Intraperitoneal involvement in rhabdomyosarcoma CT findings in a child

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Intraperitoneal neoplastic involvement in rhabdomyosarcoma is rare and its incidence and imaging characteristics need to be further described. We present the computerized tomography (CT) findings of a case with pelvic rhabdomyosarcoma and intraperitoneal neoplastic involvement. Enhanced peritoneal and retroperitoneal masses were seen around the liver, spleen, in the paracolic gutters, and in the lesser sac without evidence of ascites, mesenteric nodules or omental caking. Our case also showed that absence of ascites does not preclude the presence of peritoneal involvement. Progression in the peritoneal disease was also well demonstrated by CT.

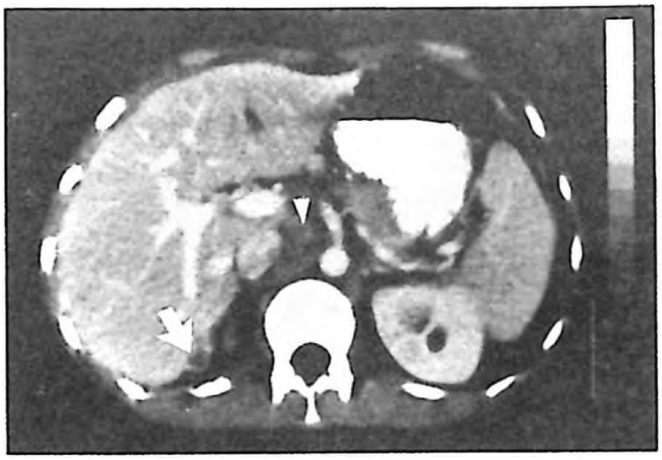
Key words: computerized tomography, rhabdomyosarcoma, peritoneum.

Rhabdomyosarcoma accounts for 5% to 8% of childhood cancer and presents in a wide variety of histologic types and of spread patterns of the tumor. The tumor infiltrates locally, invades lymphatics and blood vessels, and frequently presents with distant hematogenous metastases to lung, bone marrow and bone. Intraperitoneal rhabdomyosarcoma has been previously reported in adults; however, it presence in children was only recently described[1,2]. Chun et al.[2] reported an incidence of 11% for peritoneal involvement in rhabdomyosarcoma over the course of the disease and of 7% at the time of initial diagnosis. Despite this relatively high incidence, only six cases with radiological findings of rhabdomyosarcoma with peritoneal involvement have been described so far. Thus, imaging characteristics of intraperitoneal neoplastic involvement of rhabdomyosarcoma and its true incidence need to be further described. We present the computerized tomography (CT) findings of a child with pelvic rhabdomyosarcoma and peritoneal neoplastic involvement.

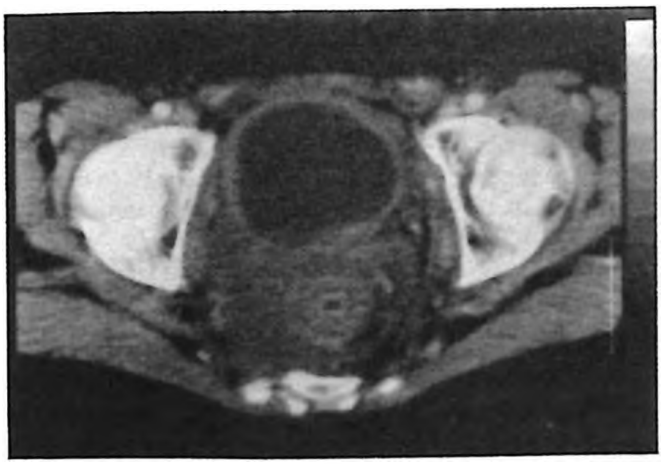
Case Report

A 16-year-old male with a 10 x 9 cm pelvic mass was dignosed as rhabdomyosarcoma by biopsy. In the initial postoperative CT, a small peritoneal nodule posterior to the right lobe of the liver and paraceliac adenopathy were seen (Fig. 1a). There was also a soft tissue mass encircling the rectum (Fig. 1b). This residual mass could not be separately identified from prostate, posterior wall of the bladder or other pelvic structures. There were no other peritoneal, mesenteric, omental or retroperitoneal masses or nodules. No ascites was noted. Histopathological subtype could not be identified. The tumor was accepted as stage III according to the IRS staging system (Intergroup Rhabdomyosarcoma Study Group)[3]. A chemotherapy regimen including vincristine, epirebicine, and cyclophoshamide plus radiotherapy was given. Since there was no response at the end of two months, chemotherapy regimen was changed to ifosphamide, cisplatin and vincristine. Six months after diagnosis a debulking surgery was performed. Nine months after diagnosis, a follow-up CT was performed, on which the previously observed perihepatic nodule and paraceliac adenopathy were increased in size. There were new perihepatic peritoneal masses with scalloped borders similar in appearance to pseudomyxoma peritonei (Fig. 2a). New peritoneal masses were also identified near the spleen, in the lesser sac, in both paracolic gutters, anterior to the transverse colon and in the falciform ligament (Fig. 2b). No omental

caking, mesenteric nodules or ascites was observed. All of the peritoneal masses were enhanced with central hypodense areas most likely consistent with necrosis. Peritoneal and retroperitoneal masses tended to displace the adjacent anatomic organs rather than invade them. Calcification was not observed in either the peritoneal or retroperitoneal masses. Recurrent pelvic mass was stable in size and appearance on the follow-up CT. He died with progressive disease at 22 months from diagnosis.

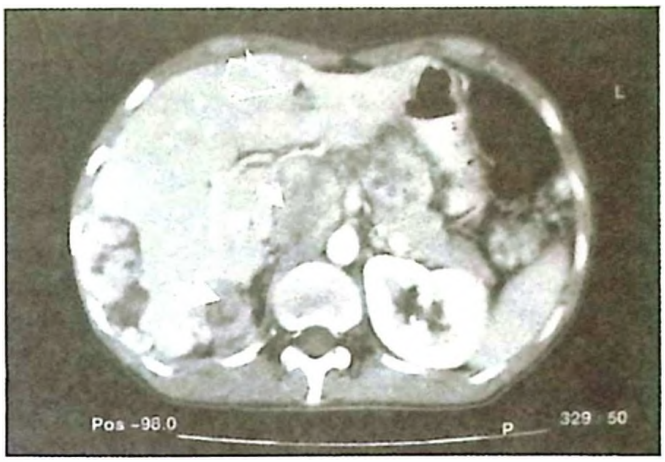


(a)

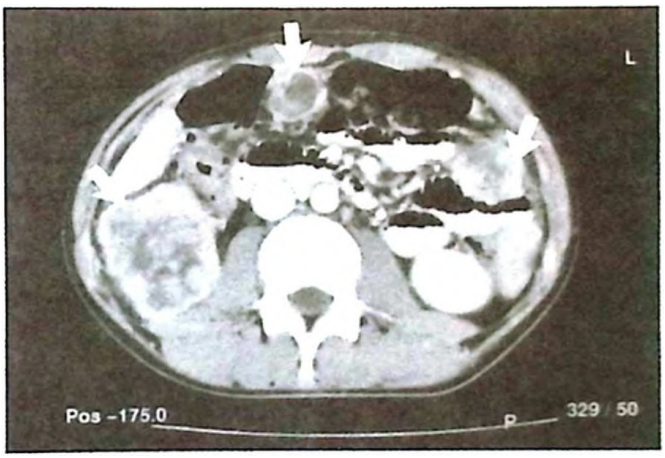


(b)

Fig. 1. Post-operative axial contrast-enhanced CT images of a 16-year-old boy with the dignosis of prostatic rhabdomyosarcoma. CT scan at the level of liver (A) shows small perihepatic peritoneal nodule (arrow) and paraceliac adenopathy (arrowhead). In pelvis (B) note the infiltrative soft tissue mass surrounding the rectum, prostate and posterior wall of the bladder.



(a)



(b)

Fig. 2. Follow-up (9 months later) axial constrast-enhanced CT scans of the same patient. CT scan at the level of liver (A) demonstrates increase in the size of the previously noted perihepatic nodule (arrow) and paraceliac adenopathy (curved arrow). Additional new peritoneal masses around liver, spleen, in the falciform ligament (open arrow), and in the lesser sac are also seen. Lower image through the abdomen (B) demonstrates preitoneal masses in the paracolic gutters and anterior to the transverse colon (arrows). Note the absence of ascites.

Discussion

Rhabdomyosarcoma is the most common soft tissue sarcoma in childhood, representing about 5-15% of all solid malignancies in children[4]. It is thought to arise from primitive mesenchymal cells, and it may arise from almost any organ in the body. Histologically there are two major subtypes (embryonal and alveolar) that have different

propensities for primary site and metastatic potential[5]. Embryonal rhabdomyosarcoma is most common in the head and neck, genitourinary tract and extremities. It has a more favorable outcome than the alveolar subtype. Alveolar rhabdomyosarcoma is more aggressive, often presenting with metastatic disease, and is more commonly located in the extremities, trunk and perineum. Head and neck (40%) and the genitourinary tract (20%) account for most primary rhabdomyosarcomas[4].

Intraperitoneal neoplastic involvement in children is less common, and limited information is available about its incidence and imaging features. There is only one series in the literature about malignant intraperitoneal neoplasms of childhood[6]. Most of the data are from case reports. Lymphoma, mesothelioma, desmoplastic small round cell tumor, malignant fibrous histiocytoma, immature teratoma, germ cell tumors, Wilm's tumor, neuroblastoma and intracranial tumors via ventriculoperitoneal shunts have been previously described to have either primary or metastatic neoplastic peritoneal involvement[7-15]. Another distinct entity causing multiple discrete tumor nodules in the peritoneum, mesentery and omentum is peritoneal leiomyosarcomatosis[16]. Recently rhabdomyosarcomas have also been reported to have intraperitoneal neoplastic involvement[2]. Detection of the peritoneal involvement is clinically very important since its presence alters the management. In their series of 55 children with rhabdomyosarcoma, Chung et al.[2] reported three cases with primary peritoneal rhabdomyosarcoma, two cases with pelvic rhabdomyosarcoma (paratesticular and prostate) and one case of extremity rhabdomyosarcoma with metastatic peritoneal involvement. Ascites, which was present in all cases with peritoneal involvement in Chung's series, was not noted either CT scan of our patient, despite the presence of extensive peritoneal metastases. Our case showed that absence of ascites does not preclude the possibility of intraperitoneal spread in rhabdomyosarcoma. Mesenteric nodules described in three of the six patients in Chung's series were not seen in our case. We did not see any omental caking, similar to the three patients in Chung's series with primary extraperitoneal rhabdomyosarcoma. All of the peritoneal masses in our case were enhanced, despite some of the reported non-enhanced peritoneal masses described by Chung et al.[2]. The presence of peritoneal involvement in children with rhabdomyosarcoma requires the use of more intensive treatment regimens. Computerized tomography could successfully monitor the peritoneal disease and demonstrate its progression.

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