·临床研究·

中轴区原发外周型原始神经外胚层肿瘤 CT 和 MRI 表现

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【摘要】 目的:探讨中轴区原发外周型原始神经外胚层肿瘤(pPNETs)的 CT 和 MRI 表现,以提高对中轴区原发 pPNETs 的影像学认识。**方法**: 回顾性分析 2008 年 10 月至 2014 年 5 月期间经病理证实的 10 例中轴区原发 pPNETs 患者临床资料,其中男 7 例,女 3 例;年龄 8~49 岁,中位年龄 23.6 岁。术前经多排螺旋 CT 平扫 3 例,平扫并增强 4 例;5 例经 MRI 平扫并增强扫描;其中 2 例同时行 CT 及 MRI 检查。**结果**:骨内型 6 例,骨外型 4 例。发病部位及例数分别为骶椎 3 例,腰椎 2 例,颈椎 1 例,颈椎椎管 1 例,尾骨 1 例,右髂骨 1 例,骶前间隙 1 例。横断位最小肿瘤最大层面大小 1.1 cm×1.2 cm,最大者 8.0 cm×9.2 cm,中位大小 4.4 cm×5.7 cm,其中 6 例最大径线>5 cm。其中溶骨性破坏 5 例,2 例伴钙化,混合性 1 例,2 例无骨质破坏。CT 以等密度为主,1 例密度均匀,余 6 例密度不均,其中 3 例可见"浮冰"样改变,增强扫描 1 例中度强化,余 3 例明显强化,2 例内见多发小血管。MRI 在 T1WI 上 5 例均为等信号,在 T2WI 上均为稍高信号,信号不均,增强后 5 例均强化明显。2 例伴椎体压缩性骨折,10 例均未见骨膜反应,5 例 MRI 检查无椎间盘破坏。**结论**:中轴区原发 pPNETs 以儿童及青年多见,肿块常较大。骨内型肿块常包绕椎体,并以椎前间隙为主,均伴椎体骨质破坏,溶骨性常见,椎体原发多见,附件原发或受累少见,可累及椎管,以前壁受累常见,可伴多发新生小血管;骨外型以深部软组织间隙常见,少数原发于椎管,多伴椎体骨质破坏,以溶骨性为主。不侵及椎间盘,椎间隙不窄。CT 可伴"浮冰"征,以骨内型常见。MRI T1WI 以等信号为主,T2WI 以稍长信号为主,增强强化明显。

【关键词】 外周型原始神经外胚层肿瘤;　中轴区;　CT;　MRI
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CT and MRI manifestations of the axial area primary peripheral primitive neuroectodermal tumors　*JIANG Kai, WANG Peng, WANG Liao, YU Zhi-hai, XU Yu, WANG Liang-jiong, TU Can, DENG Sheng-de*, and WANG Jian-hua. *The Affiliated Hospital of Medical College of Ningbo University, Ningbo* 315020, *Zhejiang, China*

ABSTRACT　**Objective**: To explore CT and MRI manifestations of the axial area peripheral primitive neuroectodermal tumors (pPNETs) in order to improve the knowledge of this disease. **Methods**: The clinical data of 10 patients with pPNETs underwent pathologically confirmed were retrospectively analyzed from October 2008 to May 2014. There were 7 males and 3 females, aged from 8 to 49 years old with median of 23.6 years. The preoperative multi-slice spiral CT scan was completed in 3 cases, plain CT scan and enhancement in 4 cases; MRI and enhancement scanning in 5 cases; and among them, 2 cases underwent both MRI and CT scan. **Results**: In-bone type was found 6 cases and out-bone type was found 4 cases. Three cases occurred in sacral vertebrae, 2 cases in lumbar vertebrae, 1 case in cervical vertebrae, 1 case in cervical spinal canal, 1 case in coccyx, 1 case in the right iliac bone, 1 case in presacral space. Cross sectional the smallest tumor maximum level was 1.1 cm×1.2 cm in size, the biggest tumor was 8.0 cm×9.2 cm, the median size was 4.4 cm×5.7 cm, of them, the tumor of maximal diameter larger than 5 cm had 6 cases. Except 2 cases without destruction of bone, the other 5 cases with osteolytic destruction, 2 cases with calcification, 1 case with mixed. Equidensite was main in CT scan, 1 case with uniform density, other 6 cases with uneven density, in which 3 cases with "floating ice" change; 1 case with moderate strengthening, other 3 cases with obviously strengthening, 2 cases with multiple small blood vessels in enhancement scanning. MRI of 5 cases showed the signal of isointensity on T1WI, the slightly high signal on T2WI and the signal was not uniform; after enhancement scan, the signal of 5 cases obviously enhanced. Two patients complicated with vertebral compression fractures, no periosteal reaction was found in all patients, and no the destruction of intervertebral disk was found in 5 patients of MRI scan. **Conclusion**: The axial area pPNETs is common among children and the youth, and the mass often is huge. The mass of in-bone type often envelopes the vertebral body, and main located on prevertebral space, all associated with bone destruction, osteolytic destruction is common, and primary vertebral bodies also is common, attachment primary or involvement is few found, it can involve the spinal canal and an-

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terior wall of spinal canal is common, some cases complicate with multiple newly born small vessels. The mass of out-bone type in deep soft tissue is common, minority primary spinal canal, many complicated with vertebral bone destruction, osteolytic destruction was main. The intervertebral disk was not invaded and intervertebral space has not stenosis. CT scan offer complicate with "floating ice" sign, and in-bone type is common. Isointensity is main on MRI T1WI and slightly longer signal is main on MRI T2WI, strengthening signal is obvious.

KEYWORDS Peripheral primitive neuroectodermal tumor; Axial area; CT; MRI

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外周型原始神经外胚层肿瘤（peripheral primitive neuroectodermal tumors, pPNETs）是起源于神经嵴向神经方向分化的具有相似细胞形态学及细胞基因学特点的恶性小圆细胞肿瘤[1]。pPNETs 分为中枢型和外周型，中枢型占多数[2-3]。pPNETs 又分为骨内型和骨外型，骨内型起源于椎体或附件骨骼，骨外型起源于原因不明的间充质，多见于椎旁及椎管内，多无钙化[4]。中轴区域原发 pPNETs 罕见，其影像学报道较少。本文通过分析 10 例中轴区域原发 pPNETs 的 CT 和 MRI 表现，探讨其影像学表现特征，以提高影像学认识。

1　资料与方法

1.1　一般资料

2008 年 10 月至 2014 年 5 月经穿刺活检及手术病理证实的中轴区原发 pPNETs 患者 10 例，其中男 7 例，女 3 例；年龄 8~49 岁，中位年龄 23.6 岁。临床表现为骶部疼痛 5 例，并 3 例伴软组织肿块进行性增大，3 例为腰部疼痛，2 例为颈部疼痛，并肩部麻木、上肢无力。

1.2　CT 扫描

3 例单行平扫，4 例行平扫及增强扫描。检查应用 PHILIPS Brilliance 16 及 PHILIPS i256 CT 扫描，管电压 120 kV，管电流 210 mAs，层厚 5mm。增强扫描采用高压注射器注射非离子型对比剂（碘海醇）80 ml，流速分别为 4 ml/s，三期扫描：动脉期 30 s，实质期 60 s 及延迟期 90 s。

1.3　MRI 扫描

6 例患者术前应用 GE Signa 1.5T 行平扫及增强检查。采用体表线圈，平扫包括常规矢状位 T1WI、T2WI，抑脂矢状位 T2WI，横断位 T2WI 4 个序列成像，增强经肘静脉注射 Gd-DTPA 15 ml，采用抑脂矢状位、冠状位及横断位 T1WI 3 个序列成像。

2　结果

本组骨内型 6 例，骨外型 4 例。发病部位：骶椎 3 例，腰椎 2 例，颈椎 1 例，颈椎椎管 1 例，尾骨 1 例，右髂骨 1 例，骶前间隙 1 例。横断位最小肿瘤最大层面大小 1.1 cm×1.2 cm，最大者 8.0 cm×9.2 cm，中位大小 4.4 cm×5.7 cm，其中 6 例最大径线大于 5 cm。其中融骨性破坏 5 例，2 例伴钙化，混合性 1 例，余 2 例无骨质破坏。2 例伴椎体压缩性骨折，10 例均无椎间隙狭窄，5 例 MRI 检查无椎间盘破坏。

CT 表现：(1)骨内型：S_3 椎体、C_6 椎体、右侧髂骨翼、S_1-S_2 椎体处等、略低密度的软组织肿块包绕椎体及髂骨，髂骨处软组织肿块以后外侧为主，边界模糊不清，骶尾椎处以椎前为主，部分边界不清，其中髂骨及颈椎、骶椎处软组织肿块巨大，并内见散在的斑片状稍低密度灶，伴髂骨及颈椎、骶尾椎不均匀溶骨性、骨质破坏、骨质硬化（图 1-4），软组织肿块以椎前为主（图 1，2a，4），部分软组织肿块内可见散在的高密度残存骨，这种散在分布于软组织密度中的高密度残存骨似"浮冰"样改变（图 1，2a），右侧髂骨处软组织占位增强后强化明显，内见片状低密度及多发迂曲小血管影（图 3a），右侧臀上动脉受累，VR 重建显示右髂骨耳状面旁不规则骨质破坏（图 3b）；C_6 椎体占位伴椎管受累，椎管前壁处可见软组织密度灶，增强后软组织肿块不均匀强化，强化明显，内见多发迂曲小血管影（图 2a）。(2)骨外型：L_5S_1 左侧椎间孔、S_3 椎体至盆底椎前间隙、L_2 椎体水平椎旁软组织密度占位灶，边界清，相邻骨骨质吸收、椎间孔扩大，骶骨无明显破坏，腰椎椎体骨质破坏伴钙化（图 5-7），其中骶、腰椎处软组织肿块巨大，增强后强化明显，并内见斑片状、片状低密度灶。

MRI 表现：(1)骨内型：L_4、S_1 椎体处巨大软组织占位灶伴椎体骨质破坏、轻度压缩性骨折，部分边界不清，T1WI 为等信号，T2WI 为稍高、高信号，信号稍不均，软组织肿块以椎前部体积相对较大（图 8-9），椎间盘均未见破坏，椎间隙均未狭窄，腰椎病变未累及椎管，骶椎病变累及椎管，位于硬膜外，椎管变窄，增强强化明显，强化不均，内见斑片状、片状低信号区，并骶椎病变可见沿椎管内硬膜外跨节段浸润性生长，上下方均可见脊膜尾征（图 9）。(2)骨外型：L_5S_1 左侧椎间孔、S_3 椎体至盆底椎前间隙、C_2 椎体水平椎管内占位灶（图 10-12），T1WI 为等信号，T2WI 为稍高信号为主，边界清，椎间孔处肿块跨椎管内外生长，颈椎椎管内肿块压迫脊髓，髓内未见水肿；增强后肿块均强化明显，强化程度略不均一。

3　讨论

pPNETs 的发生与染色体[t(11,22)(q24,q12)]

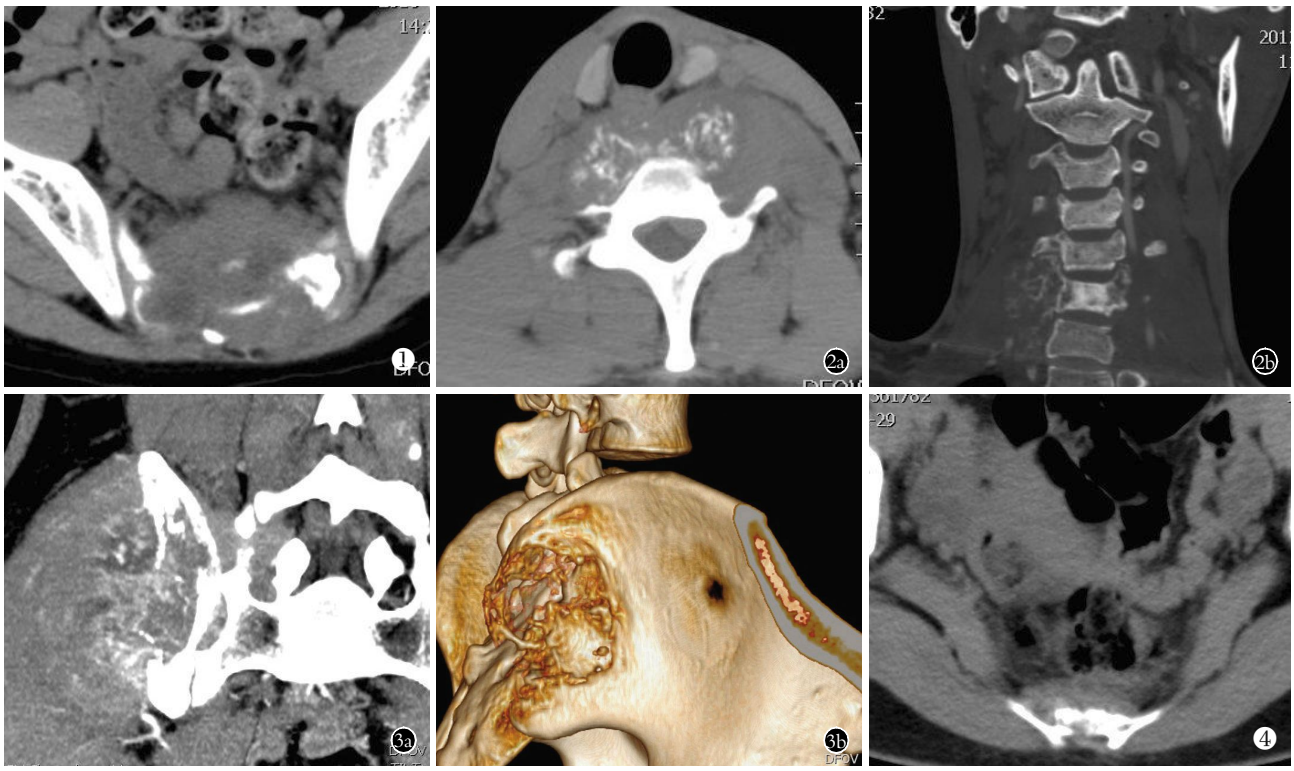

图 1　男性患者，8 岁，骶椎处巨大占位灶伴骶椎骨质破坏，周围见软组织密度包绕，并骶前间隙处肿块较大，内见散在残存骨，可见“浮冰”征　**图 2**　男性患者，22 岁，外周型原始神经外胚层肿瘤　**2a.** C_6 椎体处巨大混杂密度占位灶，椎前间隙处肿块较大，内见条状、片状致密影，可见“浮冰”征，椎管受累，狭窄不明显　**2b.** 冠状位重建示 C_6 椎旁混杂密度灶伴相邻椎体骨质密度增高，呈象牙质样变，椎间隙未狭窄　**图 3**　男性患者，18 岁，外周型原始神经外胚层肿瘤　**3a.** 右侧髂骨近耳状面处臀后缘处巨大软组织占位灶伴髂骨骨质破，边界不清，增强后强化明显，内见多发迂曲小血管　**3b.** VR 重建图像示右侧髂骨耳状面旁广泛骨质破坏缺损，边缘不规则　**图 4**　男性患者，18 岁，骶椎处占位灶伴骨质破坏，骶椎周围见软组织密度包绕，前缘处软组织灶体积相对较大，累及骶管

Fig.1　Male, 8-year-old, enormous occupying lesion complicated with sacral vertebral bone destruction in sacral vertebra, encased the surrounding soft tissue density, and the mass was larger in the presacral space, scattered remaining bone and “floating ice ” sign could be found　**Fig.2**　A 22-year-old male patient with peripheral primitive neuroectodermal tumor　**2a.** C_6 vertebral body huge mixed density accounted for range, prevertebral mass at the gap was larger, and can see the strip, sheet density, “floating ice ” sign, spinal canal was involved in image, spinal stenosis was not obvious　**2b.** The coronal reconstruction showed that the density focus of C_6 and the bone density of adjacent vertebral body increased, and the vertebral body appeared dentin change and there was no stenosis in the intervertebral space　**Fig.3**　An 18-year-old male patient with peripheral primitive neuroectodermal tumor　**3a.** The huge soft tissue tumor was found in the right ilium near to auricular surface hip trailing edge, complicated with iliac bone destruction, boundary was not clear, obvious strengthening after enhancement scanning, could see the multiple tortuous vessels　**3b.** VR reconstructed image showed that the right facies auricularis ossis ilii near to extensive bone was damaged and the edge was irregular　**Fig.4**　An 18-year-old male patient with occupying lesion complicated with bone destruction in sacral vertebra, there was the surrounding soft tissue density encasement around sacral vertebra, in front soft tissue focus there was relatively large, involving sacral canal

易位有关，具有向多方向分化的潜能，故同一瘤体内常可见分化阶段不同的组织[2]。以四肢、骨盆、胸部和腹膜后好发[2,5]。各年龄段均可发病，但儿童及青少年多见，男性略多于女性，但亦有文献报道[6]女性发病率为男性的 3~4 倍。临床表现以迅速增大的肿块伴疼痛及肿块压迫症状常见。pPNETs 罕见，恶性程度高，预后很差[7]，与其他小圆细胞肿瘤相似[8]。本组年龄 8~49 岁，儿童及青少年占 80%(8/10)；男性占 70%(7/10)。

3.1　中轴区域原发 pPNETs 的影像学表现

pPNETs 肿块常较大，多大于 5 cm[9]。本组骨内型 6 例肿块均较大，平均大小约 4.6 cm×6.8 cm；骨外型 4 例，肿块平均大小约 3.7 cm×4.6 cm，其中 2 例肿块巨大，最大径线达 9.2 cm。本组病例肿瘤大小与部位深浅及症状出现早晚有关，部位深、症状出现晚的肿瘤体积常较大，反之亦然，而与影像学分型无明显相关。

骨内型：pPNETs 为高度恶性的小圆细胞肿瘤，常呈浸润性生长，与组织分界不清楚。小圆细胞瘤常表现为软组织肿块较大，而骨质破坏相对轻。中轴区原发 pPNETs 多数起源于髓腔，均伴有骨质破坏，并以椎体溶骨性破坏常见，伴钙化及混合性少见，附件原发或受累少见。肿块常包绕椎体，体积往往较大，并以椎前间隙为主，概与椎前间隙组织结构相对疏

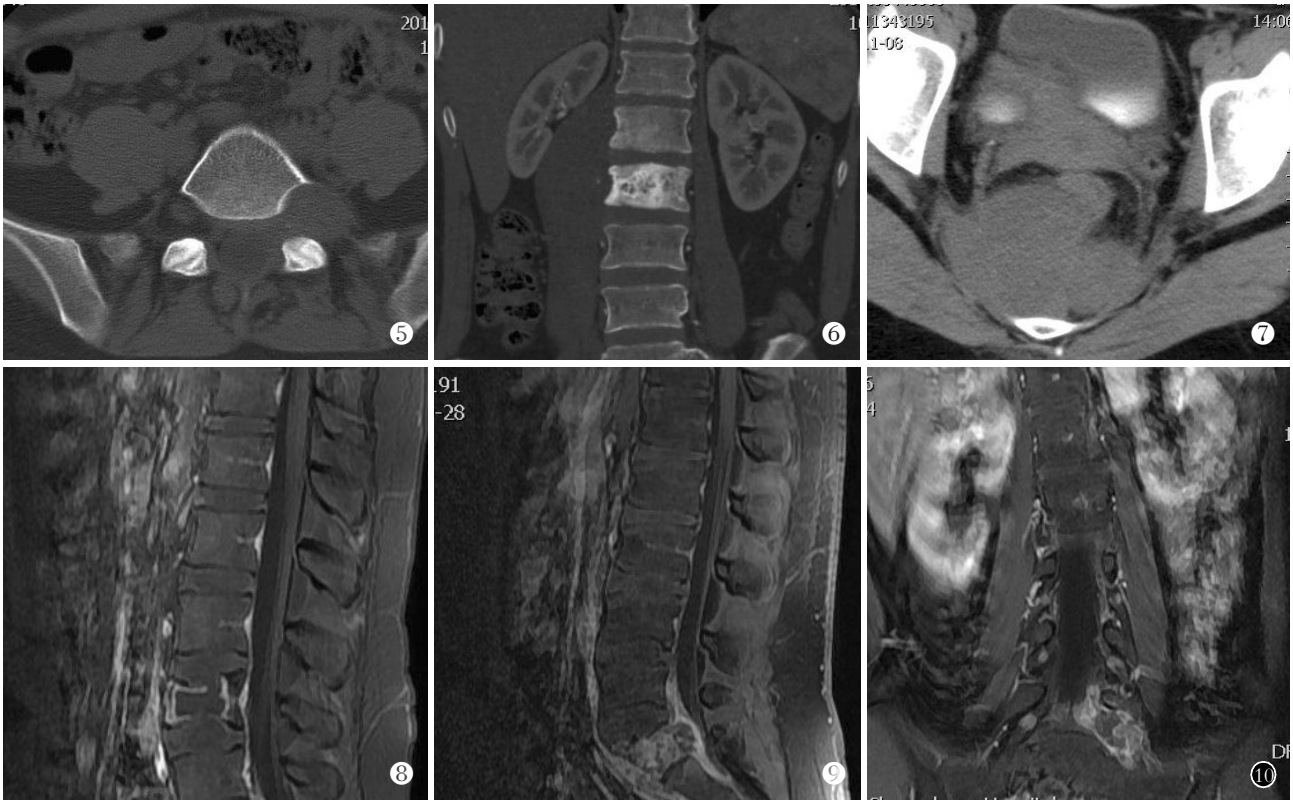

图 5　女，28 岁，L_5S_1 左侧椎间孔处跨椎管内外生长的软组织占位灶，椎间孔扩大，相邻骨空间骨质吸收　**图 6**　男，24 岁，$L_{1,2}$ 右侧椎旁巨大软组织密度占位灶，累及腰大肌，相邻椎体骨质密度不均匀增高，椎间隙无狭窄　**图 7**　女，19 岁，骶前间隙处巨大软组织密度占位灶，与骶椎、盆壁分界不清，直肠受压、前移　**图 8**　女，36 岁，L_4 椎体处占位伴骨质破坏，椎体压缩性骨折，相邻椎间盘及椎管未受累，椎体前方见软组织肿块，增强后不均匀强化　**图 9**　男，14 岁，S_1 椎体处占位伴骨质破坏，增强后不均匀强化，内见散在斑点状低信号灶，肿瘤沿椎前间隙及椎后硬脊膜外跨节段蔓延　**图 10**　女，28 岁，L_5S_1 左侧椎间孔处占位灶，增强后肿块强化明显，强化不均，沿神经走行方向跨椎管生长，伴椎间孔扩大，硬脊膜受累

Fig.5　Female, 28-year-old , the soft tissue mass growed in the L_5S_1 left intervertebral foramen across the canal, both inside and outside, the intervertebral foramen expanded, bone space adjacent to bone resorption　**Fig.6**　Male, 24-year-old, the huge soft tissue density mass in the $L_{1,2}$ right side, involving the lumbar muscle, bone density of adjacent vertebral body increased with uneven, there was no stenosis in the intervertebral space　**Fig.7**　Female, 19-year-old, the huge soft tissue density mass in the presacral space, and the boundary of sacral vertebral and pelvic wall was not clear, the rectum was compressed and antedisplaced　**Fig.8**　Female, 36-year-old, occupying lesion complicated with bone destruction on L_4 vertebral body, compression fracture of vertebral body, adjacent intervertebral disc and vertebral canal were unaffected, a soft tissue mass was in front of the vertebral body, showed inhomogeneous enhancement　**Fig.9**　Male, 14-year-old, occupying lesion complicated with bone destruction on S_1 vertebral body, the image showed inhomogeneous enhancement, there was the scattered mottled with low signal lesions, tumor along prevertebral space and dura mate spinalis cross segment spread　**Fig.10**　Female, 28-year-old, the occupying lesion on the left intervertebral foramen of L_5S_1, the image showed obviously strengthening with uneven after enhancement scanning, followed the nerve across spine to growth, with intervertebral foramen enlargement, the dura mate spinalis was involved

松有关。肿瘤常累及椎管，多位于硬脊膜外，并可沿硬脊膜外跨节段生长。肿块与周围组织分界往往不清，内常可见囊变、坏死区，部分病灶内可见散在的大小不等形态各异的残存骨，病灶内偶见肿瘤骨形成，部分瘤内可见新生迂曲的肿瘤血管。CT 表现：常见以等或稍高密度为主，边界常不清，少数可边界清，肿块密度常不均，增强后强化明显，强化不均，少数可见迂曲的肿瘤新生血管。MRI 表现：T1WI 呈不均匀低信号，T2WI 呈不均匀高信号[8,10-11]，肿瘤易变性、坏死、囊变，而实质部分细胞排列紧密、间质含水少，其信号与肌肉组织信号相近。常累及椎管硬脊膜，因硬脊膜有一定阻挡屏障作用，肿瘤常位于硬膜外，并一般不累及椎间盘，椎间隙常不窄，增强后强化明显，强化不均。本组 2 例瘤内伴多发迂曲肿瘤血管，文献少见报道。

骨外型：发生于椎旁深部软组织或椎管内，常呈浸润性生长，边界不清，肿物除体积较小外，一般密度及信号不均。中轴区域原发 pPNETs 累及骨骼时，以椎体和(或)附件的溶骨性破坏多见，并形态多保持完整，常缺乏骨膜反应，少见瘤骨形成，伴钙化及混合性少见。CT 表现：肿物多呈混杂密度，以等密度为主，内可见囊变、坏死，增强扫描明显强化，强化不均；MRI 表现：T1WI 多呈稍低或等信号，T2WI 多呈不均匀高或稍高信号，STIR 为高信号。分化好的骨

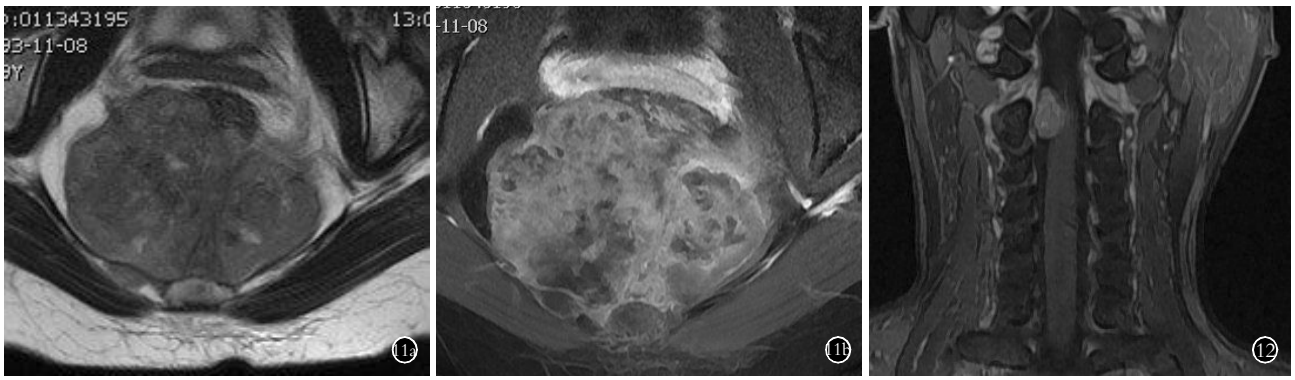

图 11　女性患者，19 岁，外周型原始神经外胚层肿瘤　**11a.** T2WI 骶尾椎前间隙巨大占位灶，与尾椎、直肠分界不清，呈分叶状改变，以稍高信号为主，内见散在斑片状高信号及条状低信号，直肠受压、前移　**11b.** 增强后肿块明显强化，强化不均，内见散在斑点状、斑片状无强化低信号灶　**图 12**　男性患者，49 岁，C_2 椎体水平椎管内见椭圆形软组织信号灶，边界清，颈髓受压、移位，增强后强化明显，强化不均

Fig.11　A 19-year-old female patient with peripheral primitive neuroectodermal tumors　**11a.** The huge occupying lesions occurred in the prevertebral space of coccygeal vertebrae, and the delimitation with caudal vertebrae and rectum was unclear, appearred lobulated change with slightly high signal, can find the scattered patchy high signal and strip like low signal, there was the rectum compression and antedisplacement　**11b.** After enhancement scanning, the mass occurred obviously strengthening with uneven, there was the low signal foci of scattered mottling and patching , and without enhancement　**Fig.12**　Male, 49-year-old, the image showed the oval soft tissue signal lesions in spinal canal of C_2, with clear boundary, cervical spinal cord compression and displacement, which had obviously enhanced with uneven after enhancement scanning

外型 pPNETs 易变性、坏死、囊变；而分化差的信号相对均匀，囊变及坏死不明显。本组 1 例位于椎管内髓外硬膜下，表现似神经鞘瘤，文献少见报道。

pPNETs 少见钙化或骨化，即使有也常较细小。丁晓毅等[12]认为骨 pPNETs 恶性程度高，理论上缺少钙化或骨化的时间。Dick 等[9]报道的 29 例中仅有 5 例发生轻度钙化。本组病例中 2 例肿块内见残存骨，1 例发生于颈椎骨内型 pPNETs 椎旁软组织内见大片钙化，椎体伴骨质硬化，罕见报道。中轴区域原发 pPNETs 软组织内散在高密度的残存骨或钙化形成“浮冰”样改变，以骨内型常见。

pPNETs 易发生远处转移。本组 10 例中有 5 例已发生肝脏、肺部的转移。

3.2　鉴别诊断

良性病变，主要是神经纤维瘤、神经鞘瘤与骨外型 pPNETs 鉴别，神经纤维瘤、神经鞘瘤常边界清，邻近骨质破坏以受压吸收为主，常边界清，并边缘常伴硬化；骨外型 pPNET 的骨质破坏以溶骨性常见，骨形态常完整，当其侵袭性小，骨质破坏不明显时，鉴别困难，当侵袭性明显、骨质破坏明显时，鉴别相对容易。

转移瘤，一般年龄较大，多有原发恶性肿瘤病史，常多发，而 pPNETs 一般发病年龄较年轻，常单发。

恶性肿瘤，主要是淋巴瘤。两者均为恶性小圆细胞肿瘤，软组织肿块往往较大，骨质破坏相对轻为其共同特点。但中轴区域原发 pPNETs 发病年龄相对淋巴瘤轻。骨内型 pPNETs 与淋巴瘤椎体周围同会出现软组织环绕，但通过复习文献[13-15]发现，脊柱原发单椎体淋巴瘤，淋巴瘤软组织肿块以后缘为主，常常累及椎管四周，形成“袖套”样改变，附件受累常见，易致椎管狭窄，与硬膜外淋巴组织丰富有关；而中轴区域原发 pPNETs 软组织肿块常以椎前为主，这大概与椎前间隙组织疏松、生长空间较大、后方受硬脊膜有阻挡作用有关，椎管以前壁受累常见，骨内型常有“浮冰”征，罕见“袖套”征，附件受累及椎管狭窄少见。骨外型 pPNETs 与淋巴瘤鉴别存在一定困难，但中轴区域原发 pPNETs 以单发病灶常见，以深部软组织常见，肿块常巨大，而淋巴瘤多发常见，淋巴组织丰富部位常见，肿块相对小。

总之，中轴区原发 pPNETs 以儿童及青少年多见，肿块常较大。骨内型肿块常包绕椎体，并以椎前间隙为主，均伴椎体骨质破坏，溶骨性常见，伴钙化及混合性少见，椎体原发常见，附件原发或受累少见，可伴椎体压缩性骨折，常有“浮冰”征，可伴多发新生小血管；骨外型以深部软组织间隙常见，少数原发于椎管，多伴椎体和(或)骨质破坏，以溶骨性为主，少数伴钙化。多不侵及椎间盘，椎间隙多不窄。CT 以等密度为主，密度不均匀，MRI 表现为等长 T1 不均匀稍长 T2 信号，增强扫描中度以上强化，强化不均匀，常伴坏死、囊变，极少数可伴瘤骨形成。治疗以手术为主，辅以放化疗，预后很差，即使治疗后也易复发和转移[16]。

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