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The Surgical Management of a Stage III Wilms Tumor Presenting With Perforated Appendicitis

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Summary

The majority of the cases of nephroblastoma do not present with abdominal pain. We present a patient with an acute surgical abdomen due to perforated appendicitis and an incidental abdominal mass. He underwent an urgent appendectomy after which tumor specific therapy was successfully initiated.

Keywords

perforated appendicitis; nephroblastoma; pediatric appendicitis

Nephroblastoma (Wilms tumor) is the most common primary malignant renal tumor found in childhood; its incidence reaching about 500 cases annually. The peak age of diagnosis occurs at around 3 years of age.¹ It is often found incidentally by a parent, who palpates a flank mass while bathing or dressing the child. The majority of these patients do not present with acute abdominal pain.² We present a patient with Wilms tumor, who presented with an acute surgical abdomen, whose diagnostic workup was complicated by concomitant acute perforated appendicitis. Although for a variety of solid tumors, there are several reports of acute appendicitis after the initiation of chemotherapy,³ this is the first report of acute perforated appendicitis preceding the diagnosis of a nephroblastoma.

CASE REPORT

A 5-year-old healthy, African American boy presented with focal right lower quadrant pain for 24 hours. He also had intermittent abdominal pain beginning a week before presentation. On examination, the patient had focal right lower quadrant tenderness with mild rebound. A right upper quadrant mass/fullness was also noted. His white blood cell count was 9.8 K/ μ L with 84% polymorphonuclear leukocytes. A urinalysis was negative. Given his pain and right upper quadrant mass/fullness, a computed tomography (CT) scan was obtained that demonstrated a dilated appendix with abscess formation, and a large heterogeneous mass arising from the superior aspect of the right kidney, extending into the posterior right hepatic lobe, likely representing a nephroblastoma (Wilms tumor).

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The patient underwent urgent open appendectomy followed by a CT-guided biopsy the following day, which yielded an inadequate tissue. The patient completed his antibiotic course and then underwent elective open biopsy and port-a-catheter placement for chemotherapy. Review of pathology sent from the open appendectomy revealed acute fibropurulent and acute peritonitis involving the appendix. The open biopsy revealed the renal mass to be a Wilms tumor with favorable histology.

He was then treated with the standard medical therapy for stage III Wilms tumor with favorable histology.¹ After 12 weeks of chemotherapy, a repeat CT scan demonstrated a markedly reduced tumor load. A right radical nephrectomy was performed along with a nonanatomic hepatic wedge resection (due to local invasion of the liver). Review of pathology showed no residual viable Wilms tumor in the renal or hepatic portions of the surgical specimen. Postoperatively, the patient has done well and will complete his chemotherapy.

DISCUSSION

This is an unusual presentation of Wilms tumor. A search of the literature revealed 1 case report that found acute perforated appendicitis associated with Wilms tumor, though only in the context of preoperative chemotherapy for the malignancy.² In fact, the majority of Wilms tumor patients do not present with acute abdominal pain, and reported rates of these symptoms have ranged from 25% to 42% of cases.³⁻⁵ The mechanisms of pain vary, from tumor-mediated distention of Gerota capsule and surrounding tissues, hemorrhage within the capsule, ureteral obstruction,⁶ to spontaneous rupture of the hemorrhagic tumor causing peritoneal irritation,⁷ a rare event with reported incidences ranging from 0.8% to 3%.^{8,9}

As a surgical team, we were posed with 2 novel issues concerning the optimal diagnostic decision making. There was no question that this patient needed an urgent operation for his perforated appendicitis. However, whether or not we ought to perform a biopsy of the tumor concomitant to the appendectomy was in question, considering that the tumor was unresectable at the time of discovery on CT scan due to liver invasion. The advantage of performing the 2 procedures together laid in the fact that this patient was very young, and any ability to limit the amount of procedural sedation and anesthesia in our young patient was viewed upon favorably. However, performing the 2 procedures concomitantly created the concern that any intra-abdominal infection may seed into the renal mass upon biopsy, which would likely complicate future surgical management of the tumor.

Had the tumor been resectable at the time of discovery, our approach to management would have been similar with respect to the prioritization of the patient's surgical issues. Specifically, we would first control the intra-abdominal infection by performing an urgent appendectomy. This would then be followed by appropriate postoperative antibiotic therapy. Only upon completion of the course of antibiotics would we then consider performing a radical nephrectomy to definitively treat the Wilms tumor.

The second issue we faced was whether to perform the appendectomy through a laparoscopic or open approach. The general concern with the laparoscopic technique was whether impeccable control of the intra-abdominal infection could be obtained, given the renal mass that would need a biopsy and, eventually, surgical resection.

Patients with Wilms tumor do not usually present with an acute surgical abdomen. Our patient presented with focal peritonitis and right upper quadrant mass/fullness. A CT scan was performed, revealing perforated appendicitis and a large, unresectable, right renal mass. Our management of this patient included an urgent open appendectomy with an appropriate course of antibiotics, followed by an open renal biopsy. Once the diagnosis of Wilms tumor

was confirmed, the patient was given preoperative chemotherapy followed by a radical nephrectomy.

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