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·临床医学图像·

小脑发育不良性节细胞瘤/Lhermitte-Duclos病

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Dysplastic cerebellar gangliocytoma/Lhermitte-Duclos disease

HAN Tong

Department of Neuroradiology, Tianjin Huanhu Hospital, Tianjin 300350, China (Email: mrbold@163.com)

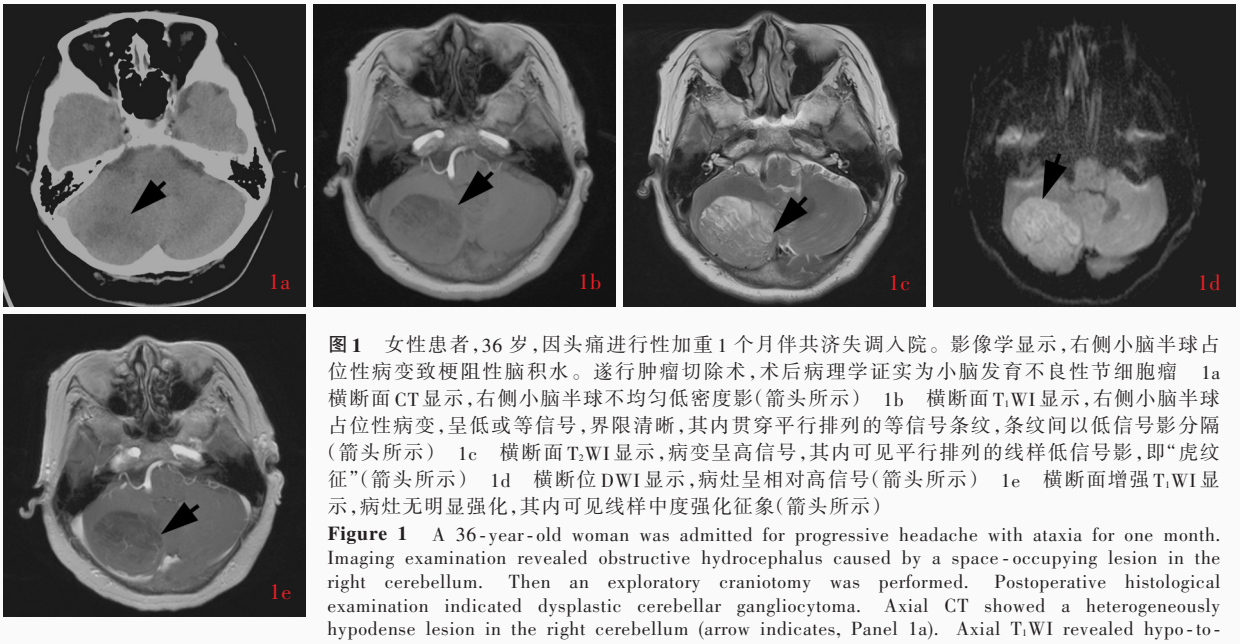

图1 女性患者，36岁，因头痛进行性加重1个月伴共济失调入院。影像学显示，右侧小脑半球占位性病变致梗阻性脑积水。遂行肿瘤切除术，术后病理学证实为小脑发育不良性节细胞瘤 1a 横断面CT显示，右侧小脑半球不均匀低密度影（箭头所示） 1b 横断面T_1WI显示，右侧小脑半球占位性病变，呈低或等信号，界限清晰，其内贯穿平行排列的等信号条纹，条纹间以低信号影分隔（箭头所示） 1c 横断面T_2WI显示，病变呈高信号，其内可见平行排列的线样低信号影，即“虎纹征”（箭头所示） 1d 横断位DWI显示，病灶呈相对高信号（箭头所示） 1e 横断面增强T_1WI显示，病灶无明显强化，其内可见线样中度强化征象（箭头所示）

Figure 1 A 36-year-old woman was admitted for progressive headache with ataxia for one month. Imaging examination revealed obstructive hydrocephalus caused by a space-occupying lesion in the right cerebellum. Then an exploratory craniotomy was performed. Postoperative histological examination indicated dysplastic cerebellar gangliocytoma. Axial CT showed a heterogeneously hypodense lesion in the right cerebellum (arrow indicates, Panel 1a). Axial T_1WI revealed hypo-to-isointense right cerebellar lesion with clear boundary. There were thin iso-intense parallel linear striations across tumor with hypointense streaks between them (arrow indicates, Panel 1b). Axial T_2WI showed hyperintense lesion with areas of parallel linear hypointense strips running throughout lesion which was called "tiger striping" (arrow indicates, Panel 1c). Axial DWI revealed relatively hyperintense signal in right cerebellum (arrow indicates, Panel 1d). Enhanced axial T_1WI showed no obvious enhancement of the lesion. There existed several moderate enhancements of linear signals within the lesion (arrow indicates, Panel 1e).

小脑发育不良性节细胞瘤（WHOⅠ级）是罕见的中枢神经系统良性肿瘤，亦称为Lhermitte-Duclos病（LDD）。好发于成人，3~80岁均可发病，病程进展较慢。成年患者多与常染色体显性遗传性Cowden综合征密切相关。病变位于小脑半球，界限清晰，呈单发或多发，受累的小脑结构扭曲变形，发育不良的小脑脑叶增大，病灶内有时可见囊性变。CT诊断作用有限，表现为颅后窝低或等密度影（图1a），界限清晰。MRI可见病变呈长T_1、长T_2信号，强度不均匀，T_1WI可见平行排列的等信号条纹，以低信号影分隔（图1b），T_2WI呈高信号，其内可见低信号分层样条带（图1c），即“虎纹征”，与异常节细胞增生、脑沟发育不良、中心脑白质萎缩、脱髓鞘导致的异常发育的小脑脑叶肿胀和不规则排列有关，是特征性影像学表现。DWI呈高信号（图1d），推测与病灶白质神经胶质增生和T_2投射效应（T_2 shinning）相关，ADC图呈等信号，有一定特异性。另一特征性影像学表现是增强扫描病灶实性部分无强化（图1e），推测与病变区血-脑屏障无明显破坏且不存在肿瘤新生血管有关。因病灶富含扩张的薄壁引流静脉，故增强扫描可见线样中度强化影，系引流静脉强化征象（图1e）。这些引流静脉在PWI上可以导致病灶呈轻度高灌注，应与肿瘤新生血管致灌注升高相鉴别。单侧或双侧小脑病变无强化征象，伴“虎纹征”，DWI呈稍高或高信号，可以明确诊断小脑发育不良性节细胞瘤。影像学表现不典型者应与发生于小脑的胚胎发育不良性神经上皮肿瘤（DNT）、增强扫描无强化的髓母细胞瘤、小脑胶质瘤等相鉴别。DWI呈高信号可用于发现多发性小病灶，但应与缺血性卒中亚急性期相鉴别。

（天津市环湖医院神经放射科韩彤供稿）