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个案与短篇

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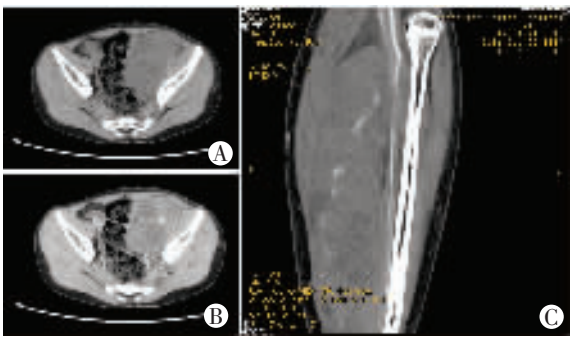
腹膜后及左下肢多发横纹肌肉瘤1例

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【关键词】 横纹肌肉瘤;计算机断层成像
【中图法分类号】 R738.7 【文献标志码】 B

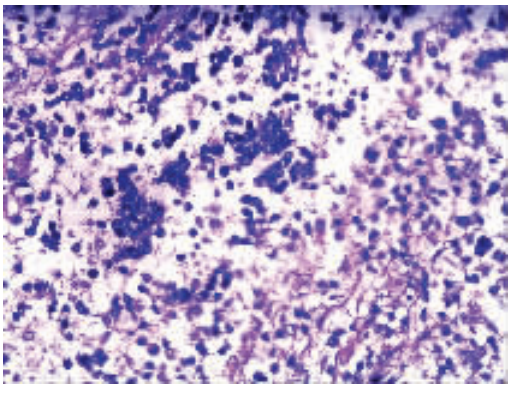
1 临床资料

患者,女性,13岁。因"左下肢肿胀1个月余,发现左腹股沟包块7 d"入院。查体见左下肢肿胀明显,左侧腹股沟区可扪及包块,质中,无压痛。下肢髋关节X线片:骨盆及髋关节未见明显骨质异常。CT检查发现腹膜后、盆腔偏左侧、左侧腹股沟区、腘窝及小腿后方软组织内多发结节状肿块影,边界清楚,密度不均匀,平扫实性部分呈等密度,CT值约40 HU,增强后呈轻度不均匀强化(图1A、B),以边缘假包膜强化为主,其内坏死区域无强化,延迟期强化程度减退不明显。最大者位于小腿后方,约4.8 cm×6.6 cm×21.8 cm,周围组织呈推压改变;骨骼未见明显侵犯(图1C)。CTA显示肿块侵犯包绕左侧髂外动脉,部分包绕左侧股动脉及腘动脉,致其轻度狭窄并推压移位呈弧形。由于多发肿块有沿着神经血管走行的特点,影像诊断考虑神经纤维瘤病可能。入院后在全麻下行腹膜后肿块姑息性切除术及腹股沟肿瘤切除术,术中探查发现左侧腹膜后约10 cm×12 cm的包块,质软,边界欠清,比较固定,沿左侧髂外动脉、髂外静脉、股动脉及股静脉走行,包裹左侧髂外动脉、髂外静脉及股动脉,上极达髂内动脉与髂外动脉分叉处,外侧缘达髂骨,内侧紧贴膀胱壁,下缘达卵圆窝。术后病理诊断:腹膜后小细胞恶性肿瘤(图2),结合免疫组化符合胚胎型横纹肌肉瘤。免疫组化:CK(-),Vim(-),WT-I(+++),Desmin(灶性+),Myo-D1(-),Myogenin(+),S-100(-),SMA(灶性+),LCA(-),Syn(+)。



A:平扫 肿块位于盆腔偏左侧,呈等低密度,边界清楚;B:增强动脉期 呈轻度不均匀强化;C:CT增强冠状重建 最大肿块位于小腿后方,其内可见液化坏死,胫腓骨未见骨质破坏

图1 多发横纹肌肉瘤患者CT表现



小圆形肿瘤细胞排列成致密区和疏松的黏液样组织交替分布,细胞大部分胞核呈梭形,核深染,胞浆嗜酸,病理性核分裂象易见

图2 多发横纹肌肉瘤病理学观察 (HE ×40)

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按需治疗)、累积暴露日和是否发生严重出血事件有关。当然,在此方面还需进一步多中心大样本的前瞻性研究。

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2 讨论

横纹肌肉瘤(rhabdomyosarcoma)是来源于横纹肌细胞或向横纹肌分化的间叶细胞的恶性肿瘤,以儿童和青少年多见,其发病率占儿童所有恶性肿瘤的5%,儿童软组织肿瘤的50%,仅次于神经母细胞瘤和肾母细胞瘤[1]。男、女性发病无明显差别,也有统计男性发病多于女性[2-3]。可发生于全身任何部位,以头颈部最为多见,其次为泌尿生殖道及腹膜后,少数发生于四肢,占15%~20%[3-4]。以单发多见,有少量多发横纹肌肉瘤文献报道,其发病部位分别为腹膜后、双侧嚼肌及左侧上下肢、鼻咽、乳腺及眼眶。该病例发生于腹膜后、盆腔、左侧腹股沟区及左下肢。最新病理分类将横纹肌肉瘤分为胚胎型(包括葡萄状肉瘤及梭形细胞型)、腺泡型、多形性3种类型。以胚胎型最常见,占50%~60%,绝大多数发生在婴幼儿,以头颈部、泌尿生殖道及腹膜后多见[3]。此例胚胎型横纹肌肉瘤发生于左下肢及腹膜后,年龄为13岁,属青少年,与文献报道不符。该病容易通过淋巴道及血行转移,以肺部多见,其次为骨髓、淋巴结及骨转移。10%~20%患者在发现时已发生远处转移[5]。本例患者目前尚未发现肺部及骨转移征象。

本例CT检查显示腹膜后、盆腔、左侧腹股沟区及左下肢多发肿块,沿血管神经分布,且长轴与神经血管走行一致,影像最初误诊为神经纤维瘤病。但回顾分析发现,多发肿块体积均较大,最大径线约22 cm,且坏死囊变明显。有作者统计四肢软组织肿瘤直径>8 cm,其恶性的可能性大[6]。并且该病例行CT血管成像检查,经多平面重建后清晰显示肿瘤侵犯包绕大血管,因此该患者就其CT特点应首先考虑恶性肿瘤的可能性。而神经源性肿瘤多呈不均匀明显强化,且钙化常见,但横纹肌肉瘤多呈轻-中度强化[3,5],该患者呈轻度持续性强化,并未见明显钙化显示,可兹鉴别。该患者未侵及骨质,对周围肌肉呈推压移位改变,导致误诊为良性病变,但此特点正是发生于四肢/腹膜后横纹肌肉瘤与头颈部的横纹肌肉瘤不同之处,头颈部病变则多呈浸润性生长,易破坏周围肌肉及骨质[2,7]。

多发性横纹肌肉瘤影像报道病例较少,影像表现总的来讲缺乏特异性,最终确诊需依靠病理诊断。

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