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(收稿日期:2014-09-03)

· 临床医学图像 ·

MELAS 综合征

doi: 10.3969/j.issn.1672-6731.2014.09.018

MELAS syndrome

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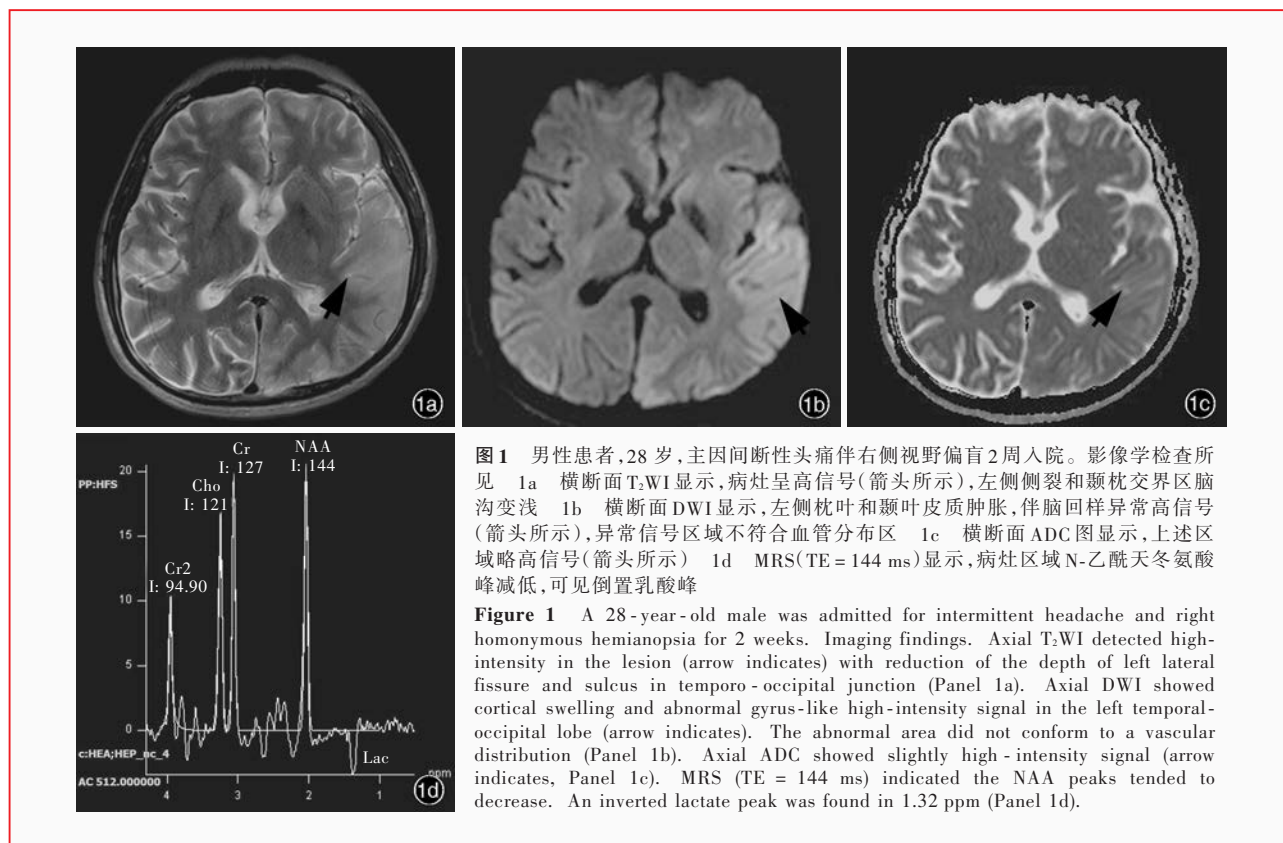


图1 男性患者, 28岁, 主因间断性头痛伴右侧视野偏盲2周入院。影像学检查所见 1a 横断面T₂WI显示, 病灶呈高信号(箭头所示), 左侧侧裂和颞枕交界区脑沟变浅 1b 横断面DWI显示, 左侧枕叶和颞叶皮质肿胀, 伴脑回样异常高信号(箭头所示), 异常信号区域不符合血管分布区 1c 横断面ADC图显示, 上述区域略高信号(箭头所示) 1d MRS(TE = 144 ms)显示, 病灶区域N-乙酰天冬氨酸峰减低, 可见倒置乳酸峰

Figure 1 A 28-year-old male was admitted for intermittent headache and right homonymous hemianopsia for 2 weeks. Imaging findings. Axial T₂WI detected high-intensity in the lesion (arrow indicates) with reduction of the depth of left lateral fissure and sulcus in temporo-occipital junction (Panel 1a). Axial DWI showed cortical swelling and abnormal gyrus-like high-intensity signal in the left temporal-occipital lobe (arrow indicates). The abnormal area did not conform to a vascular distribution (Panel 1b). Axial ADC showed slightly high-intensity signal (arrow indicates, Panel 1c). MRS (TE = 144 ms) indicated the NAA peaks tended to decrease. An inverted lactate peak was found in 1.32 ppm (Panel 1d).

MELAS 综合征是一组以卒中样发作为主要特征的线粒体疾病, 全称为线粒体脑肌病伴高乳酸血症和卒中样发作。任何年龄均可发病, 大多数患者为母系遗传, 少数呈散发。头部CT表现为脑白质, 尤其是皮质下白质多发低密度病灶, 部分患者可出现基底节区对称性或全脑弥漫性钙化。MRI则以不对称累及颞顶枕叶的卒中样病灶为重要特征, 累及皮质或皮质下, 呈脑回样长T₁、长T₂信号改变(图1a), 病变分布不符合血管分布区域; 急性期DWI呈高信号(图1b), 表现扩散系数(ADC)值于正常值范围(图1c); 脑卒中样病灶具有反复发作、不断迁移之特点。MRA或DSA检查无明显血管狭窄等相关征象。灌注成像显示脑血容量(CBV)和脑血流量(CBF)轻度升高。MRS最具诊断优势, 特征性表现为1.32 ppm邻近区域双乳酸峰波峰(图1d)。

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