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· 临床医学图像 ·

颅内孤立性纤维性肿瘤

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Intracranial solitary fibrous tumor

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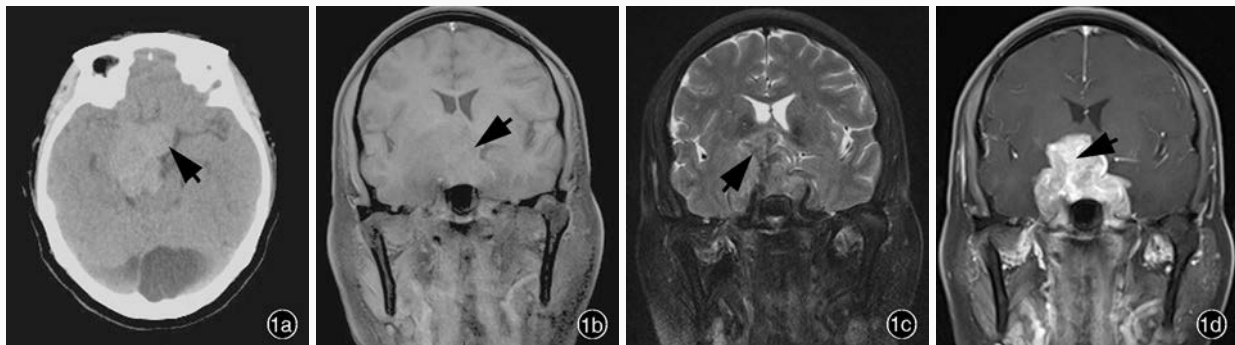


图 1 女性患者, 63 岁, 主因头痛伴视物不清 1 年就诊。头部 MRI 显示鞍区和双侧海绵窦区占位性病变, 诊断为垂体腺瘤。术后病理诊断为孤立性纤维性肿瘤 1a CT 显示, 鞍上池略高密度影, 强度欠均匀, 病灶呈浅分叶状(箭头所示), 与相邻脑实质分界清晰, 压迫右侧颞叶内侧面和大脑脚 1b 冠状位 T₁WI 显示, 鞍区不规则占位性病变, 呈不均匀略低或等信号(箭头所示), 病变位于鞍区和鞍上, 双侧海绵窦受累 1c 冠状位 T₂WI 显示, 肿瘤呈略高信号, 强度欠均匀, 其内可见斑片状低信号(箭头所示) 1d 冠状位延迟增强 T₁WI 显示, 病灶呈明显不均匀强化, T₂WI 低信号区呈现明显渐进性强化(箭头所示)

Figure 1 A 63-year-old female patient had suffered from headache and blurred vision for one year and came to clinic. MRI showed a space-occupying lesion located in sellar region and bilateral cavernous sinus. Preoperative diagnosis was pituitary adenoma. Postoperative pathological diagnosis was solitary fibrous tumor. CT scan showed an irregular slightly high-density lobulated lesion in supra sella cistern (arrow indicates) with compression of medial aspect of the right temporal lobe and cerebral peduncle. There existed a clear margin between normal brain parenchyma and tumor tissue (Panel 1a). Coronal T₁WI indicated an irregular lesion with slightly hypointense and iso-intense signals located in sellar region and supra sella cistern (arrow indicates), involving bilateral cavernous sinus (Panel 1b). Coronal T₂WI showed a heterogeneous hyperintense lesion with patchy hypointense regions within the tumor (arrow indicates, Panel 1c). Delayed enhancement of coronal T₁WI showed obvious heterogeneous enhancement in the lesion, with marked progressive delayed enhancement in the region which showed hypointensity in T₂WI (arrow indicates, Panel 1d).

孤立性纤维性肿瘤(SPT)是一种少见的间叶组织起源的梭形细胞肿瘤,首次报道发生于胸腔,陆续报道见于全身多部位,发生于颅内者罕见。2007年世界卫生组织(WHO)中枢神经系统肿瘤分类将其归为非脑膜上皮细胞起源肿瘤,多位于与硬脑膜毗邻的大脑镰和额颞枕叶凸面、小脑幕、颅后窝和脑桥小脑角区,脑室、鞍区和海绵窦区极为罕见,属WHO I级。该肿瘤具有颅外肿瘤特征,与硬脑膜关系密切,与脑实质界限清晰,表现为宽基底生长、“硬膜尾征”、“假包膜征”。由于肿瘤的不均质性,多呈分叶状改变。CT显示等或略高密度,与肿瘤富含胶原纤维和细胞密集排列有关(图1a),体积较小的病灶密度均匀,体积较大者密度不均,钙化少见,相邻骨质可见吸收压迫,少见反应性骨质增生。MRI能够分辨肿瘤内胶原纤维密集区、肿瘤细胞密集区与粗大供血动脉等软组织成分,在诊断上具有一定优势:T₁WI呈等皮质信号(图1b),T₂WI呈高或高低混杂信号,均匀分布的低信号与稍高信号分界清晰,形成典型的“阴阳征”(图1c),前者为细胞稀疏伴胶原纤维密集区,后者为肿瘤细胞密集区含密集薄壁血管。增强扫描呈中度或明显强化,其强化程度与间质血管和胶原纤维含量相关,动态延迟增强扫描显示胶原纤维密集的T₂WI低信号区呈明显渐进性强化(图1d)。肿瘤体积较小时,信号强度、增强方式和生长形态与脑膜瘤区分困难,肿瘤体积较大时,具有“阴阳征”的分叶状病变伴T₂WI低信号区呈现渐进性强化等典型特征,可作为诊断依据。少数肿瘤内含均匀一致的梭形细胞或胶原纤维,影像学征象不典型,明确诊断须依靠免疫组织化学染色。鉴别诊断包括脑膜瘤、血管外皮细胞瘤,发生于鞍区和脑桥小脑角区者应注意与垂体瘤、脑膜瘤、神经鞘瘤相鉴别。

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