

复发性上段颈椎脊索瘤一例

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【关键词】 脊索瘤； 颈椎； 复发； 神经外科手术； 病例报告

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Recurrent upper cervical chordoma: one case report

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患者 男性, 58 岁, 主因颈部疼痛 3 个月, 于 2009 年 2 月 10 日入院。患者 3 个月前无明显诱因出现颈部疼痛, 以左后侧显著, 呈持续性钝痛, 转头时加重、休息后缓解, 同时伴转头活动受限, 于当地医院行颈椎 CT 和 MRI 检查证实上段颈椎占位性病变(图 1)。遂行病变组织穿刺活检术, 术后病理学检查证实为脊索瘤。为求进一步手术治疗至我院就诊。患者自发病以来精神、睡眠好, 饮食正常, 大小便正常, 体重无明显变化。既往史、个人史和家族史均无特殊。入院后体格检查: 神志清楚, 语言流利, 脑神经检查未见明显异常, 颈部强迫体位, 上肢外形和活动度无异常, 四肢肌力和肌张力正常、腱反射正常、位置觉和痛温觉正常。左侧下颌角触及皮下肿物, 圆形、质地较硬、不能活动、触之无疼痛。数字减影血管造影术(DSA)和球囊闭塞试验(BOT)显示, 左侧椎动脉被肿瘤推向外侧, 管径通畅, 右侧椎动脉代偿良好(图 2)。遂行颈部肿瘤全切除术。患者右侧卧位, 头架固定, 气管插管全身麻醉, 采取侧方入路, 手术切口自左侧乳突沿胸锁乳突肌后缘斜向下至 C₅ 水平, 长度约 10 cm, 逐层切开皮肤和皮下组织, 分离胸锁乳突肌和头夹肌, 显露肿瘤包膜, 肿瘤体积大小约 6.00 cm × 4.50 cm × 4.20 cm, 呈灰白色、质地柔软、血运中等, 肿瘤破坏枢椎(C₂)椎体和横突, 于手术显微镜下包膜内分块

全切除肿瘤, 术中左侧椎动脉破裂, 予电凝切断后结扎; 然后植入左侧寰椎(C₁)侧块螺钉和 C₃ 椎弓根螺钉各 1 枚, 钛棒连接, 电钻磨薄 C₁ 后弓和 C₃ 椎板, 于 C₁ 后弓、C₃ 椎板、钛棒和螺钉周围植入人工骨(山西奥瑞生物材料有限公司); 留置引流管, 术后 24 h 拔除; 逐层关闭手术切口, 行间断缝合。术后临床症状消失, 恢复良好, 无手术相关并发症。患者共住院 14 d, 出院时自觉颈部疼痛明显缓解, 活动自如, 四肢肌力同术前。术后 6 个月随访时复查颈椎 CT 显示螺钉位置和植骨生长良好, 肿瘤无复发迹象(图 3)。随访至术后 3 年(2012 年 8 月)患者出现颈部疼痛伴吞咽不适, 复查颈椎 MRI 显示肿瘤复发, 咽后壁受累, 脊髓受压(图 4)。患者拒绝再次手术, 予伊马替尼(imatinib)400 mg/d 口服, 共治疗 2 个月, 同时将 ¹²⁵I 粒子植入肿瘤内行放射治疗(于外院植入 ¹²⁵I 粒子 2 次, 具体剂量不详), 带瘤生存, 生活可自理。随访至术后 6 年(2015 年 12 月), 患者因呼吸困难合并肺部感染死亡。

讨 论

脊索瘤是临床少见的骨组织恶性肿瘤, 源自残留的胚胎脊索组织, 好发于颅底、脊柱和骶尾部等中轴骨。肿瘤恶性程度相对较低, 生长缓慢, 但常侵及周围组织, 且发生部位较为特殊, 故手术全切除较为困难, 而传统放射治疗和药物化疗效果不佳, 具有极高的复发率^[1]。有文献报道, 患者 5 年生存率为 50% ~ 60%^[2]。发生于颈椎的脊索瘤相对少见, 约占脊柱脊索瘤的 10%。由于肿瘤周围毗邻气管、食道和颈内动脉等重要结构, 肿瘤压迫可引起

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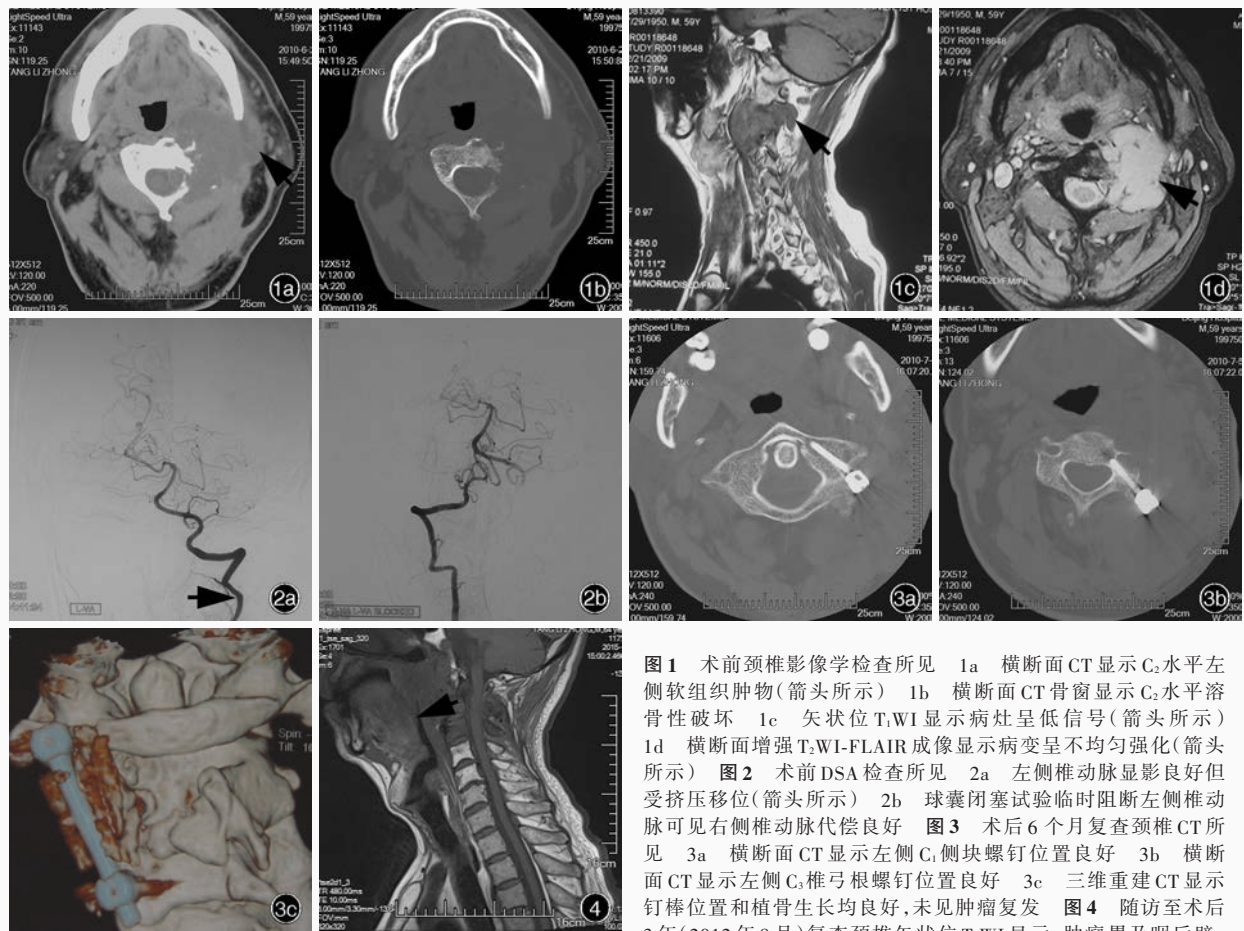


图 1 术前颈椎影像学检查所见 1a 横断面 CT 显示 C₂ 水平左侧软组织肿物(箭头所示) 1b 横断面 CT 骨窗显示 C₂ 水平溶骨性破坏 1c 矢状位 T₁WI 显示病灶呈低信号(箭头所示) 1d 横断面增强 T₂WI-FLAIR 成像显示病变呈不均匀强化(箭头所示) 图 2 术前 DSA 检查所见 2a 左侧椎动脉显影良好但受挤压移位(箭头所示) 2b 球囊闭塞试验临时阻断左侧椎动脉可见右侧椎动脉代偿良好 图 3 术后 6 个月复查颈椎 CT 所见 3a 横断面 CT 显示左侧 C₁ 侧块螺钉位置良好 3b 横断面 CT 显示左侧 C₂ 椎弓根螺钉位置良好 3c 三维重建 CT 显示钉棒位置和植骨生长均良好, 未见肿瘤复发 图 4 随访至术后 3 年(2012 年 8 月)复查颈椎矢状位 T₁WI 显示, 肿瘤累及咽后壁,

口咽部明显受压(箭头所示)

Figure 1 Preoperative cervical imaging findings Axial CT showed soft tissue mass on the left side of C₂ (arrow indicates, Panel 1a). Bone window of axial CT showed osteolytic lesion at C₂ level (Panel 1b). Sagittal T₁WI showed hypo-intensity of the lesion (arrow indicates, Panel 1c). Axial enhanced fat-suppressed T₂WI-FLAIR showed heterogeneous enhancement (arrow indicates, Panel 1d). **Figure 2** Preoperative DSA findings Left vertebral artery was open but pushed outward (arrow indicates, Panel 2a). Right vertebral artery was patent after the left vertebral artery balloon occlusion test (Panel 2b). **Figure 3** Cervical CT findings 6 months after operation Axial CT showed the position of C₁ lateral mass screw on the left side was good (Panel 3a). Axial CT showed the position of C₂ pedicle screw on the left side was good (Panel 3b). Three-dimensional reconstructed CT showed good position of screws and growth of bone graft. No tumor recurrence was revealed (Panel 3c). **Figure 4** In the follow-up 3 years after operation (August 2012), cervical T₁WI showed recurrent tumor at the retropharyngeal space, and oropharynx was compressed (arrow indicates).

颈部疼痛、咽喉部异物感、吞咽困难等, 肿瘤压迫脊髓可出现神经功能缺损^[3]。

CT 和 MRI 是目前诊断脊索瘤的主要手段。CT 表现为脊柱溶骨性破坏, 周围软组织占位性病变。MRI 呈 T₁WI 低信号、T₂WI 高信号, 病变呈分叶状, 其内可见不规则分隔, 边界清楚; 增强扫描呈轻至中度不均匀强化。典型脊索瘤可呈现“蜂房征”, 反映脊索瘤内较多黏液样基质和肿瘤细胞空泡样改变。

对于任何部位的脊索瘤, 目前较为公认的治疗方法是整体综合治疗(包括手术和术后放射治疗联合药物化疗)^[1,4-5]。手术根治性切除仍在综合治疗中占首要地位, 首次手术切除程度决定肿瘤复发速

度和患者生存期。Sundaresan 等^[6]回顾分析 54 例脊柱脊索瘤患者的临床资料发现, 约 90% 行部分切除术的患者术后 2 年内肿瘤复发。由于颈椎解剖学结构特殊, 肿瘤全切除常存在较大技术挑战, 如椎动脉显露、神经根保护、脊柱稳定性重建等。由于肿瘤对椎体的溶骨性破坏, 需采用钉棒系统进行颈椎稳定性重建^[3,4]。该例患者肿瘤挤压椎动脉, 使其失去正常解剖学结构而于术中误伤, 因此, 术前采用 DSA 评价椎动脉走行或代偿情况对手术成功有重大帮助。若肿瘤全切除困难, 手术目的则是解除肿瘤对脊髓和重要结构的压迫、缩小肿瘤体积, 为后续放射治疗奠定基础; 对于无法或不能耐受手术的

患者,可于病变组织活检术后直接行放射治疗。

一般认为,脊索瘤对传统放射治疗不敏感,最新研究显示,相对于传统的光子放射治疗,碳离子或质子放射治疗可将较大放射剂量控制在肿瘤局部,从而很好地控制局部复发,延长患者总生存期,并通过将放射剂量分割的方法以减少放射治疗的毒性反应^[7-9]。但是,由于此种放射治疗方式费用昂贵,目前在我国难以全面开展。

迄今尚无化疗药物获批准用于复发进展后的脊索瘤。目前临床应用的药物多为临床试验阶段或尝试性应用,如伊马替尼、拉帕替尼(lapatinib)等,但前期临床试验显示,分子靶向治疗在延缓脊索瘤生长方面取得了令人振奋的成果,有望成为治疗脊索瘤的有效方法^[1,10]。

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Neuropsychological Formulation: A Clinical Casebook published

Neuropsychological Formulation: A Clinical Casebook (ISBN: 978-3-319-18337-4, eBook ISBN: 978-3-319-18338-1) was published by Springer International Publishing. The editor of this book is Jamie A.B. Macniven.

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