·病例报告·

鞍区脊索瘤术后椎管内多发转移一例

张培海 孙振兴 王贵怀

【关键词】 脊索瘤； 蝶鞍； 椎管； 肿瘤转移； 病例报告
【Key words】 Chordoma; Sella turcica; Spinal canal; Neoplasm metastasis; Case reports

Intraspinal cord multiple metastases postoperation of the sellar chordoma: one case report

ZHANG Pei-hai, SUN Zhen-xing, WANG Gui-huai
Department of Neurosurgery, Beijing Tsinghua Changgung Hospital, Beijing 102218, China
Corresponding author: WANG Gui-huai (Email: youngneurosurgeon@163.com)
This study was supported by Beijing Tsinghua Changgung Hospital Fund for Youth (No. 12015C1044).
Conflicts of interest: none declared

患者 男性，32岁。主因脊索瘤术后1年余，双下肢麻木、无力3个月，于2018年8月27日入院。患者1年前因“多饮多尿15天”于外院行头部MRI检查，发现鞍区占位性病变（图1），手术切除病变，术后病理诊断为脊索瘤（图2）；术后第2天发生脑脊液鼻漏，经腰大池引流术治愈，共住院25天。本次入院前3个月出现双下肢麻木、无力，逐渐进展至无法独自站立，病程中伴便秘，偶有小便失禁，遂入我院接受治疗。入院后体格检查：双下肢肌力为2级，肌张力降低，T_{12}平面以下深浅感觉减退。腰椎穿刺脑脊液检查发现可疑肿瘤细胞。头部MRI显示鞍内、鞍上及鞍背占位性病变，符合脊索瘤表现。脊椎MRI增强扫描，胸椎硬脊膜囊壁呈条形强化征象，L_{4-5}椎间隙水平及椎管内占位性病变，硬脊膜囊末端异常强化征象，考虑脊索瘤脑脊液播散可能（图3）。临床诊断考虑椎管内多发占位性病变，脊索瘤播散转移可能。征得患者及其家属同意，于2018年8月30日在全身麻醉下行椎管内肿瘤切除术。术中于L_{4-5}和L_5-S_1椎间隙硬脊膜外可见大小约为1.50 cm×2.00 cm的实性肿瘤灶，质地柔软，血供丰富，与硬脊膜粘连不紧密，手术显微镜下切除肿瘤后，进一步探查发现L_{4-5}椎间隙肿瘤灶部位有一硬脊膜小孔，蛛网膜自小孔疝出，切开并悬吊硬脊膜，可见蛛网膜明显增厚，切开蛛网膜后可见马尾神经和终丝上密布结节样肿物，呈小点样至米粒样大小不等，呈灰白色，血供不丰富，马尾神经增粗，椎管内蛛网膜下隙变窄，留取病变严重的神经根行组织病理学检查。术后病理诊断符合脊索瘤转移，组织形态学观察肿瘤组织富含黏液，黏液样间质中可见条索样或巢片状排列的立方形肿瘤细胞，部分胞质呈轻度嗜酸性；部分肿瘤细胞呈液滴状，胞质内可见大量黏液推挤胞核移位，细胞呈轻至中度异型性（图4a）。免疫组织化学染色，肿瘤细胞胞核及胞质广谱细胞角蛋白（PCK）、上皮膜抗原（EMA，图4b）、S-100蛋白（S-100）、波形蛋白（Vim）、CD31、细胞角蛋白8/18（CK8/18）呈阳性，胶质纤维酸性蛋白（GFAP）呈阴性，Ki-67抗原标记指数约为5%。结合病史、术中所见和病理检查结果，最终诊断为鞍区脊索瘤术后椎管内多发转移。术后1周腰椎MRI检查显示，L_{4-5}平面病变已切除，硬膜外积液（图5）。患者此次共住院18天，于术后1个月在外院进行质子放射治疗，照射剂量为1.80 Gy、共30天，总剂量为54 Gy。出院后随访1年，患者双下肢瘫痪，小便失禁，大便困难，未复查影像学。

讨 论

脊索瘤为临床罕见且生长较为缓慢的局部侵袭性肿瘤，起源于胚胎残留脊索组织，好发于中轴

doi：10.3969/j.issn.1672-6731.2019.12.015
基金项目：北京清华长庚医院青年启动基金资助项目（项目编号：12015C1044）
作者单位：102218 北京清华长庚医院神经外科
通讯作者：王贵怀，Email：youngneurosurgeon@163.com

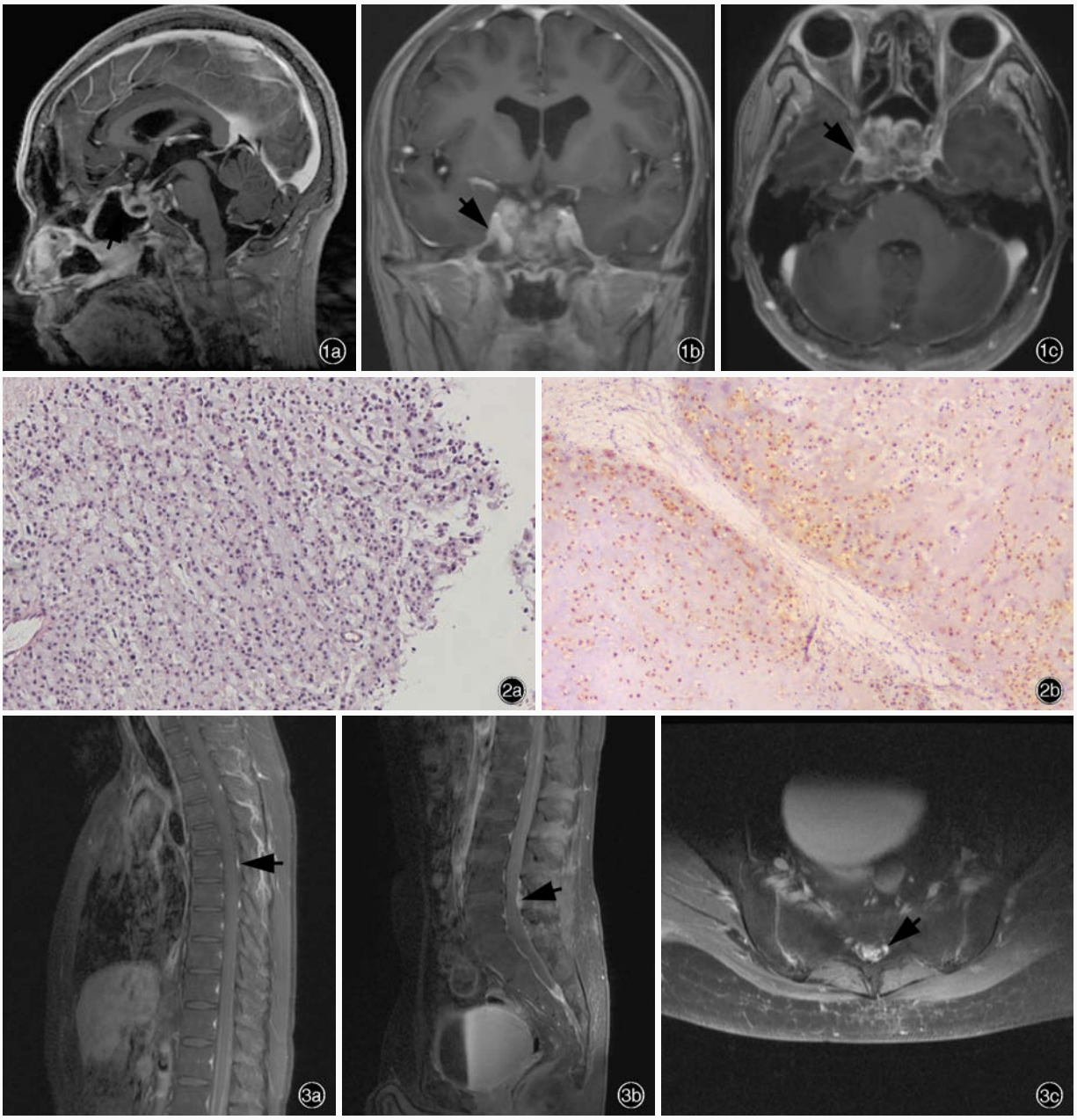

图1 首次入院时头部MRI增强扫描显示，鞍区病灶呈明显不均匀强化，大小约2.90 cm×2.80 cm×1.50 cm，边界清晰，其内多发分隔，包绕垂体柄（箭头所示） 1a 矢状位增强T_1WI 1b 冠状位增强T_1WI 1c 横断面增强T_1WI **图2** 首次术后病理学检查所见 ×100 2a 肿瘤组织富含黏液，可见呈巢片状排列的立方体状肿瘤细胞 HE染色 2b 肿瘤细胞胞质丰富，胞核呈轻至中度异型性，胞核及胞质S-100阳性 免疫组织化学染色（EnVision二步法） **图3** 再次入院时脊椎MRI增强扫描所见 3a 胸椎矢状位增强T_1WI显示，硬脊膜囊壁呈条状强化征象（箭头所示） 3b 腰骶椎矢状位增强T_1WI显示，L_{4-5}椎间隙水平及骶管内呈局限性结节样病变（箭头所示） 3c 骶椎横断面增强T_1WI显示，骶管内呈明显强化征象（箭头所示），局部可见明显不规则硬脊膜增厚、强化

Figure 1 Cranial enhanced MRI during the first admission showed a significant uneven enhancement in the sellar region, with a size of about 2.90 cm × 2.80 cm × 1.50 cm. The border was clear and there were multiple separations around the pituitary stalk (arrows indicate). Sagittal enhanced T_1WI (Panel 1a). Coronal enhanced T_1WI (Panel 1b). Axial enhanced T_1WI (Panel 1c). **Figure 2** The first postoperative pathological examination findings ×100 Tumor tissue was rich in mucus, and could be seen in a nested sheet of cubic tumor cells (Panel 2a). HE staining Tumor cells were abundant in cytoplasm, and the nucleus was mild to moderately heterotypic. The expression of S-100 in the nucleus and cytoplasm was positive (Panel 2b). Immunohistochemical staining (EnVision) **Figure 3** Spine enhanced MRI at the time of readmission Thoracic sagittal enhanced T_1WI showed signs of strip enhancement in the dural wall (arrow indicates, Panel 3a). Lumbar sacral sagittal enhanced T_1WI showed L_{4-5} intervertebral space level and localized nodular lesions in the sacral canal (arrow indicates, Panel 3b). Lumbar sacral axial enhanced T_1WI showed obvious signs of enhancement in the fistula (arrow indicates). The local area showed obvious irregular dural thickened and enhanced (Panel 3c).

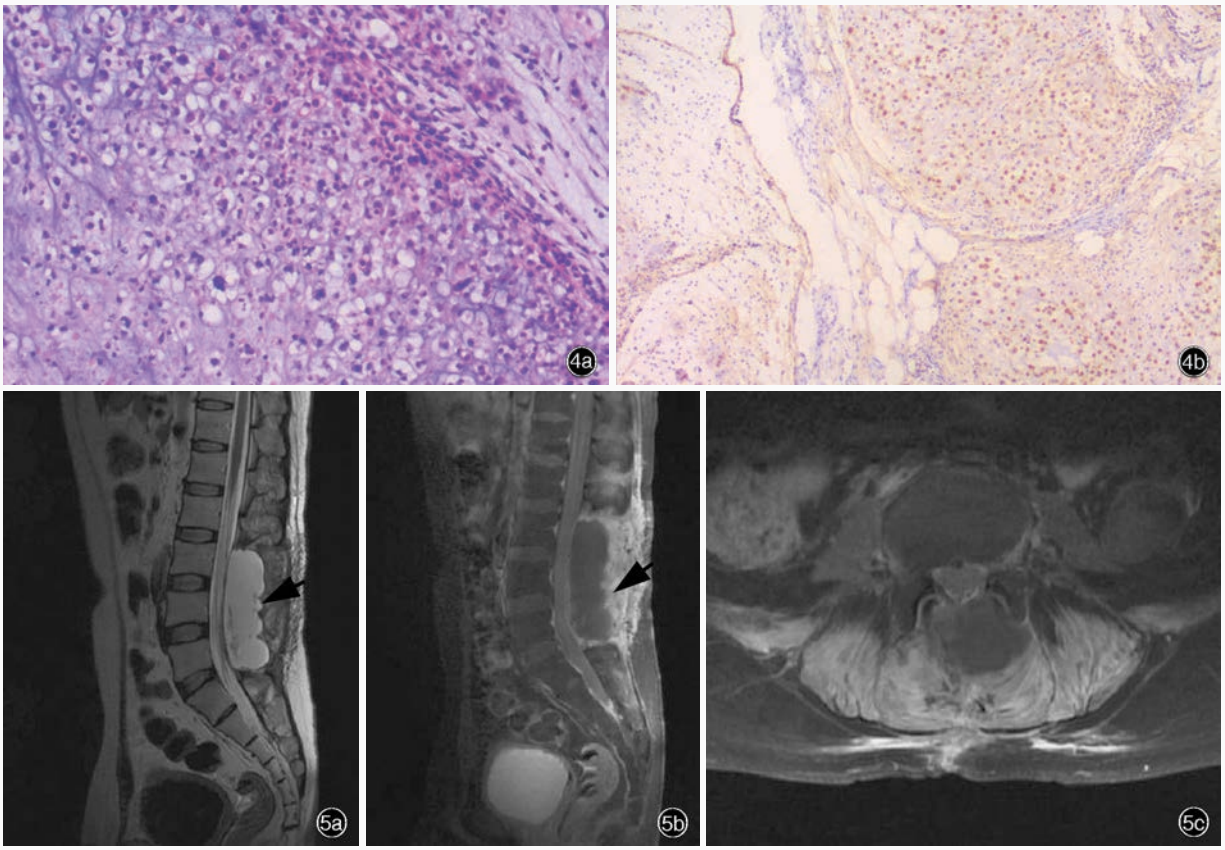

图4 再次入院术后病理学检查所见 ×100 4a 肿瘤组织富含黏液，黏液样间质中可见呈条索状或巢片状排列的肿瘤细胞，呈立方形，部分胞质轻度嗜酸性；部分肿瘤细胞呈液滴状，胞质内可见大量黏液推挤胞核移位，细胞呈轻至中度异型性 HE染色 4b 肿瘤细胞胞核呈中度异型性，胞质丰富，EMA阳性 免疫组织化学染色（EnVision二步法） **图5** 术后1周腰椎MRI检查所见 5a 矢状位T_2WI可见，术区硬脊膜外积液（箭头所示） 5b 矢状位增强T_1WI显示，L_{4-5}椎间隙水平结节状病灶已切除，硬膜外积液（箭头所示） 5c 横断面增强T_1WI显示，L_{4-5}椎间隙水平结节状病灶切除术后改变

Figure 4 Pathological examination findings after readmission surgery ×100 The tumor tissue was rich in mucus, and the tumor cells in the mucus-like interstitial were arranged in a strip-like or nest-like arrangement, which was cuboidal, and some cytoplasm was mildly eosinophilic; some tumor cells were droplet-like and visible in the cytoplasm. A large amount of mucus pushes the nucleus shift in the cytoplasm, and the cells were mild to moderately shaped (Panel 4a). HE staining The EMA was positive in nucleus and cytoplasm (Panel 4b). Immunohistochemical staining (EnVision) **Figure 5** Lumbar spine MRI findings of one-week after operation Sagittal T_2WI showed epidural effusion in the operation area (arrow indicates, Panel 5a). Sagittal enhanced T_1WI showed the L_{4-5} localized nodular lesions resected, and epidural effusion (arrow indicates, Panel 5b). Axial enhanced T_1WI revealed changes after resection of localized nodular lesions in L_{4-5} intervertebral space level (Panel 5c).

骨，尤以颅底斜坡和骶骨区最为常见，骶尾区、颅底和脊柱其他部位所占比例分别为50%、35%和15%，较少发生转移[1-5]。临床症状主要表现为头痛和外展神经麻痹导致的复视[6]。脊椎MRI可辅助评估肿瘤大小，T_2WI呈不同程度高信号，增强扫描后病灶呈不均匀强化；CT所显示的特征性环形钙化可能与钙化的组织学类型相关[7]，明确诊断仍依靠组织活检术。

本文患者肿瘤首发于鞍区，经蝶窦鞍区占位性病变切除术后出现脑脊液鼻漏，术后短期内出现下肢神经功能障碍，考虑为肿瘤细胞随脑脊液播散所致，疾病迅速进展，再次行椎管内肿瘤切除术，术中尽量维持硬脑膜完整，避免发生脑脊液鼻漏，必要时可进行鞍底重建。因此，对于术前即存在脊髓功能障碍的患者，应进行脊椎MRI检查以筛查远处转移灶[8-10]。

脊索瘤首选手术治疗，既可明确诊断，又可减轻肿瘤负荷[11-12]。对于发生于颅底的脊索瘤和软骨肉瘤，最理想的是手术全切除，然而由于解剖结构对手术入路的限制且肿瘤邻近重要脑结构，难以达到手术全切除，术后需辅助放射治疗[13-14]，但远期疗效欠佳，且存在神经放射性损伤的风险，为再次手术治疗增加难度[15-16]。质子放射治疗适用于术后神经血管重要区域的残留肿瘤，由于能够集中能量释

放并对周围组织无损伤，可实现针对病灶的精准治疗，但目前尚缺乏大样本脊索瘤病例的临床研究报道。颅内脊索瘤椎管内播散十分罕见，但当出现脊髓和神经根症状时，应及时进行脊椎MRI检查以评估是否发生转移。

利益冲突 无

参 考 文 献

[1] Heffelfinger MJ, Dahlin DC, MacCarty CS, Beabout JW. Chordomas and cartilaginous tumors at the skull base[J]. Cancer, 1973, 32:410-420.

[2] Perzin KH, Pushparaj N. Nonepithelial tumors of the nasal cavity, paranasal sinuses, and nasopharynx: a clinicopathologic study. Ⅳ: chordomas[J]. Cancer, 1986, 57:784-796.

[3] Harbour JW, Lawton MT, Criscuolo GR, Holliday MJ, Mattox DE, Long DM. Clivus chordoma: a report of 12 recent cases and review of the literature[J]. Skull Base Surg, 1991, 1:200-206.

[4] Volpe R, Mazabraud A. A clinicopathologic review of 25 cases of chordoma (a pleomorphic and metastasizing neoplasm)[J]. Am J Surg Pathol, 1983, 7:161-170.

[5] Volpe NJ, Liebsch NJ, Munzenrider JE, Lessell S. Neuro-ophthalmologic findings in chordoma and chondrosarcoma of the skull base[J]. Am J Ophthalmol, 1993, 115:97-104.

[6] Leong JL, Batra PS, Citardi MJ. CT-MR image fusion for the management of skull base lesions[J]. Otolaryngol Head Neck Surg, 2006, 134:868-876.

[7] Rassekh CH, Nuss DW, Kapadia SB, Curtin HD, Weissman JL, Janecka IP. Chondrosarcoma of the nasal septum: skull base imaging and clinicopathologic correlation[J]. Otolaryngol Head Neck Surg, 1996, 115:29-37.

[8] Fernandez-Miranda JC, Gardner PA, Snyderman CH, Devaney KO, Mendenhall WM, Suárez C, Rinaldo A, Ferlito A. Clival chordomas: a pathological, surgical, and radiotherapeutic review[J]. Head Neck, 2014, 36:892-906.

[9] Yamaguchi T, Imada H, Iida S, Szuhai K. Notochordal tumors: an update on molecular pathology with therapeutic implications[J]. Surg Pathol Clin, 2017, 10:637-656.

[10] Sakakibara Y, Taguchi Y, Nakamura H, Onodera H, Wakui D, Ikeda T, Aida Y. A case of primary intrasellar chondroid chordoma[J]. No Shinkei Geka, 2019, 47:901-907.

[11] Liu S, Zhou X, Song A, Huo Z, Yao S, Wang Y, Liu Y. Surgical treatment of giant chordoma in the thoracic spine combining thoracoscopic and posterior spinal surgery: a case report[J]. Medicine (Baltimore), 2019, 98:E16990.

[12] Abuzayed B, Alawneh K, Al Qawasmeh M, Raffee L. Clivus intraosseous meningioma mimicking chordoma[J]. J Craniofac Surg, 2019.[Epub ahead of print]

[13] Li L, Wang K, Ma X, Liu Z, Wang S, Du J, Tian K, Zhou X, Wei W, Sun K, Lin Y, Wu Z, Tian J. Radiomic analysis of multiparametric magnetic resonance imaging for differentiating skull base chordoma and chondrosarcoma[J]. Eur J Radiol, 2019, 118:81-87.

[14] Gutierrez A, Rompokos V, Li K, Gillies C, D'Souza D, Solda F, Fersht N, Chang YC, Royle G, Amos RA, Underwood T. The impact of proton LET/RBE modeling and robustness analysis on base-of-skull and pediatric craniopharyngioma proton plans relative to VMAT[J]. Acta Oncol, 2019, 20:1-10.

[15] Chetan MR, Lyon PC, Wu F, Phillips R, Cranston D, Gillies MJ, Bojanic S. Role of diffusion-weighted imaging in monitoring treatment response following high-intensity focused ultrasound ablation of recurrent sacral chordoma[J]. Radiol Case Rep, 2019, 14:1197-1201.

[16] Usher I, Flanagan AM, Choi D. Systematic review of clinical, radiologic, and histologic features of benign notochordal cell tumors: implications for patient management[J]. World Neurosurg, 2019, 130:13-23.

（收稿日期：2019-11-07）

55th Annual Congress of Canadian Neurological Sciences Federation 2020

Time: June 7–10, 2020

Venue: Banff, Alberta, Canada

Website: http://congress.cnsfederation.org/

The 55th Canadian Neurological Sciences Federation (CNSF) Congress 2020 will take place in Banff, Alberta, Canada on June 7–10, 2020. This is a collegial meeting providing multidisciplinary courses relevant to all neuroscience specialties.

By the end of the Congress, delegates will be able to discuss advances in the management of acute and chronic neurological and neurosurgical disorders and their imaging appearances, discuss new findings in neurological and neurosurgical disorders and the role of neuroimaging in diagnosis and management, describe advances in neurological, neurosurgical and/or and neuroimaging care and techniques; and identify areas where there are gaps in learning (unperceived needs) not realized before attending the Congress and extend this professional learning after the Congress to the enhanced care of patients.