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·临床医学图像·

脱髓鞘性假瘤

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Demyelinating pseudotumor

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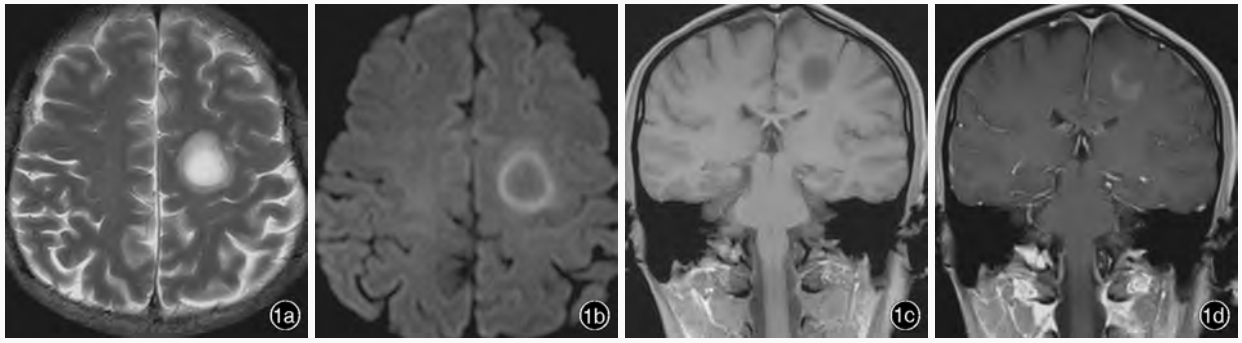

图1 男性患者，49岁。主因右侧肢体无力3 d，于2012年4月9日入院。临床诊断为左侧额叶占位性病变。手术后病理报告为脱髓鞘性假瘤 1a 横断面T_2WI显示，左侧半卵圆中心（中央沟前方）圆形占位，病灶中央区呈高信号、边缘呈环形略高信号，病变周围轻度水肿，左侧侧脑室轻度受压 1b 横断面DWI显示，病灶中心呈低信号、周围为高信号 1c 冠状位平扫T_1WI显示，病灶呈低于脑皮质的低信号，可见“垂直脱髓鞘征” 1d 冠状位增强T_1WI序列显示，病灶呈环形强化，可见“开环征”，开口朝向脑皮质

Figure 1 A 49-year-old male patient suffered from right sided hemiparesis for 3 d and was hospitalized at April 9th, 2012. He was diagnosed as space-occupying lesions in frontal lobe and proved to be demyelinating pseudotumor by postoperative pathological report. Axial T_2WI shows a round mass with central high signal intensity at left centrum semiovale (in front of the central sulci) surrounded by slightly high signal intensity with slight edema. The left ventricle was mildly compressed (Panel 1a). Axial DWI shows central zone of the lesion with low signal intensity surrounded by high signal intensity (Panel 1b). Coronal T_1WI shows the lesion with lower signal intensity than cerebral cortex. "Vertical demyelinating sign" can be seen (Panel 1c). Coronal enhancement T_1WI shows enhanced circular lesion as an "open-ring sign" with opening facing to the cortex (Panel 1d)

脱髓鞘性假瘤（demyelinating pseudotumor）是一种表现为占位效应的中枢神经系统脱髓鞘病变，临床较为少见，影像学检查常以局灶、孤立的肿块存在，故易被误诊为肿瘤。该病在1979年首先由Van der Velden报告，各年龄段均可发病，以青少年多见；无性别差异。大多位于大脑半球白质，偶尔累及小脑白质和脊髓。病理学表现与多发性硬化和急性播散性脑脊髓炎既有相似之处，又存在差异，为二者的独立中间型。临床上多呈急性发病，影像学检查表现为实性肿瘤样占位性病变，糖皮质激素类药物对治疗有效。CT平扫显示，圆形或不规则形进行性占位，周围组织水肿及占位效应较轻。急性或亚急性发病者主要表现为低密度、少数呈等或高密度，伴急性出血时可出现片状高密度区；慢性发病者可表现为低、等或高密度影，水肿程度及占位效应弱于急性者。MRI平扫为长T_1、长T_2改变，合并出血时呈短T_1、长T_2信号；增强扫描病灶多呈弥漫性强化或环形强化，部分呈实质样强化，少数不强化（图1）。强化提示病灶处于活动期，强化程度与巨噬细胞浸润和血-脑脊液屏障破坏的程度相关。“开环征”的出现对诊断脱髓鞘性假瘤具有高度特异性，MRI检查表现为非闭合环形强化，由于强化代表脱髓鞘的前沿，故强化环通常朝向白质，灰质侧不强化。“垂直脱髓鞘征”也有一定的特异性，于矢状位、冠状位影像可观察到病灶长轴有垂直于侧脑室表面的倾向。对于该病的单发病灶需注意与胶质瘤、淋巴瘤及脑脓肿相鉴别；多发病灶应与多发性硬化、急性播散性脑脊髓炎、颅内多发性转移瘤及淋巴瘤相鉴别。若临床拟诊为脱髓鞘性假瘤，可通过糖皮质激素类药物治疗后的MRI结果证实病灶缩小而明确诊断。据文献报道，大多数患者预后良好，约有10%可进展为多发性硬化。

（天津市环湖医院神经放射科韩彤供稿）