

## ·椎管内肿瘤神经外科临床研究·

## 颅颈交界区神经鞘瘤的外科手术治疗

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**【摘要】目的** 探讨颅颈交界区神经鞘瘤的临床特点及外科手术治疗原则。**方法** 共29例颅颈交界区神经鞘瘤患者经后正中入路行肿瘤切除术,术前影像学观察肿瘤未影响脊柱稳定性者仅行肿瘤切除术;肿瘤侵蚀破坏骨质、造成脊柱失稳者同期行枕颈融合内固定术。**结果** 29例患者均达显微镜下肿瘤全切除,术后病理证实为神经鞘瘤。术后枕颈部疼痛症状明显缓解,其中13例枕颈部麻木感者术后3个月随访时症状完全消失;无一例肢体麻木、肌无力症状加重,无新发神经功能缺损、脑脊液漏和死亡病例。术后均获3~24个月随访,未见肿瘤复发及脊柱畸形和椎体滑脱。**结论** 颅颈交界区神经鞘瘤采取经后正中入路即可全切除肿瘤;对于肿瘤侵蚀破坏骨质而影响脊柱稳定性的患者,术中需辅助枕颈融合内固定术。

**【关键词】** 神经鞘瘤; 脊髓; 寰枢关节; 内固定术(非MeSH词); 外科手术

### Surgical management of schwannomas in the craniocervical junction region

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**【Abstract】Objective** To investigate the clinical characteristics and surgical management of schwannomas in the craniocervical junction region. **Methods** Clinical data of 29 patients with craniocervical schwannomas was retrospectively studied. According to MRI examination, tumors in 3 cases were located in anterolateral region of the spinal cord and tumors in 26 patients were located in posterolateral side of the spinal cord. Among the 29 patients, 4 patients had intra- and extra-spinal schwannomas. All the tumors were excised through posterior approach. The extent of bone window was to the lateral side of the spinal cord. If spinal stability was destroyed by osteotomy intraoperatively or by bone erosion of the tumor preoperatively, internal fixation of occipitocervical fusion was performed after tumor excision. **Results** Total resection was achieved in all of the 29 patients, and postoperative pathological result was schwannoma. The pain of the back of neck and occipital region was relieved after operation and numbness of neck and occipital region in 13 patients disappeared in the follow-up of 3 months. There was no new neurological defect, no cerebrospinal fluid fistula and no dead patient. During the follow-up period of 3–24 months, there was no tumor recurrence and no spinal spondylolisthesis or deformity. **Conclusion** Total resection of tumor can be achieved by posterior approach for schwannomas in the craniocervical region. If spinal stability was destroyed by the tumor, internal fixation will be performed.

**【Key words】** Neurilemmoma; Spinal cord; Atlanto-axial joint; Internal fixation (not in MeSH); Surgical procedures, operative

颅颈交界区神经鞘瘤约占椎管内神经鞘瘤的5%<sup>[1]</sup>,因其毗邻延髓、颈髓上段及椎动脉等重要解剖结构,手术切除一直是神经外科一大难题。笔者对首都医科大学宣武医院神经外科2009–2012年诊

断与治疗的颅颈交界区神经鞘瘤患者的临床资料进行总结,分析其临床特点及手术治疗原则,以为临床提供参考。

### 资料与方法

#### 一、一般资料

共29例患者,男性15例,女性14例;年龄30~68岁,平均46岁;病程1周至12年,平均37.42个月。临床主要表现为枕颈部疼痛(23例)、肢体无力

doi:10.3969/j.issn.1672-6731.2013.11.005

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(20例)或麻木(14例)、头晕和吞咽困难(各1例)。患者入院后均行MRI检查,以明确肿瘤累及范围及其与周围组织关系,肿瘤灶一般呈等T<sub>1</sub>、T<sub>2</sub>信号,增强后呈明显均匀强化,部分患者因肿瘤灶内出血、坏死、囊性变而呈不均匀强化;根据肿瘤与脊髓之间的关系,肿瘤位于脊髓侧前方者3例、脊髓侧后方者26例,其中4例肿瘤经椎间孔沟通椎管内外(图1a)。有23例患者行CT检查,观察肿瘤是否侵蚀颅颈交界区骨质,同时经三维CT对需接受枕颈融合治疗的患者行螺钉植入可行性分析。本组有6例患者因肿瘤累及椎管外而行CTA检查,以明确肿瘤与椎动脉之间的关系(图1b~1d)。

## 二、手术方法

患者全身麻醉、俯卧位,Mayfield头架固定头部,取后正中直切口,逐层分离皮下组织,骨膜下分离脊柱两侧椎旁肌,咬除寰椎(C<sub>1</sub>)后弓、部分枢椎(C<sub>2</sub>)棘突、椎板上缘和枕大孔后缘。由于神经鞘瘤好发于脊髓侧后方,故骨窗向侧方咬除范围显露脊髓侧方即可,仅有少数位于脊髓腹侧,鲜见位于脊髓正前方者,且肿瘤与脊髓之间多无粘连,故术前根据肿瘤生长偏向某一侧咬除骨窗,最大程度显露脊髓侧方,且以不破坏侧方小关节稳定性为宜;术中手术床向对侧倾斜,结合显微镜角度,也可使脊髓腹侧神经鞘瘤达全切除。骨窗形成后纵行剪开硬脊膜,带线针悬吊于两旁、纵行剪开软脊膜,同时银夹固定双侧硬脊膜,手术显微镜下仔细分离肿瘤与脊髓和周围神经根的粘连。囊性肿瘤可先行囊腔穿刺,然后整块切除;对于小的实质性肿瘤可直接整块切除;大的实质性肿瘤可先行瘤内减压,待其体

积缩小后再整块切除残留肿瘤。肿瘤切除后彻底止血,严密缝合硬脊膜,逐层缝合肌肉、皮肤。本组有1例寰椎神经根神经鞘瘤患者,肿瘤呈“哑铃”形沟通椎管内外,由于肿瘤侵蚀破坏寰枕关节,肿瘤切除后出现寰枕关节不稳,术中同时行枕颈融合内固定术,缝合硬脊膜后植入枕骨板和枢椎椎弓根螺钉,截取长度适宜钛棒,塑形后植入钉尾,取螺帽在枕颈部处于中立位时加压固定。将咬除的椎板和枕骨制成直径约0.30 cm的骨颗粒,植于钉棒周围。其余患者因肿瘤未影响枕颈部稳定性,故未行枕颈融合内固定术(<http://www.cjcn.org/index.php/cjcn/pages/view/v13n11a5>)。

## 结 果

本组29例患者均达显微镜下肿瘤全切除,术后病理检查证实为神经鞘瘤(图2,3)。所有患者术后枕颈部疼痛症状均明显缓解,其中13例枕颈部麻木感者术后3个月随访时已完全消失;无一例术后肢体麻木、肌无力症状加重,且随访期间症状明显改善;术前表现为头晕和吞咽困难的患者术后症状仅轻微减轻,术后3个月时症状完全好转。本组无一例患者术后新发神经功能缺损和脑脊液漏,无死亡病例。所有患者均获3~24个月随访,复查头颈部MRI未发现肿瘤复发、脊柱畸形或椎体滑脱。

## 讨 论

颅颈交界区系指下斜坡、枕骨大孔及枢椎以上区域<sup>[2-3]</sup>,该区域肿瘤临床较为少见,常见肿瘤包括

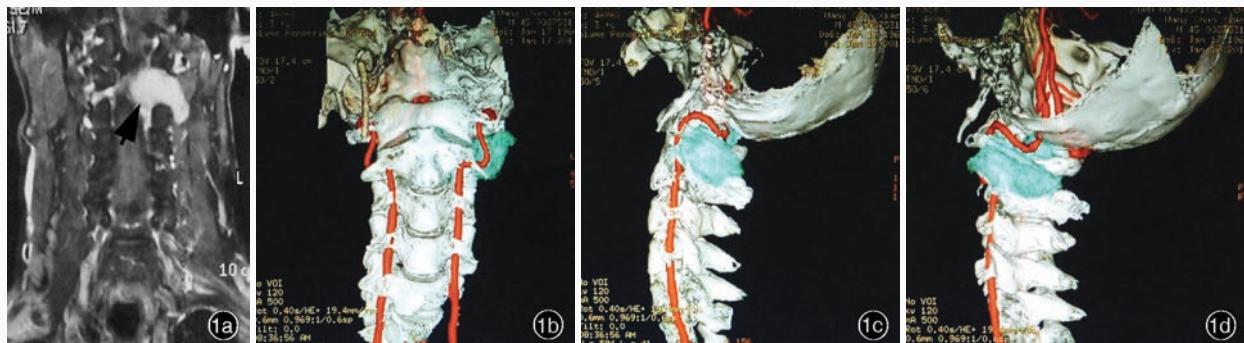
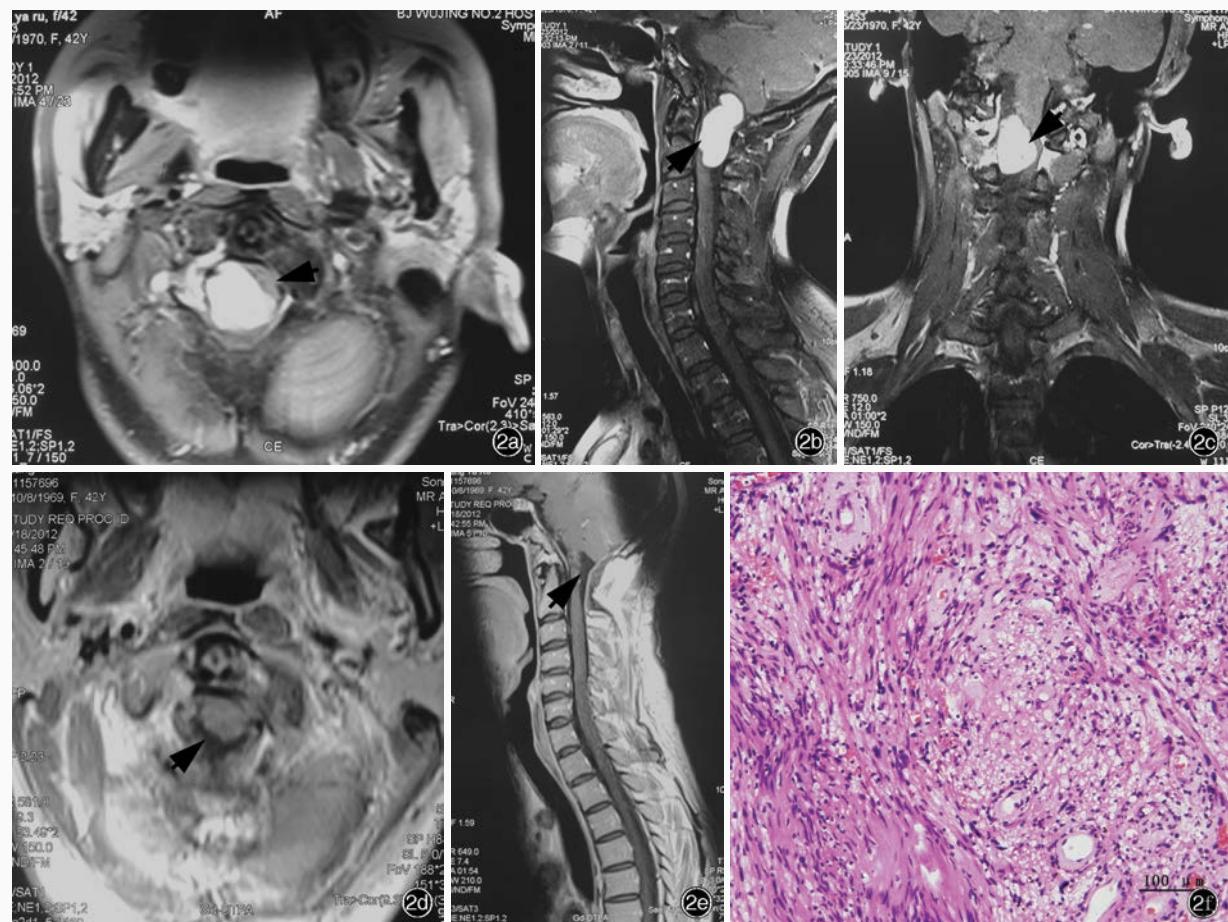


图1 颅颈交界区神经鞘瘤影像学检查所见 1a 冠状位增强T<sub>1</sub>WI扫描显示肿瘤沟通椎管内外(箭头所示) 1b~1d 正位、侧位和后斜位CTA显示,椎动脉位于肿瘤前方,未被肿瘤包裹

**Figure 1** Imaging examination findings of schwannomas in the craniocervical region. Coronal enhanced T<sub>1</sub>WI showed the tumor located intra- and extra-spine (arrow indicates, Panel 1a). Cervical CTA demonstrated that the vertebral artery was located in front of the tumor and was not covered by tumor (Panel 1b~1d).



**图2** 女性患者,42岁。主因四肢麻木、无力5个月,加重1个月入院 2a~2c 术前横断面、矢状位和冠状位增强T<sub>1</sub>WI扫描显示,肿瘤位于颅颈交界区胸椎椎管内脊髓右后方,压迫脊髓(箭头所示) 2d,e 术后横断面和矢状位增强T<sub>1</sub>WI扫描显示肿瘤全切除(箭头所示) 2f 术后病理诊断为神经鞘瘤 HE染色 低倍放大

**Figure 2** A 42-year-old female patient suffered from numbness and weakness of upper and lower extremities for 5 months, and the symptoms aggravated for 1 month. Preoperative axial, sagittal and coronal enhanced T<sub>1</sub>WI showed the tumor was located in the right backward of craniocervical thoracic spinal canal and compressed the spinal cord (arrows indicate, Panel 2a~2c). Postoperative axial and sagittal enhanced T<sub>1</sub>WI showed the tumor was totally removed (arrows indicate; Panel 2d, e). Postoperative pathological diagnosis was schwannoma (Panel 2f). HE staining low power magnified

脊膜瘤、神经鞘瘤、脊索瘤、神经节细胞瘤及骨、软骨肉瘤等。神经鞘瘤为常见椎管内肿瘤,约占椎管内肿瘤的30%<sup>[4]</sup>,大部分起源于脊神经的感觉神经根<sup>[5]</sup>,少部分起源于运动神经根<sup>[6]</sup>。尽管神经鞘瘤为临床常见椎管内肿瘤,但寰枢椎(C<sub>1~2</sub>)神经根神经鞘瘤发生率相对较低,约占所有椎管内神经鞘瘤的5.30%、占颈椎椎管内神经鞘瘤的18%<sup>[2]</sup>。颅颈交界区神经鞘瘤除寰枢椎神经根神经鞘瘤外,还包括起源于舌下神经和颈静脉孔的神经鞘瘤。舌下神经神经鞘瘤最早由De Martel等在1933年报告,既可位于颅内亦可位于颅内外,甚至可完全位于颅外。起源于颈静脉孔的神经鞘瘤占颅内神经鞘瘤的2.89%~4.00%<sup>[7~8]</sup>,其中多数起源于舌咽神经<sup>[9]</sup>。

神经鞘瘤的临床表现根据其累及范围的不同而有所不同,疼痛是最主要的临床表现,以头痛和颈部疼痛常见;此外,根据肿瘤累及范围还可以出现肢体麻木、肌无力、共济失调、声音嘶哑、吞咽困难和听力减弱等症状与体征。

神经鞘瘤多为良性肿瘤,手术全切除即可治愈。关于颅颈交界区神经鞘瘤的手术入路,文献报道较多,如前方经口入路、侧方入路及后正中入路。经口入路适用于肿瘤位于脊髓前方者,尤其是肿瘤基底部位于脊髓前方的脊膜瘤,可先处理肿瘤血供,但经口入路要求手术医师熟悉口咽部解剖、口腔外科及耳鼻咽喉科的相关手术技巧,与此同时,经口入路存在手术入路深、视野狭小及术后感

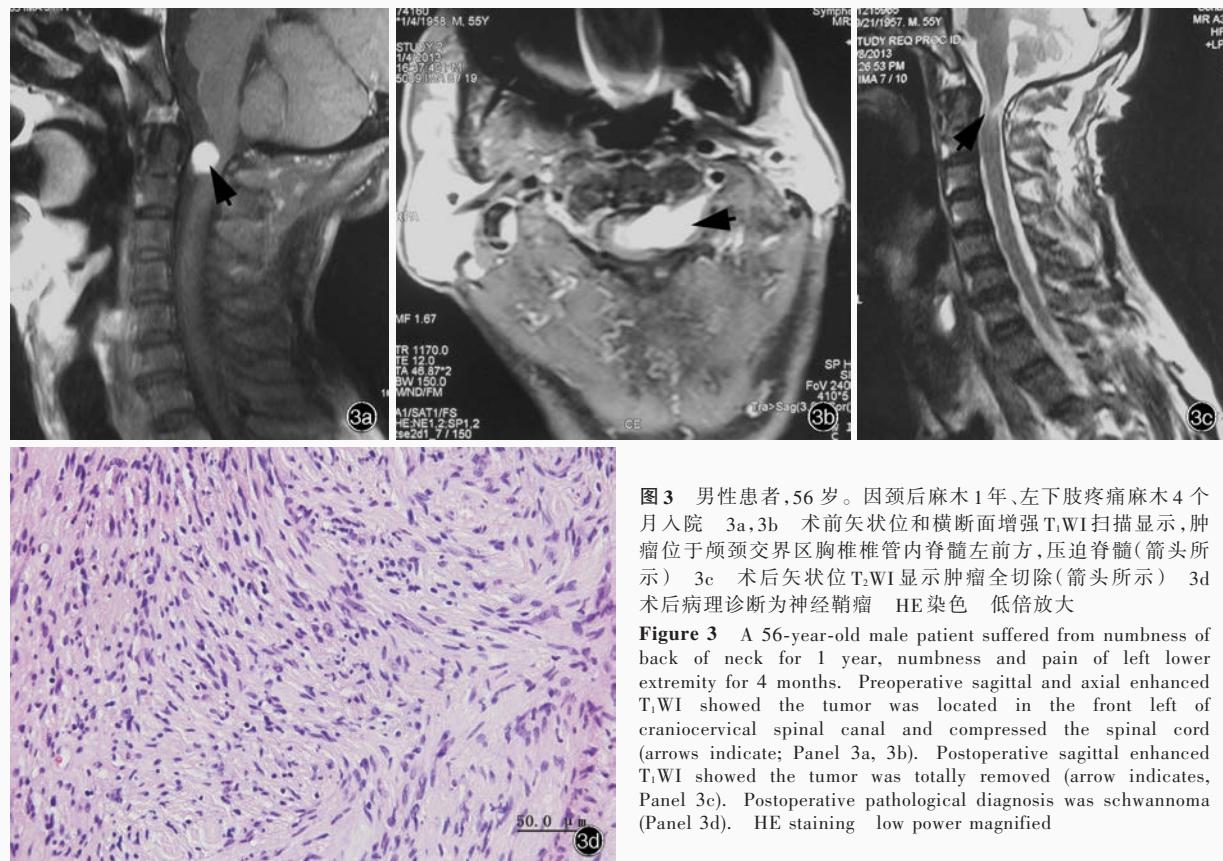


图3 男性患者,56岁。因颈后麻木1年、左下肢疼痛麻木4个月入院 3a,3b 术前矢状位和横断面增强T<sub>1</sub>WI扫描显示,肿瘤位于颅颈交界区胸椎椎管内脊髓左前方,压迫脊髓(箭头所示) 3c 术后矢状位T<sub>2</sub>WI显示肿瘤全切除(箭头所示) 3d 术后病理诊断为神经鞘瘤 HE染色 低倍放大

**Figure 3** A 56-year-old male patient suffered from numbness of back of neck for 1 year, numbness and pain of left lower extremity for 4 months. Preoperative sagittal and axial enhanced T<sub>1</sub>WI showed the tumor was located in the front left of craniocervical spinal canal and compressed the spinal cord (arrows indicate; Panel 3a, 3b). Postoperative sagittal enhanced T<sub>2</sub>WI showed the tumor was totally removed (arrow indicates, Panel 3c). Postoperative pathological diagnosis was schwannoma (Panel 3d). HE staining low power magnified

染率高等缺点<sup>[10]</sup>。侧方入路可根据病变累及部位,分为经颞入路、经岩骨入路、经乳突后入路、侧方入路、远外侧入路和极外侧入路,以及经枕骨髁、髁上及髁旁等亚型。上述手术入路有些仅是名称不同,有些可能仅是局部细微的改变,目前临床应用较广泛的人路为侧方入路、远外侧入路和极外侧入路,适用于处理脑干及颈髓上段前方或前外侧病变,但手术操作复杂,手术时间长,创伤大,术后易出现切口不愈合、脑脊液漏等并发症。对于颅颈交界区神经鞘瘤,我们认为采取单纯经颈椎后正中入路更为适宜,因为神经鞘瘤多位于脊髓侧后方,即使少数位于脊髓前方,由于神经鞘瘤与脊髓无明显界面,因此采取单纯后正中入路即可全切除肿瘤。而且相关研究显示,经后正中入路、侧方入路或极外侧入路切除颅颈交界区腹侧或侧方肿瘤,其肿瘤全切除率无明显差异<sup>[11-12]</sup>。

颅颈交界区神经鞘瘤以寰枢椎神经根神经鞘瘤多见,手术过程中若采取单纯经颈椎后正中入路,仅需咬除寰椎后弓,与正常的颅后窝减压时寰椎后弓咬除范围相当,对脊柱稳定性无明显影响,

一般术中无需行内固定,术后佩戴颈托外固定即可。但是对于肿瘤侵蚀颅颈交界区骨质者,或手术过程中切除肿瘤及其侵蚀骨质影响颅颈交界区稳定性者,术中应同时行内固定术。如颈静脉孔内神经鞘瘤,肿瘤增大破坏一侧枕髁结构,肿瘤切除后失去肿瘤的占位效应可出现颅颈交界区骨折,导致突发性呼吸骤停等严重后果。目前,常用的枕颈融合内固定技术主要为枕骨板与枢椎椎弓根螺钉固定,椎弓根螺钉内固定系统因其能有效达到脊柱“三柱”固定,符合脊柱固定的生物力学要求<sup>[13]</sup>。但是,如果枢椎椎弓根被破坏或因先天性发育不良而致枢椎椎弓根较细,无法植人椎弓根螺钉时,则需向下延续至C<sub>3</sub>或C<sub>4</sub>进行固定,但中段颈椎由于椎弓根细小,螺钉植人风险较大,我们建议选择侧块螺钉进行固定,这也是目前已经取得临床共识的内固定技术<sup>[14]</sup>。

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(收稿日期:2013-10-09)

## · 读者·作者·编者 ·

### 《中国现代神经疾病杂志》编辑部关于稿件统计分析方法的要求

《中国现代神经疾病杂志》编辑部对来稿中的统计分析方法一律要求明确研究设计方法,以及详细描述资料性质和结果,具体要求如下:

1. 研究设计方法 要求交代研究设计的名称和主要方法。如调查设计应写明是前瞻性、回顾性还是横断面调查研究;实验设计应写明具体设计类型,如自身配对设计、成组设计、交叉设计、析因设计或正交叉设计等;临床试验设计应写明属于第几期临床试验,采用何种盲法措施等。应围绕“重复、随机、对照、均衡”四项基本原则进行概要说明,尤其要说明如何控制重要的非试验因素的干扰和影响。

2. 资料及结果的表达与描述 采用均数±标准差( $\bar{x} \pm s$ )表示近似服从正态分布的定量资料,采用中位数和四分位数间距 [ $M(P_{25}, P_{75})$ ]表示呈偏态分布的定量资料;采用相对数构成比(%)或率(%)表示计数资料,用相对数构成比时分母不能小于20。应写明所用统计分析方法的具体名称、统计量具体值,应尽可能给出确切的P值;当涉及总体参数时,在给出显著性检验结果的同时,给出95%可信区间。

### 《中国现代神经疾病杂志》编辑部关于稿件图表格式的要求

《中国现代神经疾病杂志》编辑部对来稿中的图表一律以其在正文中出现的先后次序连续编码。每帧图表应冠以图(表)题,并配以英文图(表)题目。图(表)内容均采用中英文对照形式。说明性资料应以中英文对照格式置于图(表)下方注释中。

1. 表格 采用三横线表(顶线、表头线、底线)格式,如遇有合计和统计学处理内容(如t值、P值等),则在此行上面加一条分界横线;应使表中每一列数据的单位相同,有效位数一致。

2. 图片 (1)以计算机制图者应提供单张的原始图片(无箭头、无图号),以图形文件格式(.jpg)Email至编辑部(xdsjjbzz@263.net.cn)。(2)照片图要求有良好的清晰度和对比度,提供单张的原始图片(无箭头、无图号),以图形文件格式(.jpg)Email至编辑部。图中需标注的符号(包括箭头)请另纸标明,并注明图号及图的上下方向。(3)大体标本照片务必在图内有尺度标记。(4)病理图请提供单张的原始图片(无箭头、无图号),大小8 cm×6 cm,分辨率300 dpi,以图形文件格式(.tif)Email至编辑部,并请另纸注明染色方法和放大倍数。