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中分化松果体实质瘤1例

钟儒婷 梁 奕 王焕明

【摘要】中分化松果体实质瘤是一种罕见的中度恶性肿瘤，影像学表现有时难以鉴别。本文报道1例中年男性发现松果体区占位，影像学检查示胚胎性肿瘤，最终行松果体区占位性病变部分切除加取活检术，术后病理诊断为中分化松果体实质瘤。术后进行放疗，随访12个月，无复发。中分化松果体实质瘤影像学表现有一定的特征，但最终确诊需要结合病理学检查；手术切除是首选治疗方法，术后辅以放、化疗，可改善病人预后。

【关键词】松果体肿瘤；中分化松果体实质瘤；影像学表现；显微手术

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One case of pineal parenchymal tumor of intermediate differentiation

ZHONG Ru-ting[1], LIANG Yi[1], WANG Huan-ming[2]. 1. Department of Medical Imaging, General Hospital of The Yangtze River Shipping, Wuhan 430010, China; 2. Department of Neurosurgery, General Hospital of The Yangtze River Shipping, Wuhan 430010, China

【**Abstract**】Pineal parenchymal tumors of intermediate differentiation (PPTID) are a kind of rare moderately malignant tumors, and their imaging manifestations are sometimes difficult to differentiate. This paper reports a case of a middle-aged male with a space-occupying lesion in the pineal region. The imaging examination indicated an embryonic tumor. Eventually, a partial resection of the space-occupying lesion in the pineal region combined with biopsy was carried out, and the postoperative pathological diagnosis was PPTID. Postoperative radiotherapy was administered, and the patient was followed up for 12 months without recurrence. The imaging manifestations of PPTID have certain characteristics, but the final diagnosis requires a combination of pathological examination. Surgical resection is the preferred treatment method, and postoperative radiotherapy and chemotherapy can improve the prognosis of patients with PPTID.

【**Key words**】Pineal tumor; Pineal parenchymal tumors of intermediate differentiation; Imaging manifestations; Microsurgery

1 病例资料

52岁男性，因头晕、头痛、视物重影1年伴加重3个月入院。外院MRI检查发现松果体区占位，行脑室-腹腔分流术，为求进一步诊治转入我院。入院体格检查：神志清晰；双侧瞳孔等大等圆，直径约3 cm，对光反射灵敏；四肢肌力Ⅴ级，肌张力正常，病理征阴性。入院头颅MRI检查显示第三脑室后、松果体区不规则的T_1WI低信号(图1A)、T_2WI高信号(图1B)，FLAIR呈稍高信号(图1C)，病灶大小约48.5 mm×23.4 mm×61.2 mm，边界欠清，部分突入侧脑室内，双侧侧脑室扩大；增强扫描病灶呈明显不均匀强化，内可见流空血管影(图1D)。影像诊断考虑胚胎性肿瘤。完善术前准备后，全麻下行经右枕部开颅松果体区占位性病变部分切除+活检术，术中见松果体区肿块比邻侧脑室枕角脉络丛，质软，灰色鱼肉状，肿瘤血供较丰富，切除部分肿瘤。术后病理检查：镜下可见肿瘤细胞呈小圆形，肿瘤细胞弥漫分布或假菊形团样排列(图1E)。免疫组织化学染色：AE1/AE3(-)，EMA(-)，Vim(-)，GFAP(-)，Olig-2(-)，CD99(-)，Syn(+)，CgA(+)，NF(-)，NSE(+)，CD56(+)，ki-67增生指数为8%，LCA(-)，BCL-2(-)。病理诊断：中分化松果体实质瘤(pineal parenchymal tumor of intermediate differentiation，PPTID)。术后于外院进行放疗，随访12个月，肿瘤无复发。

2 讨 论

2021年，世界卫生组织(World Health Organization，WHO)将松果体实质细胞肿瘤分为松果体细胞瘤(WHO分级Ⅰ级)，PPTID(WHO分级Ⅱ/Ⅲ级)，松果体母细胞瘤(WHO分级Ⅳ级)。PPTID是一种罕见的中度恶性肿瘤，在原发性中枢神经系统肿瘤中占比不到1%[1]。PPTID好发年龄为15~57岁，平均年龄为37岁，无显著性别差异[2]。PPTID病人多表现为颅内压增高症状，还可表现为步态不稳、癫痫等，大多数因中脑导水管、第三脑室后部受压而引起梗阻性脑积水。本文病人因肿瘤压迫第三脑室后部，导致梗阻性脑积水，故产生了头晕、头痛症状。PPTID的影像学特点为松果体区不规则分叶状肿块，大多边界不清。CT平扫表现为等或稍高密度，边缘可见“爆裂状”钙化[3]。MRI表现为T_1WI等/稍低信号，T_2WI等/稍高信号，增强后明显不均匀强化(与肿瘤伴有坏死、囊变有关)，肿块内可见流空血管影，可能与肿瘤间质富含毛细血管，肿瘤细胞包绕周围血管有关，DWI弥散受限[4]。本病例的MRI表现与相关文献报道较一致。

虽然PPTID影像学表现有一定的特征，但该疾病发病率

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作者单位：430010 武汉，长江航运总医院医学影像科(钟儒婷、梁奕)，神经外科(王焕明)

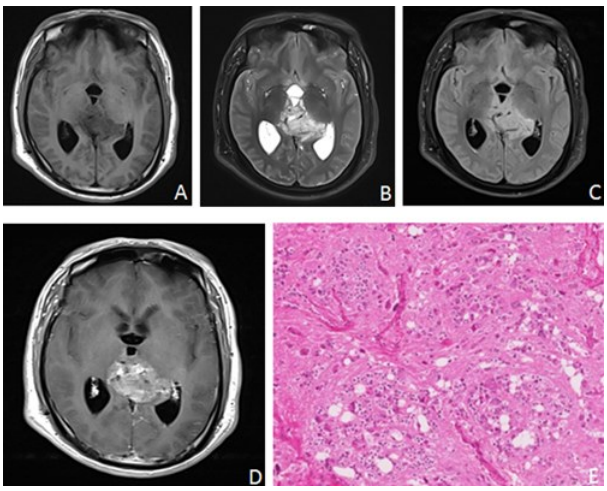

图1 中分化松果体实质瘤术前影像表现及术后病理表现

A-D. 术前头颅MRI显示第三脑室后部、松果体区不规则肿块影，边界欠清，部分突入侧脑室内，双侧脑室扩大，T_1WI呈低信号，T_2WI呈高信号，FLAIR呈稍高信号，增强扫描病灶呈明显不均匀强化，内可见流空血管影；E. 术后病理检查(HE，×100)，显微镜下可见肿瘤细胞呈小圆形，弥漫分布或假菊形团样排列

Figure 1 Preoperative imaging features and postoperative pathological features of a patient with pineal parenchymal tumor of intermediate differentiation

A-D: Preoperative head MRI reveals an irregular mass shadow in the posterior part of the third ventricle and the pineal region, with indistinct boundaries, partially protruding into the lateral ventricles, and bilateral ventricular dilation; the mass shows a low signal on T_1WI, a high signal on T_2WI, a slightly high signal on FLAIR, and significantly heterogeneous enhancement on enhanced scan, with visible flow-void vascular shadows. E: Postoperative pathological examination (HE, ×100), the tumor cells are small and round, diffusely distributed or arranged in pseudorosette-like patterns under the microscope.

较低，术前明确诊断仍然比较困难，最终确诊需要结合病理学检查。其镜下特点表现为瘤细胞呈弥漫或分叶状分布，可见假菊形团，其免疫组化标记Syn、NF、CgA可呈阳性，增殖指数在3%~10%[5]。手术切除是目前PPTID的首选治疗方法，可减轻肿块占位效应并获取肿瘤标本，术后再辅以放、化疗，可改善病人预后。

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(上接第697页)

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