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Case Report

Desmoplastic small round cell tumor: postoperative retroperitoneal mass

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ABSTRACT

We describe the case of a 14-year-old boy who presented with a large, 17.6-cm retroperitoneal mass, along with multiple metastases, and was diagnosed with desmoplastic small round cell tumor. After initial chemotherapy, he underwent gross total resection with a positive margin. On postoperative radiation planning computed tomography, a 6.8-cm heterogeneous mass was noted in the surgical bed. Given the tumor's aggressive nature and positive surgical margins, there was real concern for recurrent disease. Further evaluation with magnetic resonance imaging elucidated that the mass consisted of simple fluid and fat, without contrast enhancement, suggesting a postoperative fluid collection. He was able to continue with adjuvant treatment as planned. This case example illustrates that even large postoperative heterogeneous masses may still be related to postoperative fluid collection in patients with aggressive tumor. However, it is important to rule out recurrent disease before starting adjuvant therapy given improved outcomes with gross total resection in desmoplastic small round cell tumor.

Case presentation

A 14-year-old boy with a previous history of acute myeloid leukemia treated with chemotherapy and bone marrow transplant presented with nausea, vomiting, and epigastric pain. Initial workup revealed elevated liver function tests, a left-sided abdominal mass, and multiple liver lesions on abdominal ultrasound. Computed tomography (CT; Fig. 1, Panel A) demonstrated a large, heterogeneous, left retroperitoneal mass measuring 17.1 cm × 14.4 cm × 17.6 cm that crossed midline, replaced the left kidney, displaced the aorta toward the right, displaced the pancreas superior and anterior, displaced the spleen superiorly and laterally, and caused mass effect on the stomach. Retroperitoneal lymphadenopathy, multiple hypodense liver lesions in both the right and left lobes with the largest measuring 6.0 cm in maximal dimension, multiple bilateral pulmonary nodules with the largest measuring 2.3 cm in maximal dimension, and sclerotic lesions in bilateral iliac bones and the L4 vertebral body were also noted. Liver core biopsy indicated desmoplastic small round

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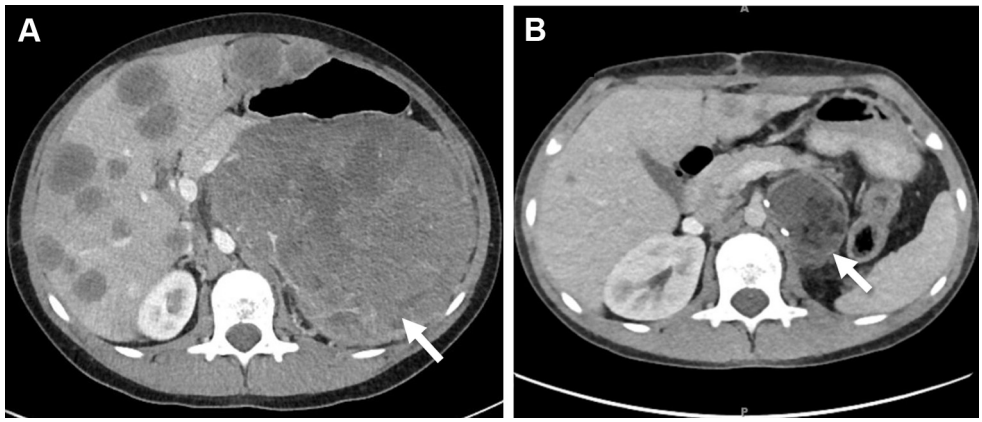

Fig. 1 – CT abdomen and pelvis at initial presentation (A) and 6 weeks after surgical resection (B). Arrows indicate preoperative and postoperative masses.

cell tumor (DSRCT) with immunohistochemical staining positive for desmin, keratin AE1/2, CAM5.2, and WT-1 and negative for CD99, myogenin, and S100 protein.

He initiated treatment with a chemotherapy regimen consisting of irinotecan, temozolomide, and temsirolimus, followed by vincristine, doxorubicin, and cyclophosphamide then ifosfamide and etoposide. A good response to chemotherapy was noted radiographically, with decreased size of the primary mass to 14.1 cm × 10.8 cm × 13.1 cm, as well as decreased size of the liver and lung lesions and retroperitoneal lymphadenopathy. He then underwent resection of the primary mass, which was found to involve the left kidney, renal sinus, and perirenal soft tissue, with attachment to the aorta. Pathology indicated a positive peripheral soft-tissue margin but a negative hilar margin. On CT scan obtained six weeks later (Fig. 1, Panel B) for postoperative radiation planning, a new 5.2 cm × 5.6 cm × 6.8 cm hypodense, heterogeneous mass was noted in the surgical bed, concerning for recurrent disease.

Further evaluation with magnetic resonance imaging (MRI) of the abdomen and pelvis (Fig. 2) with and without gadolinium and with T1-, T2-, and diffusion-weighted images helped clarify that the mass consisted of simple fluid and fat, with no internal enhancement on any phase of contrast administration, suggesting postoperative seroma and/or lymphocele and not recurrent disease. The patient then proceeded with postoperative radiation therapy to the whole abdomen and pelvis to a total dose of 30 Gy (liver to only 24 Gy). He has since received additional chemotherapy and haploidentical bone marrow transplant, with no evidence of disease progression.

Discussion

The differential diagnosis of a postoperative surgical bed lesion in the retroperitoneum is broad and includes lymphocele, seroma, hematoma, omental infarction, or recurrent disease. DSRCT is a rare and highly aggressive tumor containing desmoplastic stroma that affects primarily young men (median age of 19 years), with a 3-year overall survival of 44% and 5-year overall survival of 15% based on a larger review of 66 patients treated at Memorial Sloan-Kettering Cancer Center from 1972 to 2003 [1]. In the case of incomplete, margin-positive surgical resections, locoregional recurrence is of particular concern, even soon after surgery. Gross total resection is essential for improved survival, as 3-year survival is 58% in patients with gross total primary tumor resection (>90% tumor resection), versus 0% in those with <90% primary tumor resection [1]. Thus, it was particularly important in this case to rule out local tumor recurrence before proceeding with adjuvant radiation therapy.

DSRCT usually presents as a primary abdominal mass along with multiple peritoneal and liver lesions and can also metastasize to the lungs, bone marrow, and lymph nodes in the neck, groin, and mediastinum [1–4]. Patients are managed with aggressive multimodal therapy, including neoadjuvant chemotherapy, maximal surgical debulking, intraperitoneal chemotherapy in some cases, adjuvant

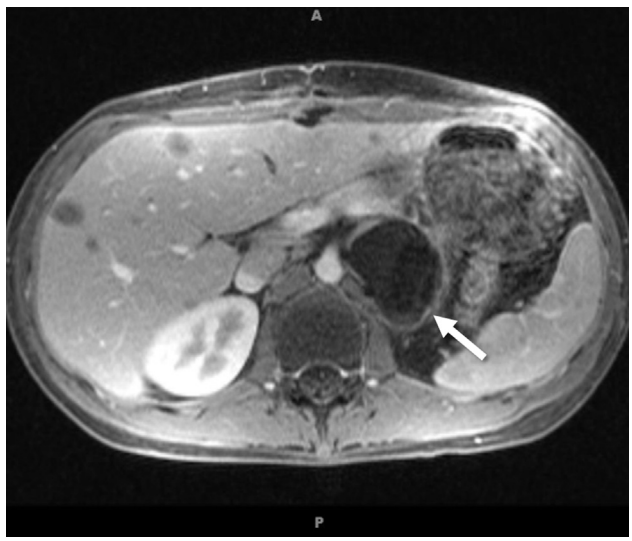

Fig. 2 – MRI abdomen and pelvis, T1-weighted image after administration of gadolinium, 7 weeks after surgical resection. Arrow indicates postoperative mass.

whole abdominopelvic radiation therapy, and stem cell or bone marrow transplant [1,4–8]. Patients receiving trimodality therapy with chemotherapy, surgical resection, and radiation therapy have improved 3-year survival (55%) compared with patients who are not treated with all three modalities (27%) [1]; however, outcomes remain poor despite aggressive treatment. A more recent study from M.D. Anderson does suggest potentially greater survival with improved salvage therapies, with a median overall survival of 60 months despite a median disease-free survival of 10 months [8].

Regarding imaging characteristics of DSRCT, CT findings typically include heterogeneous enhancement with contrast, cystic areas in approximately 50% of cases, and calcification within soft-tissue lesions in approximately 20%-30% of cases [2,9]. However, CT imaging is suboptimal for differentiating DSRCT from other processes because of poor soft-tissue contrast. On MRI, these tumors have heterogeneous hypointense or isointense T1 and hyperintense T2 signal, as well as heterogeneous contrast enhancement with gadolinium [9–11]. This patient's MRI demonstrated no significant enhancement with gadolinium within the lesion but rather characteristics of simple fluid and fat. Thus, postoperative seroma or lymphocele was favored, as opposed to tumor recurrence. Although the role of PET-CT is not well established for evaluating DSRCT, reports have shown that many of these tumors have intense FDG avidity [2,10,12], and thus PET-CT could also have been considered in the evaluation of this lesion. In the event that additional imaging could not definitively rule out recurrence, biopsy or surgical evaluation would have been considered, given the importance of maximal surgical resection before proceeding with adjuvant therapy for DSRCT.

Conclusions

This case example illustrates that even large postoperative heterogeneous masses may still be related to postoperative fluid collection in patients with aggressive tumor and that it is particularly important to investigate such findings before proceeding with adjuvant therapy.

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