

## · 临床病理报告 ·

# 鞍区和鞍上脑室外神经细胞瘤

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**【摘要】目的** 探讨发生于鞍区和鞍上的脑室外神经细胞瘤的临床病理学特征。**方法** 对1例鞍区和鞍上脑室外神经细胞瘤患者的临床表现、影像学特征、组织学形态、免疫表型和分子遗传学特征进行回顾分析并复习相关文献。**结果**女性患者,27岁,临床表现为反复头痛伴双眼视物模糊5个月。头部MRI显示鞍区和鞍上占位性病变,T<sub>1</sub>WI呈等或低信号,T<sub>2</sub>WI呈高或低混杂信号,扩散加权成像呈稍高信号,界限清晰,正常垂体结构显示不清。临床诊断为垂体腺瘤,行经鼻蝶入路垂体腺瘤切除术+脑脊液鼻漏修补术+视神经减压术,手术全切除肿瘤。组织学形态可见肿瘤细胞呈弥漫浸润性生长,部分区域可见神经毡背景;肿瘤细胞大小和形态相对一致,胞核圆形或卵圆形,染色质细腻深染,未见核分裂象。免疫组织化学染色可见肿瘤细胞胞核表达神经元核抗原和甲状腺转录因子-1,胞核和胞质表达钙视网膜蛋白,胞质表达突触素、嗜铬素A、上皮钙黏素和基质金属蛋白酶-9;胞核局灶性表达S-100蛋白,胞质局灶性表达神经微丝蛋白、细胞角蛋白8和波形蛋白;Ki-67抗原标记指数约为3%。网织纤维染色呈阴性。基因检测可见肿瘤细胞无异柠檬酸脱氢酶基因突变,无1p/19q-共缺失。最终病理诊断为脑室外神经细胞瘤(WHOⅡ级)。**结论**鞍区和鞍上脑室外神经细胞瘤临床极为罕见,组织学形态与发生于脑室的中枢神经细胞瘤相似,表现为肿瘤弥漫浸润性生长,肿瘤细胞形态较一致,胞核圆形,可见神经毡背景和“树枝”状薄壁毛细血管。应注意与垂体腺瘤、少突胶质细胞瘤和透明细胞型室管膜瘤等相鉴别。

**【关键词】** 神经细胞瘤; 蝶鞍; 病理学; 免疫组织化学

## Sellar/suprasellar extraventricular neurocytoma

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**【Abstract】Objective** To explore the clinicopathological features of extraventricular neurocytoma located in the sellar/suprasellar region. **Methods** The clinical manifestations, neuroimaging, histopathological, immunohistochemical and molecular genetic features were retrospectively analyzed in one case of sellar/suprasellar extraventricular neurocytoma, and the related literatures were reviewed. **Results** A 27-year-old female presented with intermittent headache, accompanied by blurred vision for 5 months. Head MRI demonstrated a mass with a well-defined margin measuring 3.80 cm × 2.50 cm × 3.40 cm located in the sellar/suprasellar region. The tumor showed isointense to hyperintense signals on T<sub>1</sub>WI and hyperhypointense mixed signals on T<sub>2</sub>WI, and slightly hyperintense signal on diffusion-weighted imaging (DWI). The pituitary was not shown. A transsphenoidal sellar tumor resection, cerebrospinal fluid (CSF) rhinorrhea repairing and optic decompression were performed. The mass was lightly yellow and tough with abundant blood supply and filled with old hemorrhage. The pituitary tissue was pushed to the left rear. Microscopy examination showed a diffuse invasive growth pattern with neuropil background in some area. The tumor cells were uniform on size and shape with round to oval, exquisite and hyperchromatic nuclei. No mitosis was found. Immunohistochemical staining showed the tumor cells were positive for neuronal nuclei (NeuN) and thyroid transcription factor-1 (TTF-1) in nuclei, calretinin (CR) in nuclei and cytoplasm, synaptophysin (Syn), chromogranin A (CgA), E-cadherin, matrix metalloproteinase-9 (MMP-9) in cytoplasm, and focally positive for S-100 protein (S-100) in nuclei, and neurofilament protein (NF), cytokeratin 8 (CK8) and

doi:10.3969/j.issn.1672-6731.2017.12.009

基金项目:福建省自然科学基金资助项目(项目编号:2014J01413)

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vimentin (Vim) in cytoplasm. The Ki-67 labeling index was about 3%. The tumor tissue was negative for reticular fiber staining. Molecular genetic analysis showed that isocitrate dehydrogenase 1 (IDH) gene was not mutated, and 1p/19q was intact in tumor cells. The final pathological diagnosis was extraventricular neurocytoma, WHO grade II. **Conclusions** Extraventricular neurocytoma located in the sellar/suprasellar region is very rare. The histological features are similar to central neurocytoma in ventricle. Tumor cells were in diffusely invasive growth and were uniform in size and shape, with round nuclei. Fibrillary areas mimicking neurophil and branching thin-walled capillaries can be seen. The differential diagnosis includes pituitary adenoma, oligodendrogloma, clear cell ependymoma, and so on.

**【Key words】** Neurocytoma; Sellar turcica; Pathology; Immunohistochemistry

This study was supported by Natural Science Foundation of Fujian Province, China (No. 2014J01413).

中枢神经细胞瘤(CN)是一种发生于中枢神经系统的相对罕见肿瘤,占全部中枢神经系统肿瘤的0.3%~0.6%<sup>[1]</sup>,好发于脑室系统,亦见于脑室系统以外,即脑室外神经细胞瘤(EVN)<sup>[2]</sup>。发生于鞍区和鞍上的脑室外神经细胞瘤极为罕见,目前仅见数例报道<sup>[1-7]</sup>。本文回顾分析1例鞍区和鞍上脑室外神经细胞瘤患者的诊断与治疗过程,并复习相关文献,探讨其临床表现、影像学特征、组织学形态和免疫表型、基因检测以及诊断与鉴别诊断要点。

### 病历摘要

**患者** 女性,27岁,主因反复头痛伴双眼视物模糊5个月,于2016年2月23日入院。患者5个月前无明显诱因出现头部闷痛,中等程度,偶伴头昏,每日均有发作,可自行缓解,伴双眼视物模糊、视力下降、复视,症状反复发作,同时伴性功能减退。门诊行头部MRI检查显示,鞍内和鞍上占位性病变,增强扫描病灶呈不均匀强化,考虑垂体肿瘤,遂以“垂体肿瘤”收入院。患者自发病以来,精神、饮食佳,睡眠尚可,大小便正常,体重无明显变化。

既往史、个人史及家族史均无特殊。

**体格检查** 患者体温36.8℃,心率86次/min,呼吸20次/min,血压128/84 mm Hg(1 mm Hg=0.133 kPa)。神志清楚,语言尚流利,矫正视力右眼0.8、左眼0.4,双侧瞳孔等大、等圆,直径约3mm,对光反射灵敏,眼球各向活动充分,无眼震、双睑下垂;四肢肌力5级,肌张力正常,腱反射对称活跃,深浅感觉大致正常,共济运动正常,病理反射未引出,颈部柔软,双侧Kernig征阴性。

**辅助检查** 实验室检查:血尿便常规、血液化学、下丘脑-垂体激素均于正常值范围。影像学检查:头部MRI显示,鞍内和鞍上团块状混杂信号影,T<sub>1</sub>WI呈等或低信号(图1a)、T<sub>2</sub>WI呈高或低混杂信

号、扩散加权成像(DWI)呈稍高信号,病灶内可见多发液液平面,界限清晰,大小约3.80 cm×2.50 cm×3.40 cm,蝶鞍扩大,正常垂体结构不清,视交叉受压上移,鞍上池明显受压变窄,双侧颞叶呈弧形受压改变;增强扫描显示,鞍内和鞍上团块状病灶边缘不规则环状强化,病灶中央未见明显强化(图1b)。

**诊断与治疗经过** 临床诊断为垂体腺瘤。遂于入院后第4天(2016年2月26日)于全身麻醉下行经鼻蝶入路垂体腺瘤切除术+脑脊液鼻漏修补术+视神经减压术,术中于鞍底可见黄色肿瘤,大小约3.80 cm×3.50 cm×3.00 cm,质地坚韧,血运丰富,其内充满大量陈旧性出血,垂体受压向左后方移位,部分肿瘤组织质地较软,与鞍膈粘连紧密。手术全切除肿瘤,切除标本行组织病理学检查。(1)大体标本观察:手术切除标本呈灰褐色破碎组织一堆,大小约2.00 cm×1.00 cm×0.30 cm,质地柔软,无明显包膜。经体积分数为10%中性甲醛溶液固定,常规脱水、透明、石蜡包埋,制备3 μm层厚脑组织切片,行HE染色、免疫组织化学染色和特殊染色。(2)HE染色:肿瘤组织呈弥漫浸润性生长,由较一致的呈片状排列的中等大小肿瘤细胞组成(图2a),具有圆形胞核和“椒盐”样染色质,伴少量胞质,局灶细胞增生活跃,部分区域可见神经毡背景(图2b),薄壁毛细血管呈“树枝”状。(3)免疫组织化学染色:采用EnVision二步法,检测用试剂盒购自丹麦Dako公司,检测用抗体主要包括神经元核抗原(NeuN)、钙视网膜蛋白(CR)、突触素(Syn)、甲状腺转录因子-1(TTF-1,克隆号:SPT24)、嗜铬素A(CgA)、上皮钙黏素(E-cadherin)、基质金属蛋白酶-9(MMP-9)、神经微丝蛋白(NF)、细胞角蛋白8(CK8)、波形蛋白(Vim)、S-100蛋白(S-100)、上皮膜抗原(EMA)、少突胶质细胞转录因子2(Olig-2)、胶质纤维酸性蛋白(GFAP)、下丘脑-垂体激素和Ki-67抗原,均购自丹

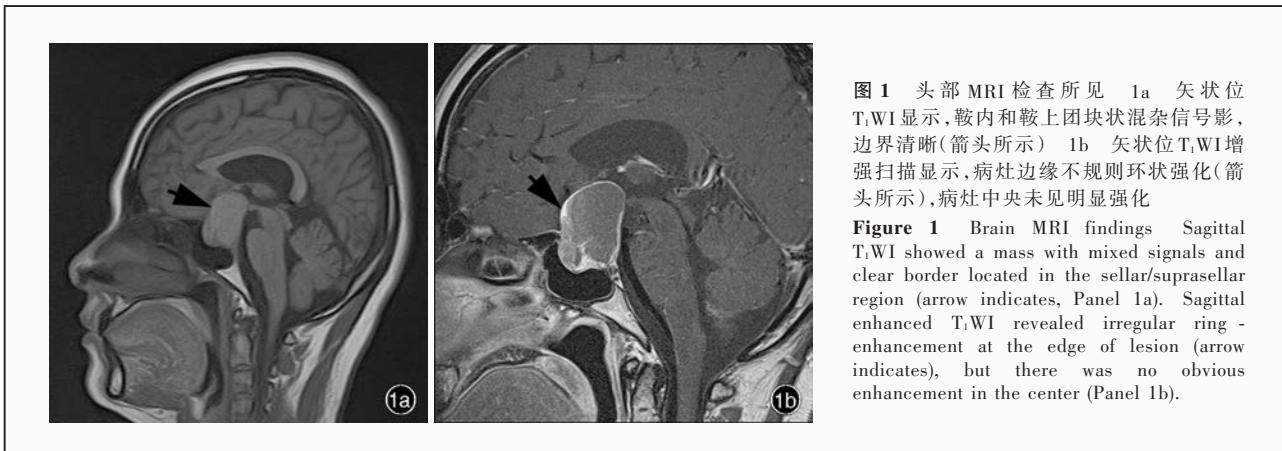


图 1 头部 MRI 检查所见 1a 矢状位 T<sub>1</sub>WI 显示,鞍内和鞍上团块状混杂信号影,边界清晰(箭头所示) 1b 矢状位 T<sub>1</sub>WI 增强扫描显示,病灶边缘不规则环状强化(箭头所示),病灶中央未见明显强化

**Figure 1** Brain MRI findings. Sagittal T<sub>1</sub>WI showed a mass with mixed signals and clear border located in the sellar/suprasellar region (arrow indicates, Panel 1a). Sagittal enhanced T<sub>1</sub>WI revealed irregular ring-enhancement at the edge of lesion (arrow indicates), but there was no obvious enhancement in the center (Panel 1b).

麦 Dako 公司。结果显示,肿瘤细胞胞核表达 NeuN (图 3a)、CR(图 3b) 和 TTF-1(图 3c),胞质表达 CR(图 3b)、Syn(图 3d)、CgA、E-cadherin 和 MMP-9,胞核局灶性表达 S-100,胞质局灶性表达 NF、CK8 和 Vim;不表达 EMA、Olig-2、GFAP 和下丘脑-垂体激素;Ki-67 抗原标记指数约为 3%。(4)特殊染色:采用福建医科大学附属第一医院病理科自行制备的 Gomori 银氨溶液进行网织纤维染色,肿瘤细胞间网织纤维染色呈阴性。(5)基因检测:采用 Sanger 测序检测异柠檬酸脱氢酶(IDH)基因,上下游引物和检测用试剂盒为福建医科大学附属第一医院分子病理实验室自行制备,采用荧光原位杂交(FISH)检测 1 号染色体短臂和 19 号染色体长臂(1p/19q)状态,检测用试剂盒购自美国 Abbott 公司。结果显示,肿瘤细胞无 IDH 基因突变,无 1p/19q 缺失。结合免疫组织化学染色、特殊染色和基因检测结果,最终病理诊断为脑室外神经细胞瘤(WHO II 级)。患者共住院 8 d,术后予醋酸去氨加压素片(0.05 mg/8 h)和尼莫地平(20 mg/次、3 次/d)口服,连续 7 d。出院时未诉头痛、头晕,自觉视力提高。术后随访 14 个月,肿瘤无复发。

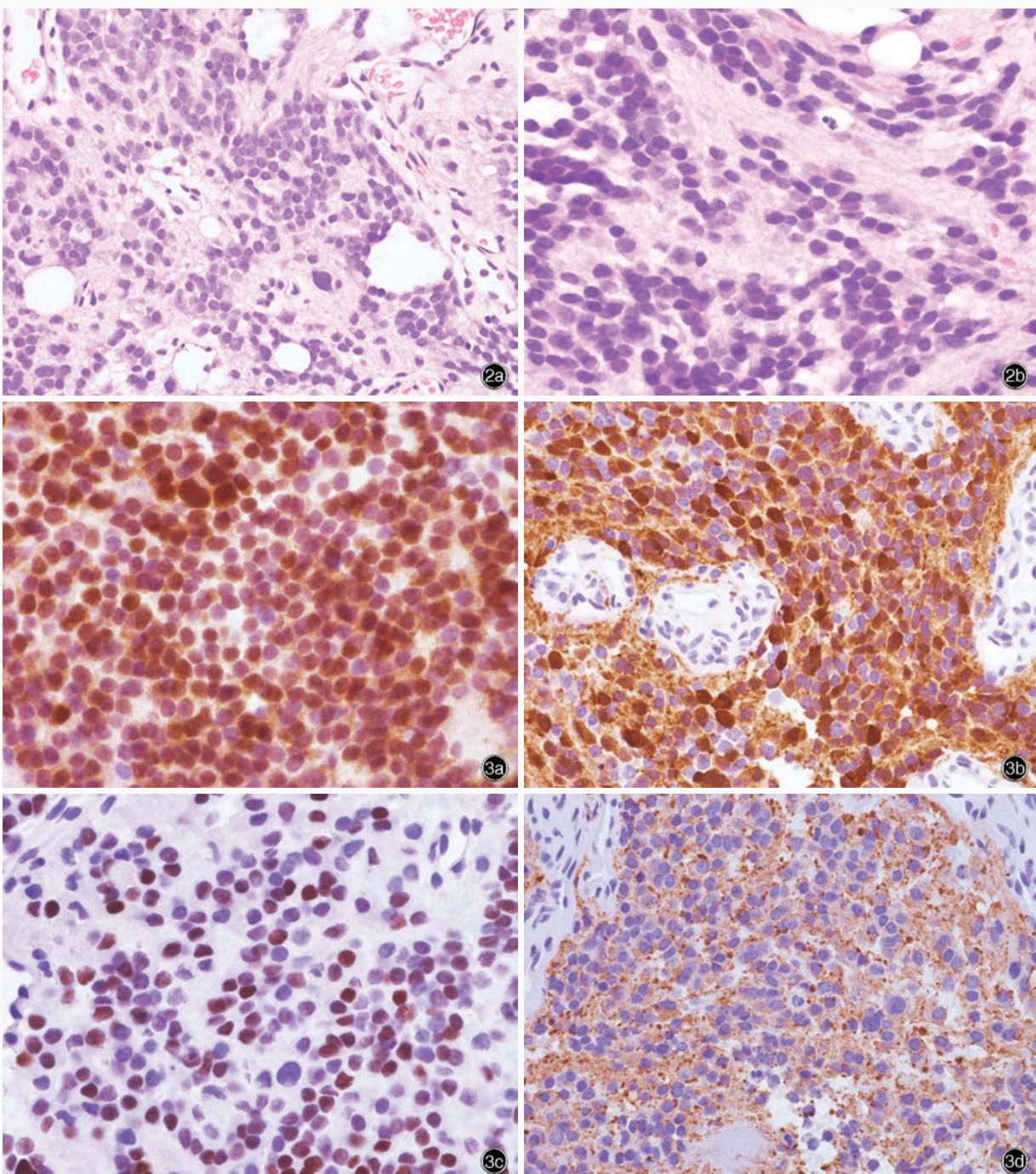
## 讨 论

神经细胞瘤源于具有双重分化潜能的神经胶质祖细胞,向神经元和星形胶质细胞分化<sup>[1]</sup>。中枢神经细胞瘤由 Hassoun 于 1982 年命名<sup>[1]</sup>,占所有中枢神经系统肿瘤的 0.1%~0.5%<sup>[2,4]</sup>,并被 1993 年世界卫生组织(WHO)中枢神经系统肿瘤分类第 2 版收录<sup>[1]</sup>。中枢神经细胞瘤可发生于任何年龄段,好发生于 30 余岁青壮年<sup>[2]</sup>,也可发生于儿童<sup>[8]</sup>;主要发生于脑室系统,尤其是透明隔、第三脑室和侧脑

室<sup>[2]</sup>,也可发生于脑室系统以外<sup>[2]</sup>。脑室外神经细胞瘤已作为一种新的独立疾病于 2007 年纳入 WHO 中枢神经系统肿瘤分类第 4 版,属 WHO II 级<sup>[9]</sup>,预后较好<sup>[4]</sup>。脑室外神经细胞瘤好发于儿童和青壮年,中位发病年龄为 34 岁<sup>[2]</sup>,无明显性别差异<sup>[10]</sup>,亦有文献报道男性稍多发<sup>[9]</sup>,还可发生于大脑半球、丘脑、小脑、脑桥、杏仁核、松果体、下丘脑<sup>[11]</sup>、视网膜和脊髓等部位<sup>[2]</sup>,发生于鞍区和鞍上者极为少见。

结合文献,分析并总结所报道的 6 例鞍区和鞍上脑室外神经细胞瘤患者的临床特点(表 1),其中视力下降是其共同特点<sup>[1-6]</sup>,本文患者即表现为视力下降。其中 1 例还出现间歇性头痛<sup>[3]</sup>,与本文患者相似。除以上临床症状外,发生于鞍区和鞍上的脑室外神经细胞瘤无其他阳性体征,内分泌功能测定除个别患者催乳素(PRL)轻度升高、雌激素轻度下降外,余下丘脑-垂体激素均于正常值范围<sup>[1-6]</sup>。本文患者黄体生成素(LH)、卵泡刺激素(FSH)、生长激素(GH)、促肾上腺皮质激素(ACTH)等均于正常值范围。

脑室外神经细胞瘤在 MRI 上表现为界限清晰、囊性或不均匀的实质性肿物,累及大脑半球白质或灰质<sup>[5]</sup>,伴或不伴钙化,可见瘤周水肿,偶可见瘤内出血<sup>[2,12]</sup>,T<sub>1</sub>WI 呈等或稍低信号,T<sub>2</sub>WI 呈等信号,增强扫描病灶呈或不呈强化征象<sup>[1-2]</sup>。Yi 等<sup>[13]</sup>对 10 例存在 IDH1 基因突变的脑室外神经细胞瘤患者进行 MRI 研究,肿瘤组织可见囊性变和钙化,但几乎未见瘤周水肿和瘤内出血。结合文献,对包括本文患者在内的 7 例鞍区和鞍上脑室外神经细胞瘤患者的影像学资料进行分析和总结,头部 CT 可见鞍区和鞍上呈等或稍高密度影,伴或不伴钙化;MRI 表现为 T<sub>1</sub>WI 呈低或等信号,T<sub>2</sub>WI 呈低、等或高信号,增强扫



**图2** 光学显微镜观察所见 HE染色 2a 肿瘤组织呈弥漫浸润性生长,由较一致的呈片状排列的中等大小肿瘤细胞组成  $\times 200$  2b 肿瘤细胞胞核呈规则圆形,染色质呈“椒盐”样,可见典型神经毡  $\times 400$  **图3** 光学显微镜观察所见 免疫组织化学染色(EnVision二步法)  $\times 400$  3a 肿瘤细胞胞核NeuN呈弥漫性强阳性 3b 肿瘤细胞胞核和胞质CR呈强阳性 3c 肿瘤细胞胞核TTF-1呈中等至强阳性 3d 肿瘤细胞胞质Syn呈阳性

**Figure 2** Optical microscopy findings HE staining The tumor was composed of uniform, middle-sized cells with diffuse and invasive growth pattern (Panel 2a).  $\times 200$  The cells had round to oval nuclei with salt-and-pepper chromatin. Typical neuropil can be seen (Panel 2b).  $\times 400$  **Figure 3** Optical microscopy findings Immunohistochemical staining (EnVision)  $\times 400$  The tumor cells were diffusely positive for NeuN in nuclei (Panel 3a). The tumor cells were strongly positive for CR in nuclei and cytoplasm (Panel 3b). The tumor cells were moderately to strongly positive for TTF-1 in nuclei (Panel 3c). The tumor cells were positive for Syn in cytoplasm (Panel 3d).

描病灶可见强化征象。

鞍区和鞍上脑室外神经细胞瘤的组织学形态

特征与中枢神经细胞瘤相似,表现为形态一致的圆形细胞<sup>[9]</sup>,胞质少至中等,胞核形态规则,呈圆形或

**表1** 文献报道和本文鞍区和鞍上脑室外神经细胞瘤患者的临床病理学特征**Table 1.** Clinicopathological features of sellar/suprasellar extraventricular neurocytoma reported in literatures and this case

Study	Sex	Age (year)	Preoperative diagnosis	Clinical feature	Image	Histological feature	Immunohistochemistry	Resection extent	Adjuvant therapy	Follow up
Wang, et al. <sup>[1]</sup>	Female	50	Pituitary adenoma	Decreased vision for 2 months	A solid sellar/suprasellar mass with homogenous enhancement and spot calcification	The tumor cells had growth patterns of nests and islands, with neutropil and calcification	Syn, CgA, NF, CD56: +	Subtotal resection	Radiotherapy	No recurrence after follow up for 18 months
Peng, et al. <sup>[2]</sup>	Male	56	Pituitary adenoma	Found the tumor by accident on CT after a head injury	T <sub>1</sub> WI showed a circumscribed dumbbell- shaped hypointense pituitary mass; the mass was iso-intense and heterogeneous enhancement on T <sub>2</sub> WI	The tumor was composed of small- medium-sized cells with growth pattern of sheets; there were neutropil and thin- walled capillary-sized vessels	Syn, CgA, NSE: +; hypophyseal hormones, GFAP: -; Ki-67: 0%-1%	Total resection	None	None
Wang, et al. <sup>[3]</sup>	Female	23	Pituitary adenoma	Found the tumor by accident on CT after a head injury	MRI showed a solid and well-defined mass, with iso-intense and homogeneous enhancement on T <sub>1</sub> WI	The tumor had round cells, hyperchromatic nuclei and moderate amount of clear cytoplasm	Syn: +; NF, S-100, GFAP, CK, EMA: -; Ki-67: 1%	Subtotal resection	Radiotherapy	None
Kawaji, et al. <sup>[4]</sup>	Male	48	Pituitary adenoma	Decreased vision for 3 months	T <sub>1</sub> WI with contrast enhancement showed an enhanced mass	Tumor cells had round nuclei and clear cytoplasms lying like cobblestones; there were linear arborizing capillary blood vessels	Syn, NF, NeuN: +; GFAP, EMA, Olig-2: -; Ki-67: 3%	Subtotal resection	Radiotherapy	Tumor recurred after 6 years with disseminations in the spinal cord
Yang, et al. <sup>[5]</sup>	Female	46	Meningioma	Decreased vision for one year	CT showed iso-intense with calcification and MRI showed enhancement with cystic changes	The tumor was composed of uniform small, round cells	Syn, NSE, Vim: +; GFAP: -	Subtotal resection	Radiotherapy	None
Wang, et al. <sup>[6]</sup>	Male	25	Pituitary adenoma or meningioma	Decreased vision for 3 months	A round, circumscribed mass with calcification	Tumor cells were oval with schistic distribution; cytoplasm was sparse with mild acidophilia; neutropil could be found	Syn, NeuN, α- internexin: +; GFAP, CK, hypophyseal hormones: -; Ki-67: 3%	Subtotal resection	Radiotherapy	None
This case	Female	27	Pituitary adenoma	Headache and blurred vision for 5 months	Iso-intense to hypointense on T <sub>1</sub> WI and hyperintense mixed hypointense signal on T <sub>2</sub> WI	The tumor cells was uniform on size and shape with round to oval nuclei; neutropil could be found	Syn, CR, CgA, NeuN, TTF-1: +; hypophyseal hormones, EMA, Olig-2, GFAP: -; Ki-67: 3%	Total resection	None	No recurrence after follow up for 14 months

+, positive, 阳性; -, negative, 阴性。Syn, synaptophysin, 突触素; CgA, chromogranin A, 嗜铬素 A; NF, neurofilament protein, 神经微丝蛋白; NSE, neuron-specific enolase, 神经元特异性烯醇化酶; GFAP, glial fibrillary acidic protein, 胶质纤维酸性蛋白; S-100, S-100 protein, S-100蛋白; CK, cytokeratin, 细胞角蛋白; EMA, epithelial membrane antigen, 上皮膜抗原; NeuN, neuronal nuclei, 神经元核抗原; Olig-2, oligodendrocytes transcription factor-2, 少突胶质细胞转录因子 2; Vim, vimentin, 波形蛋白; CR, calretinin, 钙视网膜蛋白; TTF-1, thyroid transcription factor-1, 甲状腺转录因子-1。

卵圆形, 染色质呈“椒盐”样<sup>[2,14]</sup>, 核分裂象罕见或未见<sup>[9]</sup>, 背景中可见纤细的神经毡样物质<sup>[6,14]</sup>, 薄壁毛细血管呈“树枝”状<sup>[2]</sup>。非典型脑室外神经细胞瘤的组织学形态特征为 Ki-67 抗原标记指数 > 2% 或 > 3%<sup>[15-16]</sup>, 伴或不伴非典型组织学形态特征, 如灶性坏死、血管增生和分裂活性增加, 具有较强的侵袭性, 预后较差, 复发率较高<sup>[9,14]</sup>。但非典型脑室外神经细胞瘤未被 2016 版 WHO 中枢神经系统肿瘤分类收录<sup>[9]</sup>。免疫组织化学染色对脑室外神经细胞瘤的诊断与鉴别诊断极为重要。有 50% ~ 60% 的脑室外神经细胞瘤患者可有神经节细胞或胶质细胞分化, 与中枢神经细胞瘤有所不同<sup>[14]</sup>。神经元标志物

Syn、CgA、神经元特异性烯醇化酶 (NSE)、CD56、NeuN 和 NF 呈阳性, 而 GFAP、Olig-2、S-100、巢蛋白 (Nes)、EMA 和下丘脑-垂体激素呈阴性<sup>[1-6]</sup>, P53 亦呈阴性。脑室外神经细胞瘤的分子遗传学特征为无 1p/19q-共缺失, O<sup>6</sup>-甲基鸟嘌呤-DNA 甲基转移酶 (MGMT) 启动子甲基化和表皮生长因子受体 (EGFR) 基因扩增频率低<sup>[2]</sup>。亦有研究显示, 部分脑室外神经细胞瘤存在 1p/19q-共缺失<sup>[17]</sup>。本文患者免疫组织化学染色结果与文献报道相似, 但胞核弥漫性强阳性表达 TTF-1, 尚未见诸报道。Kristensen 等<sup>[18]</sup>对中枢神经系统肿瘤 TTF-1 免疫组织化学染色进行研究, 发现仅少部分肿瘤中呈阳性表达, 3 例中

枢神经细胞瘤患者中1例弥漫性中度表达TTF-1。神经系统发育过程中,TTF-1表达局限于端脑和间脑区域细胞,包括下丘脑神经元、第三脑室室管膜细胞、伸长细胞和垂体后叶胶质细胞<sup>[18]</sup>,但不表达于不成熟的大脑皮质或侧脑室<sup>[19]</sup>。脑组织中TTF-1分子靶点作用是上调中间丝巢蛋白、少突胶质细胞瘤特化、中间神经元迁移、GH和PRL转录、脉络丛水通道蛋白1(AQP1)合成的基因表达<sup>[18]</sup>,但中枢神经细胞瘤表达TTF-1的机制尚待进一步阐述。

鞍区和鞍上脑室外神经细胞瘤应注意与以下肿瘤相鉴别。(1)垂体腺瘤:鞍区和鞍上脑室外神经细胞瘤在影像学上常误诊为垂体腺瘤,但免疫组织化学染色LH、FSH、GH、PRL、ACTH和TSH呈阴性可以排除诊断。(2)少突胶质细胞瘤:脑室外神经细胞瘤与少突胶质细胞瘤均具有圆形一致的胞核和透亮胞质,但神经节细胞分化提示脑室外神经细胞瘤<sup>[14]</sup>,且少突胶质细胞瘤更具侵袭性,常呈浸润性生长,Syn呈阴性<sup>[20]</sup>,虽有时可呈阳性,但程度低于脑室外神经细胞瘤<sup>[14]</sup>。*IDH*基因突变和1p/19q-共缺失最具鉴别诊断价值。(3)透明细胞型室管膜瘤:组织学形态可见血管周围假“菊形团”样结构,GFAP和EMA呈阳性<sup>[21]</sup>。其他鉴别诊断还包括胚胎发育不良性神经上皮肿瘤(DNT)和松果体瘤等<sup>[1]</sup>。

手术全切除肿瘤是脑室外神经细胞瘤的首选治疗方法<sup>[22]</sup>,预后较好<sup>[10]</sup>。由于发生于鞍区和鞍上的脑室外神经细胞瘤多侵犯蝶窦和海绵窦,并常压迫视神经、紧邻或包裹颈内动脉,故全切除较为困难,常采用次全切除<sup>[1,3-6]</sup>。对于次全切除的鞍区和鞍上脑室外神经细胞瘤,术后辅助放射治疗可以提高疾病控制率和患者生存率<sup>[2]</sup>。Peng等<sup>[2]</sup>对次全切除的脑室外神经细胞瘤患者术后辅以药物化疗并取得显著治疗效果。然而目前药物化疗在该肿瘤治疗中的经验仍不足,尚待更多临床实践和经验加以证实。本文患者手术全切除肿瘤,术后未辅以放射治疗和药物化疗,术后共随访14个月,未见肿瘤复发。

综上所述,鞍区和鞍上脑室外神经细胞瘤是临床极罕见肿瘤,临床主要表现为视力减退和头痛,组织学形态特征为弥漫性生长且形态较一致的细胞,胞核圆形或卵圆形,染色质呈细颗粒状,常有特征性神经毡背景,肿瘤内血管为薄壁毛细血管,核分裂象罕见,与垂体腺瘤、少突胶质细胞瘤、透明细胞型室管膜瘤等有相似之处,应注意鉴别诊断。手

术全切除肿瘤为首选治疗方法,预后较好,次全切除者可术后辅以放射治疗并长期随访。

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(收稿日期:2017-10-06)

## · 小词典 ·

## 中英文对照名词词汇(五)

 $T_2^*$ -梯度回波序列 $T_2^*$ -gradient echo sequence( $T_2^*$ -GRE)

体重指数 body mass index(BMI)

同型半胱氨酸 homocysteine(Hcy)

统计参数图 statistical parametric mapping(SPM)

突触素 synaptophysin(Syn)

微管相关蛋白-2 microtubule-associated protein-2(MAP-2)

韦氏肉芽肿病 Wegener's granulomatosis(WG)

西班牙8小时内支架取栓与内科治疗随机对照试验

Randomized Trial of Revascularization with Solitaire FR Device versus Best Medical Therapy in the Treatment of Acute Stroke due to Anterior Circulation Large Vessel Occlusion Presenting within 8-Hours of Symptom Onset (REVASCAT)

细胞角蛋白8 cytokeratin 8(CK8)

细胞外基质 extracellular matrix(ECM)

相对脑血流量 relative cerebral blood flow(rCBF)

TG相互作用因子 TG interacting factor(TGIF)

小动脉闭塞 small artery occlusion(SAO)

血管内机械取栓作为急性缺血性卒中血管内主要治疗试验

Solitaire™ with the Intention for Thrombectomy as Primary Endovascular Treatment for Acute Ischemic Stroke (SWIFT PRIME) trial

血管内治疗急性缺血性卒中的多中心随机临床试验

Multicenter Randomized CLinical Trial of Endovascular Treatment for Acute Ischemic Stroke in the Netherlands (MR CLEAN)

血管性痴呆 vascular dementia(VaD)

血管性认知损害 vascular cognitive impairment(VCI)

血-脑屏障 blood-brain barrier(BBB)

血氧水平依赖 blood oxygenation level-dependent(BOLD)

血氧水平依赖性功能磁共振成像

blood oxygenation level-dependent functional magnetic resonance imaging(BOLD-fMRI)

烟雾病 moyamoya disease(MMD)

延长急性神经功能缺损至动脉内溶栓时间的临床试验

Extending the time for Thrombolysis in Emergency Neurological Deficits with Intra-Arterial therapy (EXTEND-IA) trial

药物洗脱支架 drug-eluting stents(DES)

一氧化氮合酶 nitric oxide synthase(NOS)

乙酰胆碱 acetylcholine(ACh)

乙酰胆碱酯酶 acetylcholinesterase(AChE)

异柠檬酸脱氢酶 isocitrate dehydrogenase(IDH)

吲哚菁绿荧光血管造影术

indocyanine green angiography(ICGA)

英国牛津郡社区脑卒中项目

Oxfordshire Community Stroke Project(OCSP)

荧光原位杂交 fluorescence in situ hybridization(FISH)

硬脑膜动-静脉瘘 dural arteriovenous fistula(DAVF)

Glasgow 预后分级 Glasgow Outcome Scale(GOS)

原始神经外胚层肿瘤

primitive neuroectodermal tumor(PNET)

运动前区皮质 premotor cortex(PMC)

运动诱发电位 motor-evoked potential(MEP)

正常灌注压突破

normal perfusion pressure breakthrough(NPPB)

支架内再狭窄 in-stent restenosis(ISR)

中国生物医学文献数据库 China Biology Medicine(CBM)

中国知识基础设施工程

China National Knowledge Infrastructure(CNKI)

中枢神经细胞瘤 central neurocytoma(CN)

蛛网膜下隙出血 subarachnoid hemorrhage(SAH)

椎动脉 vertebral artery(VA)

椎-基底动脉盗血综合征

vertebrobasilar steal syndrome(VSS)

自旋回波序列 spin echo sequence(SE)

族错误率 family-wise error(FWE)