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（收稿日期：2016-12-02）

·临床医学图像·

胚胎发育不良性神经上皮肿瘤

doi：10.3969/j.issn.1672-6731.2017.01.015

Dysembryoplastic neuroepithelial tumor

HAN Tong

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

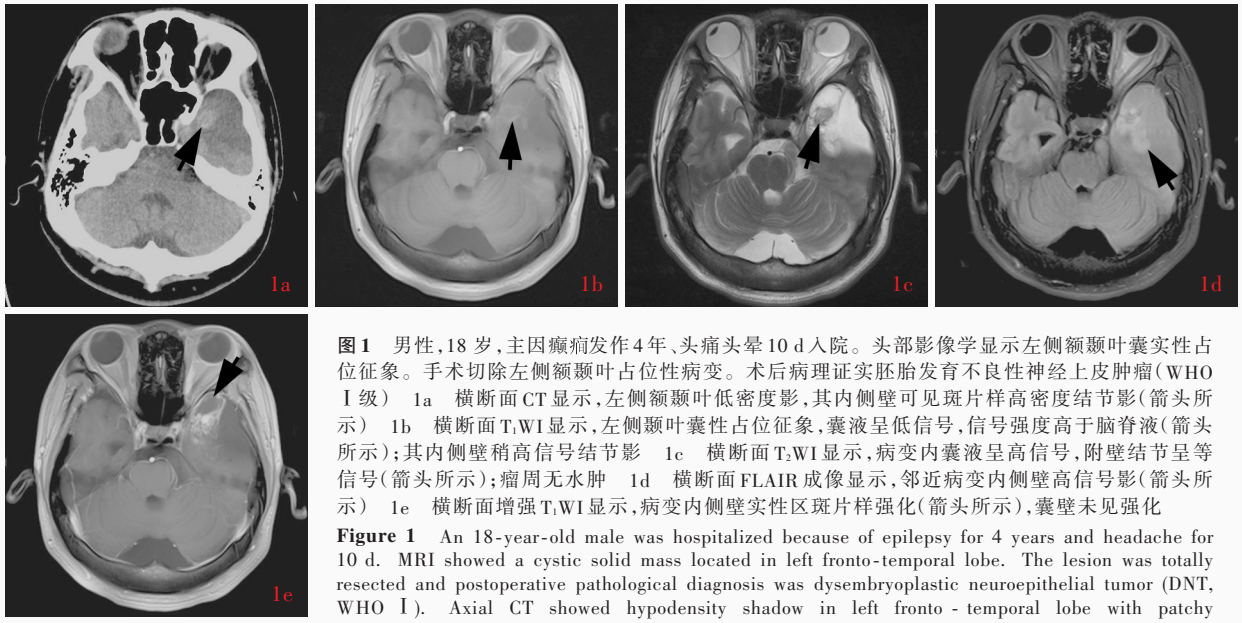

图1 男性，18岁，主因癫痫发作4年、头痛头晕10 d入院。头部影像学显示左侧额颞叶囊实性占位征象。手术切除左侧额颞叶占位性病变。术后病理证实胚胎发育不良性神经上皮肿瘤（WHO Ⅰ级） 1a 横断面CT显示，左侧额颞叶低密度影，其内侧壁可见斑片样高密度结节影（箭头所示） 1b 横断面T_1WI显示，左侧颞叶囊性占位征象，囊液呈低信号，信号强度高于脑脊液（箭头所示）；其内侧壁稍高信号结节影 1c 横断面T_2WI显示，病变内囊液呈高信号，附壁结节呈等信号（箭头所示）；瘤周无水肿 1d 横断面FLAIR成像显示，邻近病变内侧壁高信号影（箭头所示） 1e 横断面增强T_1WI显示，病变内侧壁实性区斑片样强化（箭头所示），囊壁未见强化

Figure 1 An 18-year-old male was hospitalized because of epilepsy for 4 years and headache for 10 d. MRI showed a cystic solid mass located in left fronto-temporal lobe. The lesion was totally resected and postoperative pathological diagnosis was dysembryoplastic neuroepithelial tumor (DNT, WHO Ⅰ). Axial CT showed hypodensity shadow in left fronto - temporal lobe with patchy hyperdensity nodule located in its medial wall (arrow indicates, Panel 1a). Axial T_1WI revealed a cystic mass with hypointensity, which was slightly higher than cerebrospinal fluid, located in left temporal lobe (arrow indicates). A mural nodule with slightly high-intensity was found in the medial wall (Panel 1b). Axial T_2WI showed high-intensity cystic fluid with an isointensity mural nodule (arrow indicates). No edema was found in the surrounding tissue (Panel 1c). Axial fat suppression FLAIR showed high - intensity signal adjacent to medial wall of lesion (arrow indicates, Panel 1d). Axial enhanced T_1WI showed patchy enhancement in solid component of medial wall (arrow indicates) and no enhancement was found in the cystic wall (Panel 1e).

胚胎发育不良性神经上皮肿瘤（DNT，WHO Ⅰ级）是中枢神经系统少见良性肿瘤，属神经元和混合性神经元-胶质肿瘤范畴。1988年由Daumas-Duport等首次命名。多见于儿童和青年，好发于大脑皮质，颞叶最为多见，其次为额叶，基底节、脑室、脑干、小脑、透明隔和胼胝体等亦有报道，肿瘤生长缓慢，临床主要表现为难治性癫痫，预后良好，术前明确诊断十分重要。典型胚胎发育不良性神经上皮肿瘤呈底部位于大脑皮质、尖部朝向脑深部的楔形或脑回样结构，囊性或囊实性，边界清晰，瘤周无水肿，无明显占位征象，邻近大脑皮质可并存皮质发育不良。CT表现为皮质和皮质下界限清晰的低密度影（图1a），20%病灶可见斑片样钙化；位于大脑凸面的肿瘤因生长缓慢致颅骨内板受压变薄。MRI显示病灶内多发结节样和假囊性结构，T_1WI呈低信号，信号强度略高于脑脊液（图1b）；T_2WI可见囊性或多囊性“肥皂泡”样结构，呈高信号（图1c），部分病变内有分隔；FLAIR成像呈略低或高信号，病变边缘可见线样、斑片样或环形更高信号带，即“环形征”（图1d），具有诊断特异性，可能与肿瘤边缘围绕含胶质-神经元成分的疏松组织有关。部分病变可见附壁结节，信号强度略高于大脑皮质。增强扫描可见少部分病变内或边缘线样、斑片样、结节样或环形强化（图1e），系增生的神经胶质细胞伴血管增生所致。应注意与位于皮质和皮质下的囊性肿瘤、带附壁结节的肿瘤（如节细胞胶质瘤、多形性黄色星形细胞瘤、毛细胞型星形细胞瘤、少突胶质细胞瘤）相鉴别。

（天津市环湖医院神经放射科韩彤供稿）