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• 病例报道 •

Ultrasonic manifestations of botryoid rhabdomyosarcoma of the biliary tract with pancreas invasion in child: a case report

儿童胆道葡萄簇横纹肌肉瘤并侵犯胰腺超声表现 1 例

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[中图法分类号]R445.1;R735.8

[文献标识码]B

患儿男, 3岁1个月, 因“排白陶土样大便 20 d, 皮肤巩膜黄染 10 d, 腹痛 1 d”就诊。家属述患儿无发热、呕吐, 皮肤无明显瘙痒; 小便深黄色。体格检查: 患儿皮肤、巩膜呈苍黄色。肝功能检查: 丙氨酸氨基转移酶 455 U/L, 天门冬氨酸氨基转移酶 366 U/L, 碱性磷酸酶 2331 U/L, γ -谷氨酰转氨酶 1637 U/L, 总胆汁酸 125.1 $\mu\text{m/L}$, 血清总胆红素 139.4 $\mu\text{m/L}$, 直接胆红素 109.4 $\mu\text{m/L}$; 血清肿瘤标志物: 糖类抗原(CA)19-9 9487.52 U/ml。超声检查: 胆总管中下段管腔内见一大小约 2.5 cm \times 1.3 cm \times 1.5 cm

团状低回声(图1), 形态规则, 其内回声不均匀并可见线样高回声, 与胆总管壁分界不清晰; CDFI于团状低回声内探及点状血流信号。该团块以上肝外胆管、左右肝管及肝内胆管均见不同程度扩张, 较宽处约 1.5 cm(胆总管)、0.7 cm(肝内胆管)。胆囊增大, 大小约 9.9 cm \times 1.9 cm, 壁光滑, 胆囊内未见异常回声。胰头部可见一大小约 3.4 cm \times 2.9 cm \times 3.4 cm 囊实性团状混合回声(图2), 边界欠清晰, 与胆总管内团状低回声相连续, 回声不均匀, 以低回声为主, 并可见囊状无回声区分布; CDFI于其实性部分探及点状血流信号。肝门部及腹主动脉旁可见多发增大淋巴结回声。超声提示: 胆总管中下段实性占位性病变, 胰头部囊实性占位性病变, 考虑横纹肌肉瘤可能性大。MRI及磁共振胰胆管成像提示: 胆总管中下段-胰头区占位病变, 考虑来源于胆总管内恶性肿瘤病变(横纹肌肉瘤)可能大, 伴病变上游肝内外胆管梗阻性扩张。PET-CT检查: 胆总管远端及胰头片状高密度影, 全身其他部位未见高代谢征象。

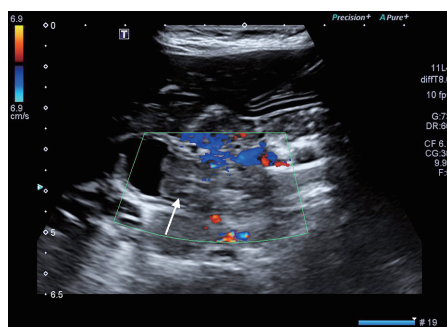


图1 肝门部斜切面声像图示胆总管中下段管腔内见团状低回声(箭头示), 可探及点状血流信号, 其以上胆总管扩张

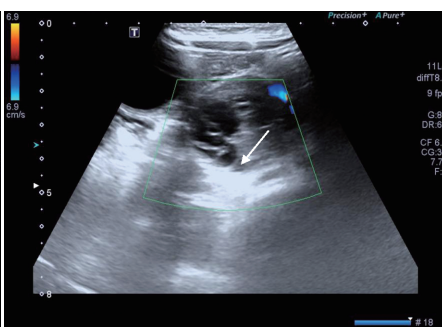


图2 上腹部剑突下横切面声像图示胰头部囊实性团状混合回声(箭头示)

部分探及点状血流信号。肝门部及腹主动脉旁可见多发增大淋巴结回声。超声提示: 胆总管中下段实性占位性病变, 胰头部囊实性占位性病变, 考虑横纹肌肉瘤可能性大。MRI及磁共振胰胆管成像提示: 胆总管中下段-胰头区占位病变, 考虑来源于胆总管内恶性肿瘤病变(横纹肌肉瘤)可能大, 伴病变上游肝内外胆管梗阻性扩张。PET-CT检查: 胆总管远端及胰头片状高密度影, 全身其他部位未见高代谢征象。

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患儿于我院胃肠外科行达芬奇机器人辅助保留幽门的胰十二指肠根治性切除术和胰肠、胆肠、十二指肠空肠吻合的消化道重建术。术后病理检查:(胆总管、十二指肠壶腹部、胰头)肿物符合葡萄簇横纹肌肉瘤;淋巴结未见肿瘤细胞浸润。术后进行化疗,随访至今,患儿恢复良好。

讨论:小儿胆道恶性肿瘤临床罕见,文献^[1-4]报道以2~6岁男童相对较多见,发病高峰为4岁,多为胚胎性横纹肌肉瘤。葡萄簇横纹肌肉瘤是胚胎性横纹肌肉瘤一种特殊亚型,好发于衬有黏膜的中空器官,如膀胱、阴道、鼻腔、鼻咽等,发生于胆道者极为少见,表现为息肉样、菜花样肿块,预后较好^[5]。横纹肌肉瘤起源于胆管壁,好发顺序依次为胆总管、左右肝管、肝内胆管^[1-2],肿瘤常沿着胆管黏膜下层生长,可蔓延侵犯周围组织器官,如肝脏、胰腺、十二指肠等部位。该病恶性程度高,常发生远处转移,因肿瘤位置不同可导致其临床表现各异。肝外胆道横纹肌肉瘤临床表现为进行性加重的阻塞性黄疸;肝内胆道横纹肌肉瘤则以腹痛、肝肿大、腹部包块就诊。CA19-9可以作为该病血清肿瘤标志物之一,本例患儿术前CA19-9高达9487.52 U/ml,术后2周降至35.54 U/ml(正常范围<37 U/ml)。目前胆道横纹肌肉瘤推荐手术、化疗和放疗的综合治疗方案,可有效提高临床治愈率。

结合本例患儿的超声图像及既往文献^[1-2,6-10],笔者总结了胆道横纹肌肉瘤的超声图像特征:①发生于肝外胆管者表现为肝外胆管内包块,以低回声为主,内部回声不均匀,可伴有不同程度、不同形态囊腔,与胆总管壁分界不清晰;②发生于肝内胆管者表现为肝内实性低回声包块或囊实性包块,边界清晰;③若肿瘤向周围组织浸润性生长,如十二指肠、胰腺等,可在局部形成一个边界模糊、与胆道肿物相连续的低回声包块或囊实性包块;④CDFI可于包块内部探及点状或点条状血流信号;⑤发生于肝外胆道伴梗阻者近端肝内外胆管可呈不同程度扩张、胆囊增大;⑥肝脏可增大,实质回声增粗、增高;⑦可伴有肝门区及腹腔内淋巴结肿大。

总之,胆道横纹肌肉瘤临床罕见,术前超声检查可为胆道

横纹肌肉瘤的临床诊断提供重要影像学依据,胆道走行区实性或囊实性包块,其内探及血流信号是本病主要的超声表现。但本病最终确诊依靠病理检查。

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