement of fourth ventricle, and minimal tonsillar herniation. (b) T2WI in para sagittal, reveals the classic laminated hypertrophic folia and alternating curvilinear bands of high and low signal intensity

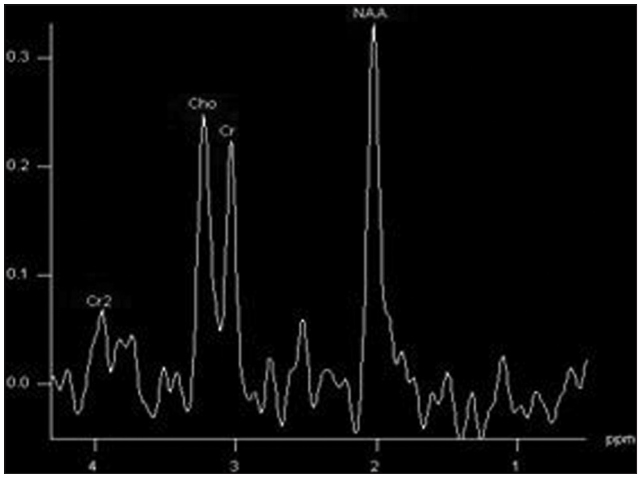

Figure 2: MR Spectroscopy demonstrated elevated creatinine and lactate peak with reduced choline/creatine ratio

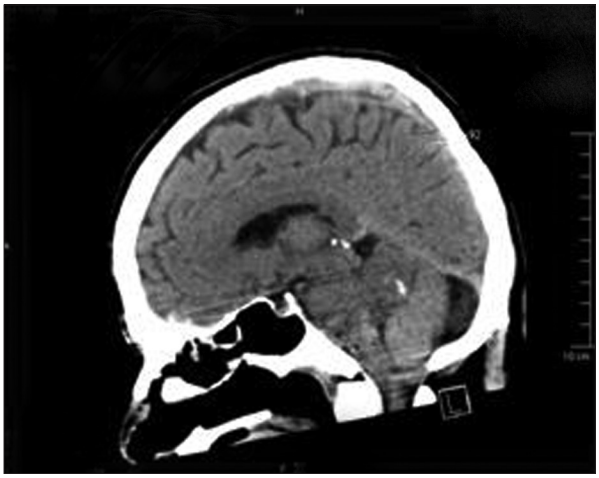

Figure 3: CT scan (sagittal reformat) shows mass in the right cerebellar lobe. Note calcific focus within and pineal and mamillary body calcifications

Diffusion/perfusion weighted imaging, MRS, FDG-PET, and SPECT in LDD can be unpredictable.[5] The standard MRI protocol and MRS was indicative of LDD in this case. It is felt that preoperative diagnosis of LDD can be made with high accuracy using MRI and MRS alone.

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