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个案与短篇

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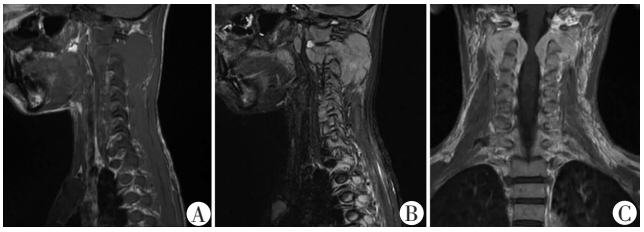
儿童颈部腺泡状横纹肌肉瘤1例

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［**关键词**］　横纹肌肉瘤；磁共振成像；儿童
［**中图法分类号**］　R738.7　［**文献标志码**］　B

1　临床资料

患者，男性，14岁。因颈部外伤后颈部疼痛、活动受限3周于2011年5月21日入我院。查体：颈部僵直，压痛，颈部屈伸、旋转、侧屈活动受限，伴疼痛，双上肢肌力、肌张力正常，皮肤感觉及血运未见明显异常。外院X线片：寰枢椎向前脱位，颈椎生理曲度变直。入院后1周行物理治疗（颌枕带牵引），颈部疼痛有所缓解。颈椎MRI平扫及增强扫描示：颈2、3椎体后缘欠连续，平扫齿状突信号略显不均匀。颈2～4椎体平面椎体、附件旁及周围软组织内见不规则形片状异常信号，T_1WI呈低信号，T_2WI呈高信号，信号欠均匀，范围约5.3 cm×5.0 cm，边界欠清，与椎管、椎体分界不清，并沿椎间孔走行，向椎管内生长，自左右方向压迫颈髓致局部变形，增强扫描病变强化明显、信号较均匀。双侧椎间孔扩大，颈髓左右方向受压、变形，前后方向受压不明显，未见异常强化；颈椎椎体信号未见明显异常强化（图1）。颈后肌肉穿刺活检见弥漫性小细胞浸润，侵犯肌组织，肿瘤细胞呈“圆形”，胞浆偏少，部分呈嗜酸性红染，胞核异型性明显，核分裂易见（图2）。免疫组化染色：MyoD1（－），Desmin（＋），Myogenin（＋），CD99（＋），FLI-1（－），CK（－），Vim（＋）。诊断：（颈后肌肉）小细胞恶性肿瘤，考虑腺泡状横纹肌肉瘤。因患者病情较重，不建议手术，继续颈部制动，并转至肿瘤科化疗，治疗后颈部包块明显缩小，颈部活动受限情况好转。



A：矢状位MRI T_1WI；B：矢状位MRI T_2WI；C：冠状位MRI T_1WI增强扫描

图1　RMS颈椎MRI平扫及增强扫描表现

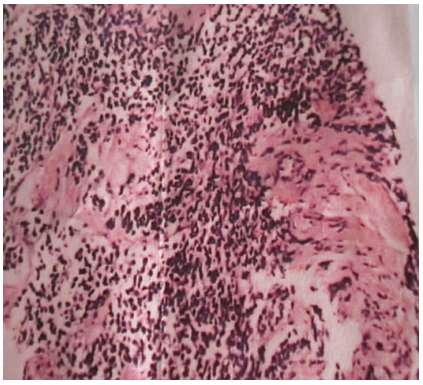


图2　RMS颈部病变穿刺活检病理学变化　（HE ×200）

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2 讨论

横纹肌肉瘤(rhabdomyosarcoma,RMS)为儿童及青少年最常见的软组织恶性肿瘤。RMS 由不同分化的横纹肌母细胞组成,源于具有肌源性分化能力的原始间充质,即可向横纹肌分化的原始间叶细胞,因此可在正常无横纹肌的解剖区域如前列腺、鼻咽部等处发生 RMS[1]。RMS 病理学分 3 型:①胚胎型最常见,约占 60%,主要发生于 15 岁以下,以头面颈部和泌尿生殖道多见;②腺泡状以 10 ~ 25 岁为主,常发于四肢深部软组织,其次为头颈部、躯干、会阴、盆腔及腹膜后;③多形性好发于老年人,四肢多见,且预后相对较差[2]。45% RMS 发生于头颈部[3],多以局部出现无外伤史的肿块,以及关节部位出现逐渐增大的肿瘤导致功能障碍而就诊,常有骨质溶骨性破坏[4]。

本例 MRI 表现特征:病变侵犯范围较广,以椎体及附件为中心,通过椎间孔向椎管内生长,与髓外硬膜下软组织相连,呈"匍匐式"浸润生长,与邻近结构分界不清;病变 T_1WI 呈低信号,T_2WI 呈较高信号,信号不均匀,边缘不规则,增强扫描呈明显强化,强化较均匀。而椎体形态、大小、信号未见异常;脊髓虽见左右方向明显受压,但信号未见明显异常。横纹肌肉瘤发生于椎体后方软组织及椎体者较为罕见,本例与成人胸椎胚胎性 RMS 描述之 MRI 表现较相似[5]。

脊柱 RMS 鉴别诊断包括:①淋巴瘤:起源于淋巴网状系统,容易侵入椎旁形成较大范围的软组织密度肿块,边缘清,范围较广,呈梭形,其纵径常常大于横径,周围可见大小不等软组织密度肿块,边界较清晰,还可经破坏的椎体后缘、椎弓根或经椎间孔侵入椎管内,围绕硬膜外生长,并向上、下方向发展呈袖套状浸润,可达 2 个椎体节段以上,其椎管内范围可明显大于椎体所累及的范围[5]。MRI T_1WI 多呈稍低或等信号,T_2WI 呈等和稍高信号。②转移瘤:患者一般年龄较大,有原发肿瘤病史,软组织肿块常破坏椎体,并以破坏椎体为中心发展,少见围绕椎管生长而不破坏椎管结构。

综上,RMS 是儿童及青少年比较常见的软组织恶性肿瘤,但像本例这样环绕椎管及椎体生长的横纹肌肉瘤比较罕见,其临床及影像表现尚无明显特征性,确诊仍依赖于病理及免疫组化检查结果。在以后的临床工作中如果遇到这种围管"匍匐式"生长的软组织病变,需要考虑到横纹肌肉瘤的可能。

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