·临床病理报告·

小脑富脂质髓母细胞瘤临床病理学特征:病例报告及文献综述

刘丽燕 王行富 张声 陈余鹏 李赞

【摘要】 **目的** 探讨小脑富脂质髓母细胞瘤临床病理学特征。**方法** 对一例小脑富脂质髓母细胞瘤患者临床表现、影像学特点,以及组织病理学和免疫组织化学特征进行回顾性分析,并复习相关文献。**结果** 男性患者,26岁。临床表现为反复头痛,伴头晕、恶心、呕吐。MRI显示病灶填充第四脑室,T_1WI呈均匀低信号、T_2WI呈不均匀高信号,增强T_1WI扫描病灶呈明显均匀强化,边界清晰。手术后光学显微镜下观察肿瘤细胞弥漫增生,大小一致,胞质淡染,呈弱酸性或嗜双色,核圆形或卵圆形,染色质细腻,核仁可见,核分裂象易见;间质中含有丰富的薄壁血管;富于脂质的细胞呈灶性分布。免疫组织化学染色肿瘤细胞CD56和突触素弥漫阳性,局灶表达神经微丝,弱表达少突胶质细胞系转录因子-2;不表达巢蛋白、神经元核抗原、S-100蛋白、胶质纤维酸性蛋白和上皮膜抗原;TP53染色阳性(约10%),Ki-67抗原标记指数约为40%。**结论** 发生于小脑的富脂质髓母细胞瘤为临床罕见的中枢神经系统肿瘤,影像学呈现发生于小脑蚓部向第四脑室生长的占位性病变;病理学表现肿瘤细胞由高度密集的一致圆形细胞组成,具有灶性脂肪样细胞分化,应注意与小脑脂肪神经细胞瘤和室管膜瘤等相鉴别。

【关键词】 髓母细胞瘤; 小脑肿瘤; 磁共振成像; 病理学; 免疫组织化学

DOI:10.3969/j.issn.1672-6731.2012.03.014

Clinicopathological features of cerebellar lipidized medulloblastoma: a case report and review of literatures

LIU Li-yan, WANG Xing-fu, ZHANG Sheng, CHEN Yu-peng, LI Zan

Department of Pathology, the First Affiliated Hospital of Fujian Medical University, Fuzhou 350005, Fujian, China

Corresponding author: ZHANG Sheng (Email: zhgshg@126.com)

【**Abstract**】 **Objective** To explore the clinicopathological features of cerebellar lipidized medulloblastoma. **Methods** The clinical manifestations, neuroimaging, histopathological and immunohistochemical features were analysed in one case of lipidized medulloblastoma in the cerebellar vermis. Related literatures were reviewed. **Results** A 26-year-old man presented with intermittent headache, accompanied by dizziness, nausea and vomiting. The magnetic resonance imaging (MRI) demonstrated a mass located the cerebellar vermis convex to the fourth ventricle. The tumor with well-demarcated boundary was homogeneous hypointense on T_1 weighted and heterogeneous hyperintense on T_2 weighted images, and enhanced brilliantly and homogenously on contrast. The patient subsequently underwent gross total mass resection. Microscopically, there was diffuse infiltration by high cellularity of tumor cells. The cytoplasm were thin eosinophilic to amphophilic. The neoplastic cells showed round to oval hyperchromatic nuclei with a delicately stippled chromatin and occasional conspicuous nucleoli and numerous mitotic figures were also present. Thin-wall vascular proliferation was detected. Lipid-laden cells were focally distributed in tumor tissue. On immunohistochemical examination, the neoplasm was reactive for CD56 and synaptophysin (Syn), focally positive for neurofilament protein (NF), weakly positive for oligodendrocyte lineage transcription factor 2 (Olig-2), and negtive for nestin, neuronal nuclei (NeuN), S-100 protein (S-100), glial fibrillary acidic protein (GFAP) and epithelial membrane antigen (EMA). TP53 protein was over expressed in 10% of tumor cells. Ki-67 antigen labeling index were about 40%.

作者单位:350005 福州,福建医科大学附属第一医院病理科[刘丽燕(现在福州,福建省级机关医院病理科,邮政编码:350003),王行富,张声,陈余鹏,李赞]

通讯作者:张声(Email:zhgshg@126.com)

Conclusion Cerebellar lipidized medulloblastoma is rare. Neuroimaging showed space occupying lesion in cerebellar vermis. Histologically, the tumor cells were consisted of monotonous, round cells with focal accumulations of lipidized cells. The differential diagnosis include liponeurocytoma and ependymoma and so on.

【Key words】 Medulloblastoma; Cerebellar neoplasms; Magnetic resonance imaging; Pathology; Immunohistochemistry

髓母细胞瘤(medulloblastoma)是发生于小脑的侵袭性胚胎性肿瘤,好发于儿童,伴明显神经元分化,易通过脑脊液途径播散,主要生长于小脑蚓部并向第四脑室生长。近年来,陆续有文献报道富脂质髓母细胞瘤(lipidized medulloblastoma)大多伴有灶性脂肪样细胞分化,但临床鲜见。笔者报告一例发生于小脑蚓部富于脂质的髓母细胞瘤,并结合相关文献对其临床表现、病理学诊断与鉴别诊断特点进行探讨。

病历摘要

患者 男性,26岁。主因反复头痛、恶心、呕吐2个月,于2011年2月18日入院。患者入院前2个月无明显诱因反复头痛,以双侧额部为甚,程度较轻,偶有疼痛程度加重至中等程度,可持续1~10 d,发作时伴有头晕,恶心、呕吐,但无抽搐,以及步伐、步态不稳等症状或体征。7~8 d前因头痛症状加重就诊于当地医院,头部MRI检查提示"第四脑室占位性病变"。为了进一步明确诊断与治疗而入住我院。患者主诉自发病以来精神状态良好,饮食、睡眠无明显异常,大小便正常,体质量无明显减轻。既往体格健康,无肿瘤家族遗传史。

诊断与治疗经过 入院后体格检查:体温36.3 ℃,脉搏76次/min,呼吸17次/min,血压120/70 mm Hg(1 mm Hg=0.133 kPa)。神志清楚。双侧眼睑无下垂;双侧瞳孔等大、等圆,直径约为3 mm,对光反射灵敏,眼球各方向运动无障碍,无眼震。双侧鼻唇沟对称,伸舌居中。四肢肌张力正常,肌力5级,腱反射对称活跃,病理征未引出;深浅感觉基本正常,共济运动无异常,双侧Kernig征阴性。头部MRI扫描提示第四脑室扩大,病灶由小脑蚓部向第四脑室生长,T_1WI呈均匀低信号、T_2WI为不均匀高信号;增强T_1WI扫描病灶呈明显均匀强化,边界清楚,大小约为2.50 cm×2.50 cm×2.00 cm(图1),病灶周围脑实质无水肿,占位效应轻微,脑桥背侧呈弧形压迹,中央导水管以及幕上脑室轻度扩张(图2)。临床初步诊断:第四脑室占位性病变,可疑室管膜瘤。

完善各项检查后于2011年2月22日经枕下正中入路施行第四脑室肿瘤切除术,术中发现肿瘤主要依附于第四脑室左侧,界限清楚,由附着面供血,血供中等,质地柔软,大小约为2.50 cm×2.50 cm×2.00 cm,呈暗红色,于手术显微镜下全切除肿瘤,并行组织病理学检查。(1)大体标本观察:肿瘤组织标本比较完整,大小约为2.50 cm×2.00 cm×2.00 cm,表面灰暗(图3a),剖面呈灰红色,质地均匀、柔软(图3b)。经体积分数为3.7%的中性甲醛溶液固定、常规脱水、石蜡包埋,3μm组织切片,分别行HE染色和免疫组织化学染色。(2)组织病理学观察:由小而一致的肿瘤细胞和脂肪样细胞组成(图4a)。肿瘤细胞密度较高,胞质淡染,弱嗜酸性,核呈圆形或卵圆形,染色质细腻,核仁可见,伴不同程度核周空晕(图4b);脂肪样细胞为圆形或多边形,呈空泡状,胞核被脂肪滴挤向一侧,呈"新月"形;单个或成片的脂肪样细胞排列于肿瘤细胞之间,分布不均匀(图4c);核分裂象易见,约为9个/10 HPF(图4d);肿瘤表面局部区域可见纤维素性坏死,间质中含丰富的由小至中等大小的弓状或分支状的薄壁血管。(3)免疫组织化学染色:采用EnVision二步法检测肿瘤细胞胶质纤维酸性蛋白(GFAP,1∶100)、S-100蛋白(S-100,1∶200)、少突胶质细胞系转录因子-2(Olig-2,1∶100)、突触素(Syn,1∶150)、神经元核抗原(NeuN,1∶100)、巢蛋白(nestin,工作液)、神经微丝(NF,1∶100)、CD56(1∶100)、上皮膜抗原(EMA,1∶100)、TP53(1∶100),以及Ki-67抗原(1∶200)表达水平,所有抗体均购自美国Dako公司。检测结果显示,细胞膜CD56表达呈弥漫强阳性(图5a),胞质突触素表达阳性(图5b),神经微丝表达呈局灶性阳性,少突胶质细胞系转录因子-2表达呈弱阳性;肿瘤细胞不表达巢蛋白、神经元核抗原、S-100蛋白、胶质纤维酸性蛋白和上皮膜抗原等;细胞核TP53阳性表达率为10%(图5c),Ki-67抗原标记指数40%

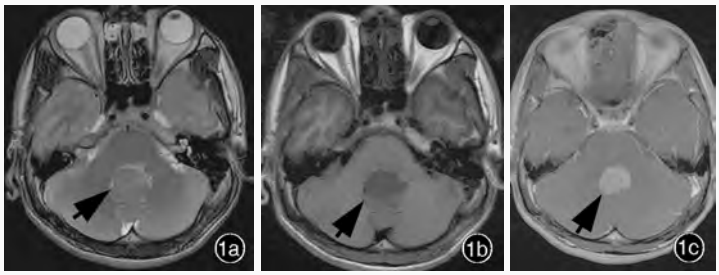

图1　手术前横断面MRI扫描显示病灶由小脑蚓部向第四脑室生长（箭头所示）　1a　T_2WI扫描病灶呈不均匀高信号　1b　T_1WI扫描病灶呈均匀低信号　1c　增强T_1WI扫描病灶呈明显均匀强化，边界清楚

Figure 1　Preoperative axial magnetic resonance imaging (MRI) revealed a mass located the cerebellar vermis convex to the fourth ventricle (arrows indicate). This lesion was heterogeneous hyperintense on T_2 weighted (Panel 1a) and homogeneous hypointense on T_1 weighted images (Panel 1b), and enhanced brilliantly and homogenously on contrast (Panel 1c), and with well-demarcated boundary

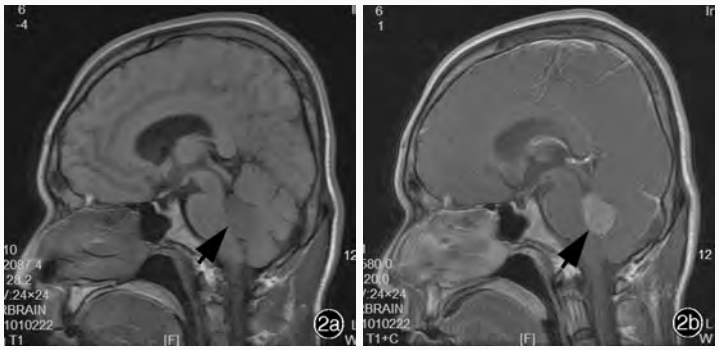

图2　手术前矢状位MRI检查显示，病灶周围脑实质无水肿，脑桥背侧呈弧形压迹，中央导水管及幕上脑室轻度扩张（箭头所示）　2a　T_1WI序列病灶呈均匀低信号　2b　T_1WI增强扫描病灶呈明显均匀强化

Figure 2　Preoperative sagittal MRI demonstrated that brain parenchyma around the lesion was without edema and the aquaeductus Sylvii and supratentorial ventricles were mild dilatation (arrows indicate). The mass was homogeneous hypointense on T_1 weighted (Panel 2a) and enhanced brilliantly and homogenously on contrast (Panel 2b)

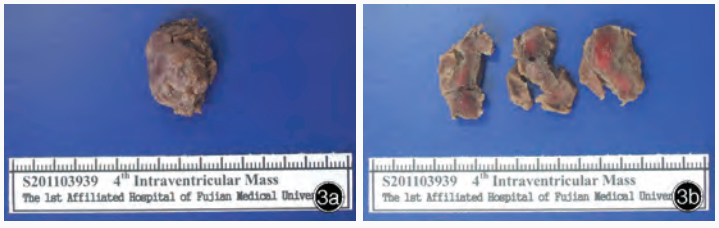

图3　手术后大体标本观察　3a　手术全切除肿瘤，无包膜、呈结节状　3b　剖面呈灰红色，质地均匀

Figure 3　General sample was observed after operation. The total resected tumor was nodosity without capsule (Panel 3a). The section was pinkish gray and uneven in texture (Panel 3b)

（图5d）。病理学诊断：（小脑蚓部）富脂质髓母细胞瘤（WHO Ⅳ级），细胞增殖活跃，核分裂象多见（9个/10 HPF）。患者术后未接受放射治疗，术后6个月随访时，MRI检查显示第四脑室及脑桥前池有明显强化的多结节状团块影，大小约为4.10 cm×2.10 cm×2.20 cm，以第四脑室左前方尤为明显（图6），提示肿瘤复发。

讨　论

髓母细胞瘤是呈侵袭性生长的胚胎性肿瘤，好发于小脑蚓部并突入第四脑室生长，多伴有不同程度的神经元和胶质细胞分化，主要通过脑脊液途径播散[1-2]。70%发生于16岁以下青少年，成人髓母细胞瘤80%发生于21～40岁，以男性好发，约占65%[1]。在2007年第4版WHO中枢神经系统肿瘤分类标准中，将髓母细胞瘤分为促纤维增生/结节型髓母细胞瘤、伴广泛结节性髓母细胞瘤、间变型髓母细胞瘤、大细胞髓母细胞瘤和伴有肌原性分化[3]及黑色素分化的髓母细胞瘤[1]。富脂质髓母细胞瘤为髓母细胞瘤中的罕见亚型，伴有局灶性脂肪样细胞分化[4]。Bechtel等[5]在1978年首次报告一例44岁男性脂肪瘤型髓母细胞瘤患者。

一、临床及影像学特征

富脂质髓母细胞瘤多发生于小脑蚓部及小脑半球，可充满第四脑室。主要表现为颅后窝占位和颅内压升高的症状与体征，如头痛、眩晕、视力障碍、渐进性步态不稳、恶心、呕吐等，但神经系统检查基本正常。影像学检查呈现占位性病变，这对诊断具有重要意义。CT平扫显示为高密度影像，密度较均匀，少数呈混杂密度和等密度，边界清晰；增强扫描病灶呈均匀强化，仅少数为不均匀强化，坏死、囊变无强化，部分病例瘤周可见水肿及梗阻性脑积水征象[6]。MRI检查，T_1WI序列肿瘤多呈略低信号，少数呈混杂及等信号，增强后呈均匀强化；T_2WI表现为略高或混杂信号，肿瘤内侧小脑深部可见低信号坏死、囊变和瘤周水肿等影像[7]。髓母细胞瘤可沿脑脊液通路在蛛网膜下隙种植转移，以粟粒状或结节状散在分布于神经轴内，其影像学表现与原发灶相似[6-7]。到目前为止，尚无富于脂质的髓母细胞瘤发生转移的报道。

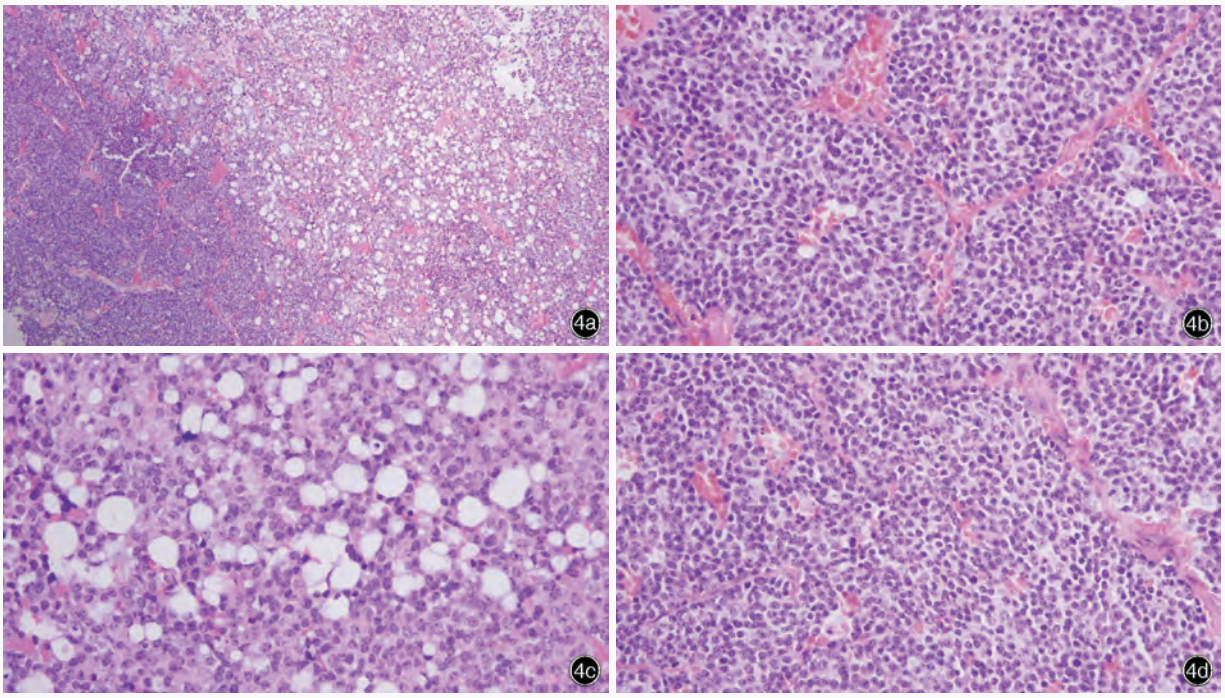

图4　光学显微镜观察　HE染色　4a　肿瘤组织主要由小而一致的肿瘤细胞和大的脂肪样细胞组成　×100　4b　肿瘤细胞密度较高，核呈圆形或卵圆形，染色质细腻　×400　4c　脂肪样细胞呈空泡状，细胞核被脂肪滴挤向一侧；单个或成片的脂肪样细胞排列于肿瘤细胞之间，分布不均匀　×400　4d　肿瘤细胞胞核分裂象易见，间质富含小至中等大小的弓状或分支状薄壁血管　×400

Figure 4　Optical microscopy findings　HE staining. The tumor showed two different cellular components: small, uniformly packed cells and large adipocyte-like cells　× 100 (Panel 4a). There was diffuse infiltration by high cellularity of tumor cells. The cells showed round to oval hyperchromatic nuclei with a delicately stippled chromatin　×400 (Panel 4b). The lipidized cells were present individually as well as in small clusters within the tumor cells and showed a single fat vacuole　×400 (Panel 4c). Numerous mitotic figures were present and thin-wall vascular proliferation was detected　×400 (Panel 4d)

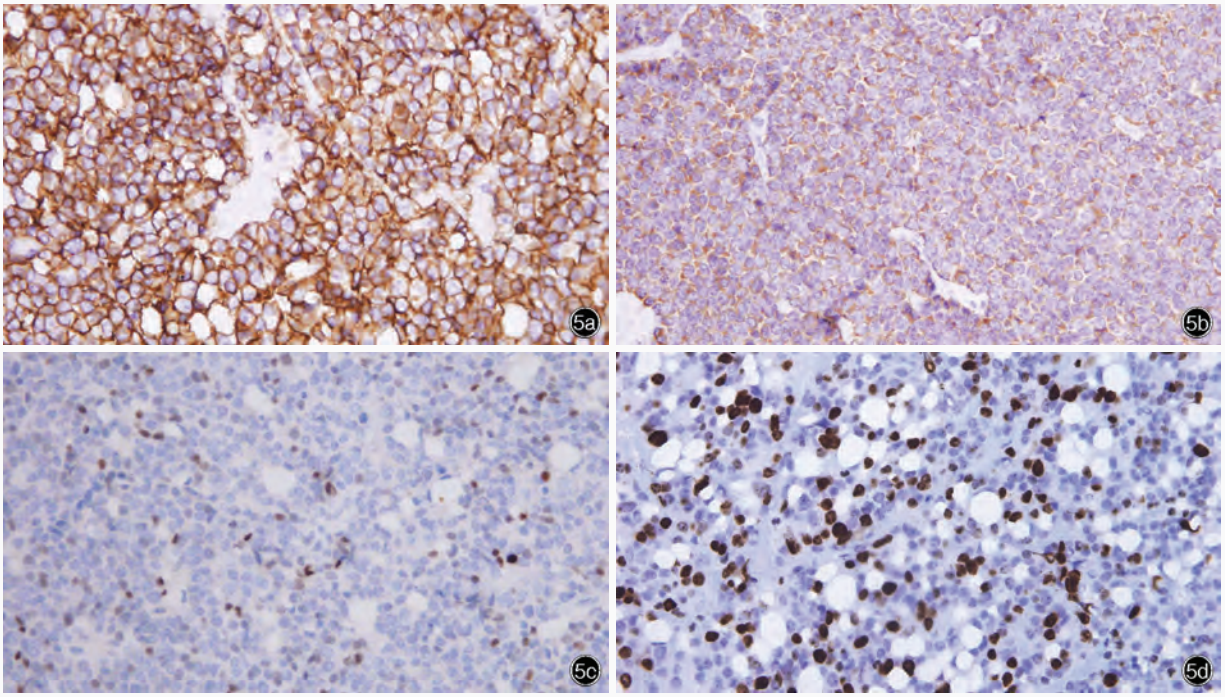

图5　光学显微镜观察　免疫组织化学染色（EnVision二步法）　×400　5a　肿瘤细胞胞膜CD56表达呈强阳性　5b　肿瘤细胞胞质突触素表达阳性　5c　肿瘤细胞胞核TP53阳性表达率约为10%　5d　肿瘤细胞Ki-67抗原标记指数约为40%

Figure 5　Immunohistochemistry (EnVision)　×400. The neoplasm was reactive for CD56 (Panel 5a) and synaptophysin (Syn, Panel 5b). TP53 protein was over expressed in 10% of tumor cells (Panel 5c). Ki-67 antigen labeling index were about 40% (Panel 5d)

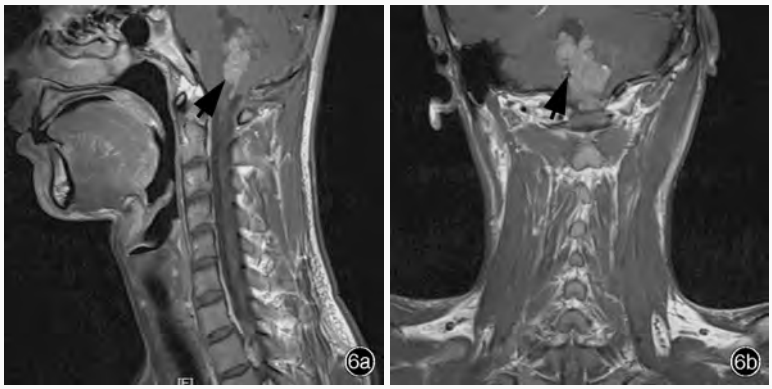

图6 随访6个月时，MRI检查可见第四脑室及脑桥前池明显强化的多结节状肿块，以第四脑室左前方明显（箭头所示）6a 矢状位增强T_1WI序列 6b 冠状位增强T_1WI序列

Figure 6 At the six month follow - up visit. MRI revealed that the lesions were enhanced brilliantly and homogenously on contrast in the fourth ventricle and cerebellopontine cistern, especially in the front - left of the fourth ventricle (arrows indicate). Contrast enhanced T_1 - weighted sagittal (Panel 6a) and coronal (Panel 6b)

二、组织病理学诊断与鉴别诊断

富脂质髓母细胞瘤大体标本表现为粉色或灰色团块，剖面呈灰红、灰白色，质地均匀。光学显微镜下观察肿瘤主要由高密度肿瘤细胞和脂肪样细胞组成，肿瘤细胞大小较为一致，胞质淡染、弱嗜酸性；核圆形或卵圆形，染色质细腻，可见核仁，伴不同程度核周空晕；脂肪样细胞为圆形或多角形、空泡状，扁形核位于细胞的一侧，呈单个或灶性分布的脂肪样细胞沿肿瘤细胞之间不均匀分布；间质可含有丰富的分支血管，将肿瘤分割成小叶状结构。免疫组织化学染色显示，肿瘤细胞和脂肪样肿瘤细胞均同时表达神经元标志物突触素[4,8]、胶质纤维酸性蛋白[4,8]、神经元特异性烯醇化酶[4,8-9]和S-100蛋白[4,8-9]等，偶可见微管相关蛋白-2（MAP-2）和神经微丝表达阳性[10]，但不表达上皮膜抗原[4,9]和细胞角蛋白（CK）[8]；脂肪样细胞不表达CD68[9]；文献报道的Ki-67抗原标记指数为15%～40%[9-11]。由于富脂质髓母细胞瘤患者的临床表现缺乏特异性，因此需结合影像学和组织病理学检查等多种手段综合分析以明确诊断，而且需注意与以下疾病相鉴别。（1）小脑脂肪神经细胞瘤：好发于成人的小脑肿瘤，亦伴有神经元/神经细胞和脂肪分化，脂肪样细胞分布也可与富脂质髓母细胞瘤相似呈簇状，故极易与富脂质髓母细胞瘤相混淆。主要鉴别特点为前者Ki-67/MIB-1抗原标记指数大多低于6%[12-14]，该例患者肿瘤细胞密度较高，且核分裂象多见，Ki-67抗原标记指数高达40%，支持富脂质髓母细胞瘤的诊断。（2）室管膜瘤：起源于脑室和脊髓中央导水管管壁的室管膜细胞，可发生于任何年龄，增强MRI扫描边界清楚，呈中等至明显强化影像，病灶内多伴囊性变且增强时囊壁强化，有时可伴出血、坏死和钙化[15-16]；而富脂质髓母细胞瘤的坏死和囊性变多不强化。组织病理学显示肿瘤细胞呈中等密度，形态一致，可见具有特征性的血管周假菊形团和室管膜菊形团，免疫组织化学染色上皮膜抗原呈胞质内斑点状阳性表达；而富脂质髓母细胞瘤不表达上皮膜抗原[4,9]。（3）血管母细胞瘤：可发生于中枢神经系统的任何部位，散发性毛细血管性血管母细胞瘤好发于小脑，影像学尤其是MRI扫描常可见出血和囊性变，以及增强时囊壁呈结节状强化，表现为大囊内含有附壁小结节，即特征性“大囊小结节”[17-18]，此为主要鉴别特点。光学显微镜下观察主要由胞质透明、泡沫状、毛玻璃样的间质细胞和丰富的毛细血管网所组成[19]。（4）多形性黄色瘤型星形细胞瘤：大多位于大脑皮质邻近大脑表面，偶可发生于小脑和脑干[20]。光学显微镜下观察肿瘤由纤维性、巨大、多核的肿瘤性星形细胞所组成，部分肿瘤细胞内含有脂肪滴，似“黄色瘤”[20]。诊断特征主要为黄色瘤样细胞表达胶质纤维酸性蛋白，致密的网状纤维和淋巴细胞浸润[20]。

三、遗传学及发生发展机制

关于髓母细胞瘤的组织起源目前尚未完全确定，主要存在两种假说：大多数学者支持起源于小脑的外颗粒层细胞[1]；部分认为起源于室管膜下的基质细胞[1]。Sharma等[9]对富脂质髓母细胞瘤进行超微结构观察发现，肿瘤细胞核圆形或卵圆形，不规则的颗粒状染色质分布于核周围，偶见核仁；含有脂质的空泡细胞或为小的单泡状或为大的多泡状；细胞界限不明显；偶见突触囊泡。免疫组织化学染色肿瘤细胞（包括含有脂肪滴空泡的细胞）神经标志物表达阳性，结合免疫组织化学染色及电子显微镜观察所见，推测脂肪样细胞的形成可能是肿

瘤细胞的不同分化或细胞代谢干扰的结果。关于髓母细胞瘤的细胞遗传学和分子遗传学研究，目前仅发现少部分髓母细胞瘤（5%～10%）存在*TP53*基因突变[1,21]，而Horstmann等[22]将小脑脂肪神经细胞瘤的基因改变与其进行对比研究，发现20%的患者存在*TP53*基因突变，明显高于前述研究所报道的5%～10%。推测髓母细胞瘤可能伴有*PTCH*、*APC*和*β-catenin*基因的失活突变，突变率分别为9%、4%和6%，而小脑脂肪神经细胞瘤几乎不存在这些基因的突变[22]；而且作为髓母细胞瘤最具特征的17q等臂染色体异常，在小脑脂肪神经细胞瘤是不存在的[14]，这些特征亦可作为富脂质髓母细胞瘤与小脑脂肪神经细胞瘤的鉴别点。

四、治疗与预后

以往的观点认为，“富脂质髓母细胞瘤”与“小脑脂肪神经细胞瘤”这两个名称是相同的[23]。近年的研究发现二者是有区别的，小脑脂肪神经细胞瘤（WHOⅡ级）具有良性的生物学行为，大部分患者生存期超过5年，单纯手术切除的患者生存时间最长可达18年。随着报道病例的增多和随访时间的延长，发现该肿瘤极易复发，复发率为31%[24]，甚至可高达50%[14]。而富脂质髓母细胞瘤相当于WHOⅣ级，易复发，手术切除后需辅助放射治疗或药物化疗。Sharma等[9]回顾16例发生于成年人的富脂质髓母细胞瘤患者，预后均较好，而发生于儿童者由于增殖潜能较高，则预后不良。因此，二者需进行鉴别。

五、结论

富脂质髓母细胞瘤为髓母细胞瘤中的罕见亚型，随着报道病例的增多，希望引起更多学者的关注，以及对其相关病例进行更加深入的研究与探讨。目前有学者认为阐明脑肿瘤干细胞发展的分子机制将成为脑肿瘤治疗的新方向，尤其是髓母细胞和恶性胶质瘤[25]，这也为我们今后的研究提供了新的思路。

参 考 文 献

[1] Giangaspero F, Eberhart CG, Haapasalo H, et al. Medulloblastoma// Louis DN, Ohgaki H, Wiestler OD, et al. WHO classification of tumours of the nervous system. Lyon: IARC, 2007: 132-140.

[2] Terterov S, Krieger MD, Bowen I, et al. Evaluation of intracranial cerebrospinal fluid cytology in staging pediatric medulloblastomas, supratentorial primitive neuroectodermal tumors, and ependymomas. J Neurosurg Pediatr, 2010, 6:131-136.

[3] Ke CS, Deng ZD, Lei T, et al. Medullomyoblastoma report of an adult case and review of literatures. Zhongguo Xian Dai Shen Jing Ji Bing Za Zhi, 2008, 8:567-571.[柯昌庶, 邓仲端, 雷霆, 等. 髓母肌母细胞瘤：一例成人病例报告及文献复习. 中国现代神经疾病杂志, 2008, 8:567-571.]

[4] Soylemezoglu F, Soffer D, Onol B. Lipomatous medulloblastoma in adults: a distinct clinicopathological entity. Am J Surg Pathol, 1996, 20:413-418.

[5] Bechtel JT, Patton JM, Takei Y. Mixed mesenchymal and neuroectodermal tumor of the cerebellum. Acta Neuropathol, 1978, 41:261-263.

[6] Dhall G. Medulloblastoma. J Child Neurol, 2009, 24:1418-1430.

[7] Sousa R, Sá G, Reimao S, et al. Adult cerebellar medulloblastoma: imaging findings in eight cases. Acta Med Port, 2006, 19:466-470.

[8] Elshihabi S, Husain M, Linskey M. Lipomatous medulloblastoma: a rare adult tumor variant with a uniquely favorable prognosis. Surg Neurol, 2003, 60:566-570.

[9] Sharma MC, Agarwal M, Suri A, et al. Lipomedulloblastoma in a child: a controversial entity. Hum Pathol, 2002, 33:564-569.

[10] Giordana MT, Schiffer P, Boghi A, et al. Medulloblastoma with lipidized cells versus lipomatous medulloblastoma. Clin Neuropathol, 2000, 19:273-277.

[11] Owler BK, Makeham JM, Shingde M, et al. Cerebellar liponeurocytoma. J Clin Neurosci, 2005, 12:326-329.

[12] Gupta K, Salunke P, Kalra I, et al. Central liponeurocytoma: case report and review of literature. Clin Neuropathol, 2011, 30:80-85.

[13] Chakraborti S, Mahadevan A, Govindan A, et al. Supratentorial and cerebellar liponeurocytomas: report of four cases with review of literature. J Neurooncol, 2011, 103:121-127.

[14] Kleihues P, Chimelli L, Giangaspero F. Cerebellar liponeurocytoma//Louis DN, Ohgaki H, Wiestler OD, et al. WHO classification of tumours of the nervous system. Lyon: IARC, 2007: 110-112.

[15] U - King - Im JM, Taylor MD, Raybaud C. Posterior fossa ependymomas: new radiological classification with surgical correlation. Childs Nerv Syst, 2010, 26:1765-1772.

[16] Chakraborti S, Govindan A, Alapatt JP, et al. Primary myxopapillary ependymoma of the fourth ventricle with cartilaginous metaplasia: a case report and review of the literature. Brain Tumor Pathol, 2012, 29:25-30.

[17] Padhi S, Sarangi R, Challa S, et al. A 10 - year retrospective study of hemangioblastomas of the central nervous system with reference to von Hippel-Lindau (VHL) disease. J Clin Neurosci, 2011, 18:939-944.

[18] Nagamatsu K, Kumabe T, Watanabe M ,et al. Differentiation of clear cell ependymoma of the cerebellum from hemangioblastoma by thallium-201 single photon emission computed tomography-case report-. Neurol Med Chir (Tokyo), 2009, 49:608-611.

[19] Plate KH, Vortmeyer AO, Zagzag D, et al. Von Hippel-Lindau disease and haemangioblastoma//Louis DN, Ohgaki H, Wiestler OD, et al. WHO classification of tumours of the nervous system. Lyon: IARC, 2007: 215-217.

[20] Giannini C, Paulus W, Louis DN, et al. Pleomorphic xanthoastrocytoma//Louis DN, Ohgaki H, Wiestler OD, et al. WHO classification of tumours of the nervous system. Lyon: IARC, 2007: 22-24.

[21] Gessi M, von Bueren AO, Rutkowski S, et al. p53 expression predicts dismal outcome for medulloblastoma patients with metastatic disease. J Neurooncol, 2012, 106:135-141.

[22] Horstmann S, Perry A, Reifenberger G, et al. Genetic and expression profiles of cerebellar liponeurocytomas. Brain Pathol, 2004, 14:281-289.

[23] Aker FV, Ozkara S, Eren P, et al. Cerebellar liponeurocytoma/ lipidized medulloblastoma. J Neurooncol, 2005, 71:53-59.

[24] Limaiem F, Bellil S, Chelly I, et al. Recurrent cerebellar liponeurocytoma with supratentorial extension. Can J Neurol Sci,

2009, 36:662-665.
[25] Dong J. Rethink and re-recognition of the 5-year study on brain tumor stem cells. Zhongguo Xian Dai Shen Jing Ji Bing Za Zhi, 2008, 8:384-387.[董军. 对脑肿瘤干细胞五年研究历程的反思及再认识. 中国现代神经疾病杂志, 2008, 8:384-387.]
（收稿日期：2012-04-16）

·临床医学图像·

血管母细胞瘤

DOI：10.3969/j.issn.1672-6731.2012.03.027

Hemangioblastoma

HAN Tong
Department of Neuroradiology, Tianjin Huanhu Hospital, Tianjin 300060, China (Email: mrbold@163.com)

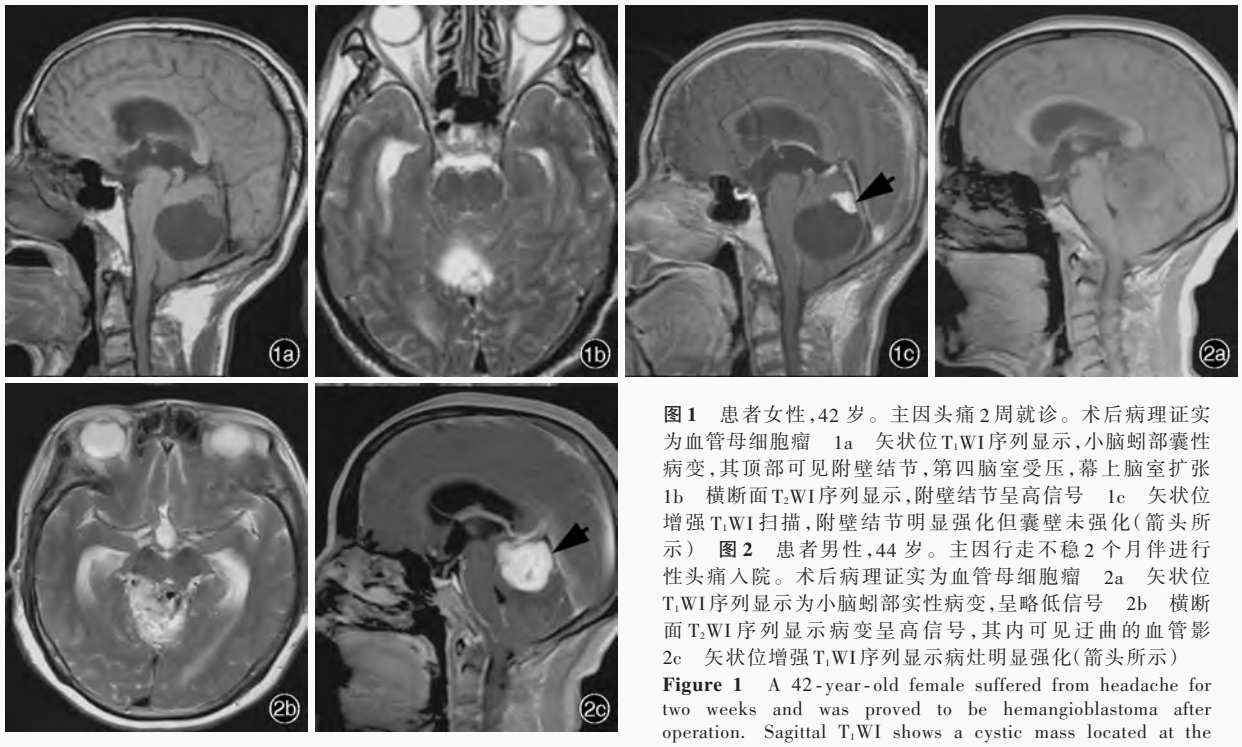

图1 患者女性，42岁。主因头痛2周就诊。术后病理证实为血管母细胞瘤 1a 矢状位T_1WI序列显示，小脑蚓部囊性病变，其顶部可见附壁结节，第四脑室受压，幕上脑室扩张 1b 横断面T_2WI序列显示，附壁结节呈高信号 1c 矢状位增强T_1WI扫描，附壁结节明显强化但囊壁未强化（箭头所示） **图2** 患者男性，44岁。主因行走不稳2个月伴进行性头痛入院。术后病理证实为血管母细胞瘤 2a 矢状位T_1WI序列显示为小脑蚓部实性病变，呈略低信号 2b 横断面T_2WI序列显示病变呈高信号，其内可见迂曲的血管影 2c 矢状位增强T_1WI序列显示病灶明显强化（箭头所示）

Figure 1 A 42-year-old female suffered from headache for two weeks and was proved to be hemangioblastoma after operation. Sagittal T_1WI shows a cystic mass located at the cerebellar vermis with a mural nodule on the roof. The fourth ventricle was compressed with enlargement of the third ventricle (Panel 1a). The mural nodule was high intensity on T_2WI transverse section (Panel 1b) and enhanced obviously after injection of contrast. The cyst wall demonstrated no enhancement (arrow indicates, Panel 1c) **Figure 2** A 44-year-old male suffered from walking instability for two months and was proved to be hemangioblastoma after operation. There is a solid lesion with slightly hypointetsity on sagittal T_1WI (Panel 2a) and high intensity on T_2WI (Panel 2b). Several serpentine tumor vessels are found within the lesion and the tumor demonstrates marked enhancement on post-contrast scans (arrow indicates, Panel 2c)

血管母细胞瘤（HGB）为起源于脑膜的中枢神经系统肿瘤，WHO Ⅰ级，占颅内肿瘤的1%～2%、颅后窝肿瘤的7%。好发于成年人。可分为散发性和家族性两种类型，前者约占75%，于中老年发病，多见于小脑半球；后者伴发于von Hippel-Lindau病（VHL），青少年期即可出现症状，主要位于小脑，小部分位于视觉通路。血管母细胞瘤由密集不成熟的血管组织构成，囊性变是其突出特点，囊变体积远超过肿瘤本身，将肿瘤推向一侧，使其成为附壁结节。影像学检查分为大囊小结节、单纯及实质肿块3种类型。（1）大囊小结节型（图1）：最为常见，结节于CT呈等或稍高密度，MRI呈等信号，囊性部分为等或高于脑脊液信号；增强扫描结节明显强化而囊壁不强化。其发病年龄和特征性强化改变具有鉴别诊断意义，应与囊性胶质瘤、转移瘤、小脑单纯囊肿或表皮样囊肿等鉴别。（2）单纯囊性型：临床少见，应与小脑囊肿相鉴别。（3）实质肿块型（图2）：较为少见，CT呈等密度或混杂密度；MRI于T_2WI序列呈不均质高信号，瘤内/瘤周多见血管影，瘤周水肿明显；肿瘤由丰富的幼稚血管所构成且无囊性变，故增强后显著强化；瘤内/瘤周异常流空血管影具有鉴别意义，应与脑膜瘤、单发转移瘤、淋巴瘤、恶性胶质瘤相鉴别。散发性囊性血管母细胞瘤手术疗效满意，而实质性、家族性、多发性血管母细胞瘤预后不良。

（天津市环湖医院神经放射科韩彤供稿）