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·临床医学图像·

Sturge-Weber综合征

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Sturge-Weber syndrome

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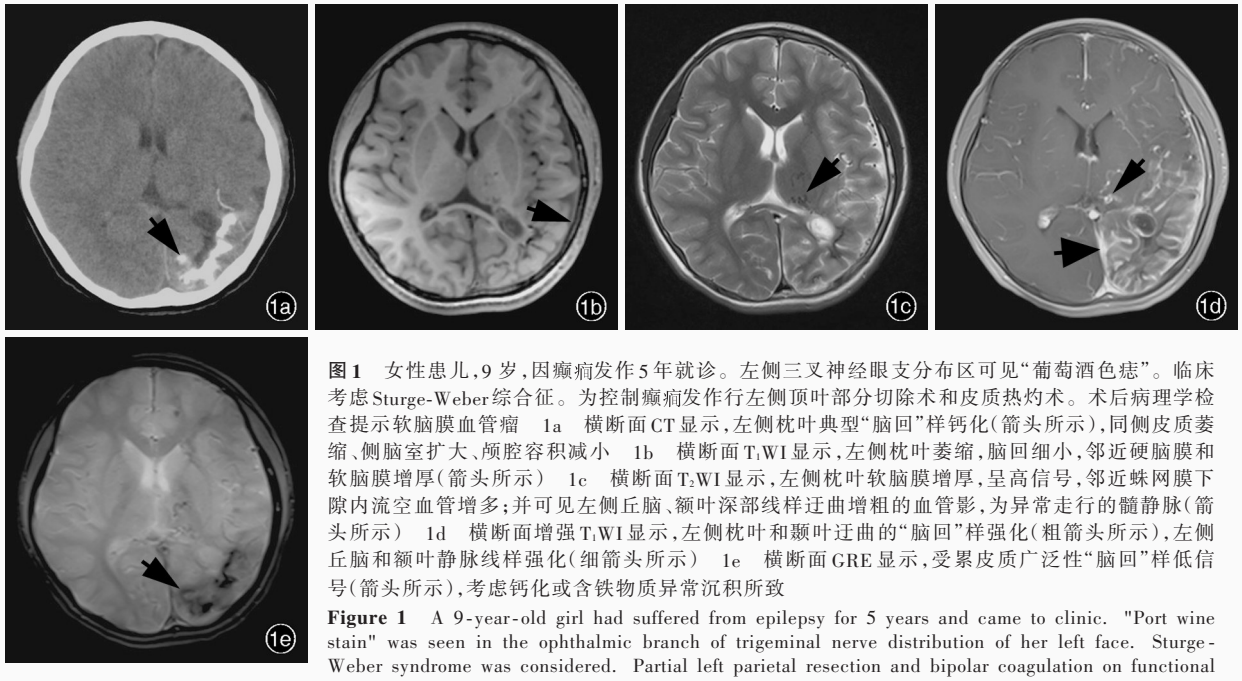

图1 女性患儿，9岁，因癫痫发作5年就诊。左侧三叉神经眼支分布区可见“葡萄酒色痣”。临床考虑Sturge-Weber综合征。为控制癫痫发作行左侧顶叶部分切除术和皮质热灼术。术后病理学检查提示软脑膜血管瘤 1a 横断面CT显示，左侧枕叶典型“脑回”样钙化（箭头所示），同侧皮质萎缩、侧脑室扩大、颅腔容积减小 1b 横断面T_1WI显示，左侧枕叶萎缩，脑回细小，邻近硬脑膜和软脑膜增厚（箭头所示） 1c 横断面T_2WI显示，左侧枕叶软脑膜增厚，呈高信号，邻近蛛网膜下隙内流空血管增多；并可见左侧丘脑、额叶深部线样迂曲增粗的血管影，为异常走行的髓静脉（箭头所示） 1d 横断面增强T_1WI显示，左侧枕叶和颞叶迂曲的“脑回”样强化（粗箭头所示），左侧丘脑和额叶静脉线样强化（细箭头所示） 1e 横断面GRE显示，受累皮质广泛性“脑回”样低信号（箭头所示），考虑钙化或含铁物质异常沉积所致

Figure 1 A 9-year-old girl had suffered from epilepsy for 5 years and came to clinic. "Port wine stain" was seen in the ophthalmic branch of trigeminal nerve distribution of her left face. Sturge-Weber syndrome was considered. Partial left parietal resection and bipolar coagulation on functional cortex were performed for seizure control. Postoperative pathological diagnosis was leptomeningeal angioma. Axial CT showed typical gyriform calcification in left occipital lobe (arrow indicates), and ipsilateral cortical atrophy, ventricular enlargement and reduced cranial cavity (Panel 1a). Axial T_1WI showed left occipital atrophy with small gyri and thickening of adjacent dura and leptomeninges (arrow indicates, Panel 1b). Axial T_2WI showed thickening of left occipital leptomeninges with high intensity and more flow-empty vessels in the adjacent subarachnoid space. Several tortuous deep medullary veins could be seen in left thalamus and frontal lobe (arrow indicates, Panel 1c). Axial enhanced T_1WI showed serpentine gyriform enhancement of left occipital and temporal lobe (thick arrow indicates) and linear enhancement of deep medullary veins (thin arrow indicates, Panel 1d). Axial GRE revealed extensive gyriform low intensity in involved cortex (arrow indicates), which may be caused by calcification or iron deposition (Panel 1e).

Sturge-Weber综合征亦称脑颜面血管瘤综合征，是一种以面部和软脑膜血管瘤病为主要特征的先天性神经皮肤综合征，侵犯面部者称“葡萄酒色痣”，主要累及单侧三叉神经分布区（眼支多见）；软脑膜血管瘤多与面部病变同侧（80%），少数累及双侧（20%）。该病多呈散发，无性别差异，常伴癫痫发作、智力障碍和偏瘫。颅骨X线可见受累区域皮质“双轨”样钙化。脑血管造影术典型表现为病变区域缺乏正常浅皮质静脉，深部髓静脉扩张、数目增加；毛细血管期和静脉期可见弥漫性染色。CT在显示皮质和皮质下钙化方面优于X线和MRI，表现为沿皮质分布的“脑回”样或“车轨”样钙化（图1a）；晚期病变区脑萎缩，致颅骨不对称、患侧颅板增厚（图1a）。MRI扫描显示：（1）异常增厚的软脑膜T_1WI呈等信号（图1b），T_2WI呈高信号，有血管流空时呈低信号（图1c）；增强扫描可见沿脑表面分布的“脑回”样强化（图1d），异常强化组织既包括软脑膜内异常血管，也包括病变区已出现血-脑屏障破坏的变性脑组织。（2）软脑膜血管瘤致皮质浅静脉数目减少，病灶向深静脉系统引流的髓静脉扩张、数目增加，呈圆形和条形低信号（图1c）。（3）病变侧脉络丛增大伴明显强化，与室管膜下静脉扩张继发脉络丛增生有关。（4）受累皮质和皮质下T_2WI和GRE低信号（图1e），可能是由于皮质营养不良性钙化或含铁物质异常沉积所致。应注意与动脉畸形、合并颅内钙化的其他神经皮肤综合征和甲状旁腺功能亢进症等其他伴发颅内病理性钙化的疾病相鉴别。

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