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· 临床医学图像 ·

室管膜下巨细胞星形细胞瘤

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Subependymal giant cell astrocytoma

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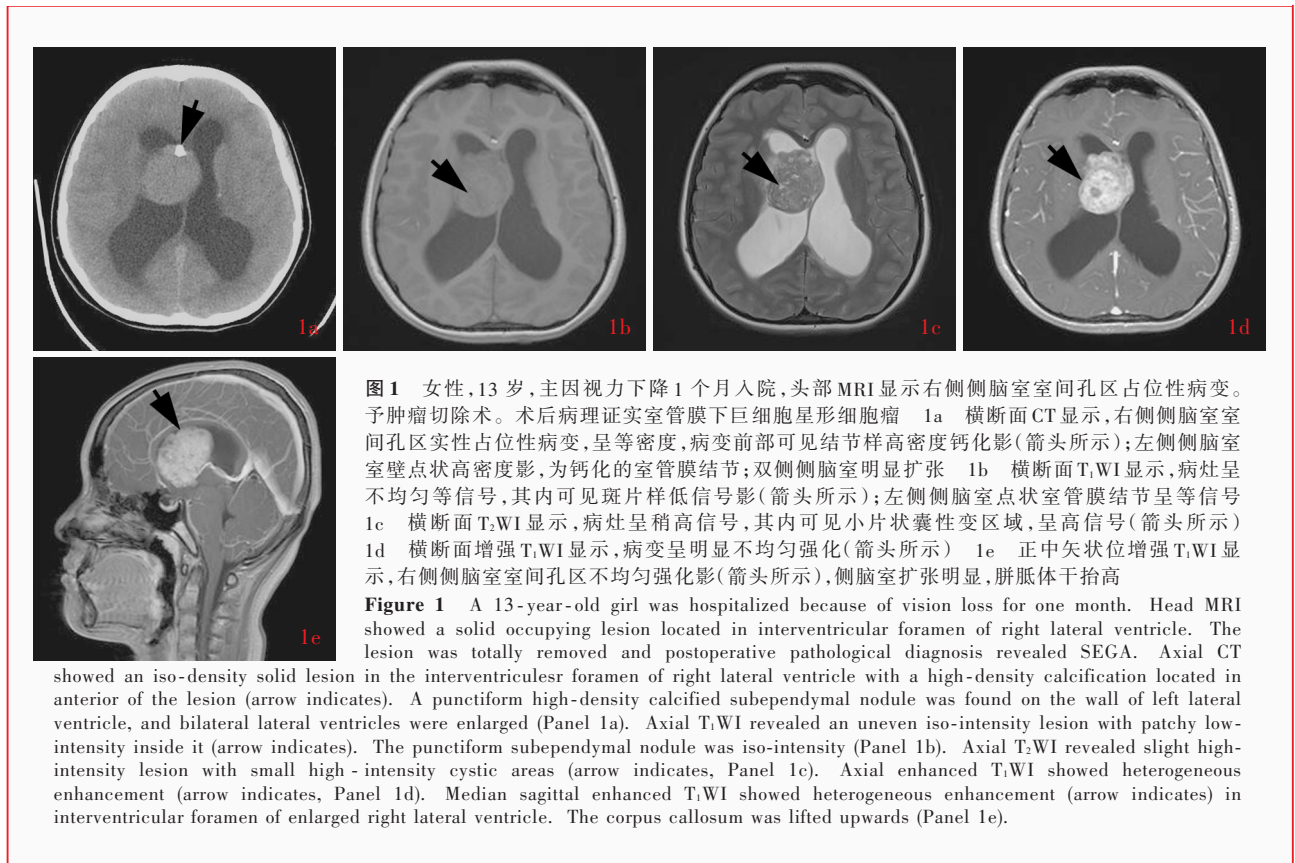


图1 女性,13岁,主因视力下降1个月入院,头部MRI显示右侧侧脑室室间孔区占位性病变。予肿瘤切除术。术后病理证实室管膜下巨细胞星形细胞瘤 1a 横断面CT显示,右侧侧脑室室间孔区实性占位性病变,呈等密度,病变前部可见结节样高密度钙化影(箭头所示);左侧侧脑室室壁点状高密度影,为钙化的室管膜结节;双侧侧脑室明显扩张 1b 横断面T₁WI显示,病灶呈不均匀等信号,其内可见斑片样低信号影(箭头所示);左侧侧脑室点状室管膜结节呈等信号 1c 横断面T₂WI显示,病灶呈稍高信号,其内可见小片状囊性变区域,呈高信号(箭头所示) 1d 横断面增强T₁WI显示,病变呈明显不均匀强化(箭头所示) 1e 正中矢状位增强T₁WI显示,右侧侧脑室室间孔区不均匀强化影(箭头所示),侧脑室扩张明显,胼胝体干抬高

Figure 1 A 13-year-old girl was hospitalized because of vision loss for one month. Head MRI showed a solid occupying lesion located in interventricular foramen of right lateral ventricle. The lesion was totally removed and postoperative pathological diagnosis revealed SEGA. Axial CT showed an iso-density solid lesion in the interventricular foramen of right lateral ventricle with a high-density calcification located in anterior of the lesion (arrow indicates). A punctiform high-density calcified subependymal nodule was found on the wall of left lateral ventricle, and bilateral lateral ventricles were enlarged (Panel 1a). Axial T₁WI revealed an uneven iso-intensity lesion with patchy low-intensity inside it (arrow indicates). The punctiform subependymal nodule was iso-intensity (Panel 1b). Axial T₂WI revealed slight high-intensity lesion with small high-intensity cystic areas (arrow indicates, Panel 1c). Axial enhanced T₁WI showed heterogeneous enhancement (arrow indicates, Panel 1d). Median sagittal enhanced T₁WI showed heterogeneous enhancement (arrow indicates) in interventricular foramen of enlarged right lateral ventricle. The corpus callosum was lifted upwards (Panel 1e).

室管膜下巨细胞星形细胞瘤(SEGA)是临床少见的生长缓慢的神经上皮组织肿瘤,属WHO I级,仅占原发性中枢神经系统肿瘤的0.1%,好发于20岁以下儿童和青少年、偶见于成人,男性多于女性,多发生于结节性硬化症(TSC)患者。肿瘤好发于侧脑室室间孔区,第三和第四脑室及基底节区少见,典型者可堵塞室间孔致梗阻性脑积水。CT呈等密度,强度不均匀,多合并低密度小囊性变区域,病灶边缘和内部常见斑片样或结节样钙化(图1a)。MRI显示肿瘤实性部分呈T₁WI等或略低信号(图1b),T₂WI等或略高信号,多合并小囊性变(图1c),DWI呈等信号,钙化不明显,病灶内出血罕见;增强扫描病灶呈明显不均匀强化(图1d,1e)。结节性硬化症患者除合并室间孔区室管膜下巨细胞星形细胞瘤外,多合并室管膜下结节及皮质和(或)皮质下错构瘤样结节。室管膜下结节在CT上表现为侧脑室室壁室管膜下散在突向侧脑室的结节影,呈等或稍低密度,可见钙化或仅表现为钙化(图1a);在MRI上呈T₁WI等或高信号(图1b)、T₂WI等或低信号、DWI等信号,增强扫描病灶无明显强化。CT和磁敏感加权成像(SWI)能够提示微小室管膜下结节。位于室间孔区以及动态随访中体积增大且明显强化的室管膜下结节均提示转化为室管膜下巨细胞星形细胞瘤的可能。皮质和(或)皮质下结节及类错构瘤样脑白质异常在MRI上呈T₁WI等或略低信号、T₂WI等或略高信号,增强扫描多无明显强化;脑白质异常表现为白质区楔形和线样自皮质指向侧脑室室壁的长T₂信号。典型的室管膜下巨细胞星形细胞瘤较易诊断,而不伴结节性硬化症典型症状者应注意与好发于脑室系统的肿瘤相鉴别,如中枢神经细胞瘤、脉络膜乳头状瘤和室管膜瘤等。

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