SPONTANEOUS RUPTURE OF WILMS' TUMOR PRESENTING AS ACUTE APPENDICITIS: A RARE CASE PRESENTATION

Henry TCL[1], Tan JH[1], and Vellusamy VM[2].
1Department of Surgery, Hospital Sultanah Aminah, Johor Bahru, Malaysia
2Department of Pediatric Surgery, Hospital Sultanah Aminah, Johor Bahru, Malaysia

Correspondence:
Henry Tan Chor Lip,
General Surgeon Department of Surgery,
Hospital Sultanah Aminah,
Johor Bahru, Malaysia
Phone number: +6072257000
Email: relos1402@gmail.com

Abstract

In Malaysia, incidence of Wilms' Tumor (WT) is rare, with only 8 cases reported over four years according to the Malaysia Cancer Registry Report (2012-2016). Reports of spontaneous rupture of WT are scarce and the commonest presentation of this entity is a palpable mass in the abdomen. Herein, we report on the unusual presentation of an 11-year-old boy with typical symptoms of acute appendicitis. The child was subjected to an initial open appendicectomy and diagnosed intra-operatively with a right sided ruptured renal mass. Subsequent investigation leading to the diagnosis of a ruptured Wilms' tumor and its treatment is discussed in this case presentation.

Keywords: *Child, Wilms' Tumor, Kidney Neoplasm*

Introduction

Pre-operative spontaneous rupture of Wilms' Tumor (WT) is defined as the presence of blood or floating tumor in the peritoneal cavity with gross rupture through the capsule or peritoneal tumor implants (1). According to the Malaysian Cancer Registry Report (2012-2016), the reported occurrence of WT is scarce with only eight cases reported amongst males aged between 10-15 years old (2). Reports of spontaneous rupture of WT are also an uncommon presentation of WT (3). To our knowledge, there were no cases reported in the literatures of WT presenting as acute appendicitis. Herein, we report on an 11-year-old boy that was admitted with typical presentation of acute appendicitis and subsequently diagnosed as a right WT with spontaneous retroperitoneal rupture.

Case Presentation

An eleven-year-old boy presented with acute lower abdominal pain for one day. The abdominal pains were associated with fever and vomiting. Further history revealed a previously healthy boy with no recent trauma nor any symptoms of urinary tract infection. Physical examination showed, dehydration with localized abdominal peritonism over the right iliac fossa radiating to the right hypochondriac region. Vital monitoring was within normal limits with a recorded low-grade fever of 37.5 degree Celsius. The white cell count was raised, 13.6 x 10^9/L [4.0 -11.0] with a low hemoglobin level of 10.9 g/dl [13.0-18.0]. Blood urea, electrolytes and creatinine levels were within normal limits. There were no red blood cells seen on urine for microscopic examination. Due to the short history of symptoms, positive peritonism over the right iliac fossa and raised white cell count, the boy was diagnosed with acute appendicitis. He underwent an emergency appendicectomy on the same day. Upon entering the peritoneum, there was presence of minimal hemoserous peritoneal effusion with a mass which initially appeared to be a sealed right sided zone two retroperitoneal hematoma. The mass was non-pulsatile without evident expansion intraoperatively. There was no gross macroscopic evidence of a breach of the posterior peritoneal abdominal surface. Appendicectomy was performed and abdomen was closed without further dissection of the mass. Post-operatively, an urgent computed tomography (CT) thorax and abdomen revealed a localized right solid cystic renal mass measuring 8.4 x 9.3 x 10.9 cm with contrast extravasation at the superior pole, which indicates tumor rupture. There was no radiological evidence of distant metastases. Ultrasound guided percutaneous biopsy tested positive for WT-1 stains, which led to the diagnosis of a right Wilms' Tumor. Pre-operative chemotherapy was commenced with actinomycin D,

vincristine and doxurubicin for eight weeks. Radiological re-evaluation of the mass on CT abdomen appeared larger measuring 9.4 x 10.1 x 13.2 cm with replacement of solid to cystic component as seen in Figure 1. An elective radical nephrectomy was performed via a right upper transverse laparotomy. Upon entering the posterior peritoneum, there was a large solid cystic right renal mass encasing the abdominal aorta and inferior vena cava. The mass was adherent to the liver and right undersurface of the diaphragm. A vascular surgeon was engaged on-table to assist in the dissection to free the tumor away from the aforementioned great vessels (Figure 2A and 2B). Despite best efforts by both the vascular and pediatric surgeons, approximately 95 % of the tumor was resected with residue mainly over the great vessels (abdominal aorta and inferior vena cava) as seen on Figure 2B. Inevitably, due to its large size, tumor rupture occurred at the supero-lateral portion of the tumor despite meticulous dissection (Figure 2C). An immediate measure to prevent further intraperitoneal contamination of tumor spillage was performed intraoperatively. The resected specimen revealed a solid cystic renal mass measuring 15 x 9.5 x 9 cm, weighing 670 gm. There were large areas of necrosis, which indicates response to pre-operative chemotherapy. On microscopic examination, the tumor was found to contain 80 % of epithelial cells, 15 % of stromal cells and less than 5 % of blastemal cells, which were positive for WT-1 stains (Figure 3A, 3B 3C). There were no features of anaplasia (extreme nuclear and cytologic atypia). Evidently, tumor rupture was revealed by microscopic visualization of focal disruption of the renal capsule (Figure 3D). With the classical triphasic cells, the tumor was classified as a Wilms' tumor, intermediate risk epithelial type with evidence of tumor perforation. Recovery was uneventful and the patient was subsequently discharged on post-operative day 15. The child was transferred to the pediatric oncology department for chemotherapy and subsequent radiotherapy.

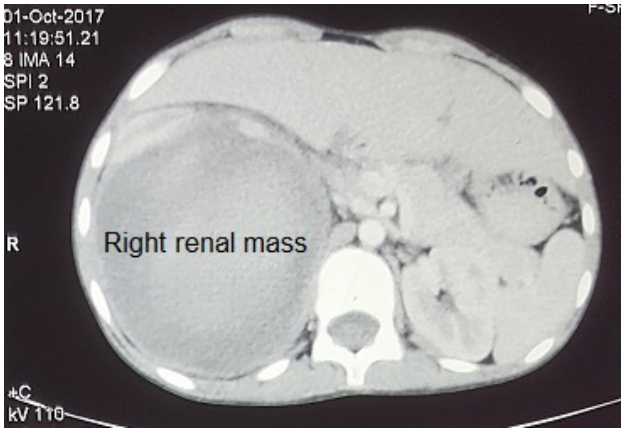
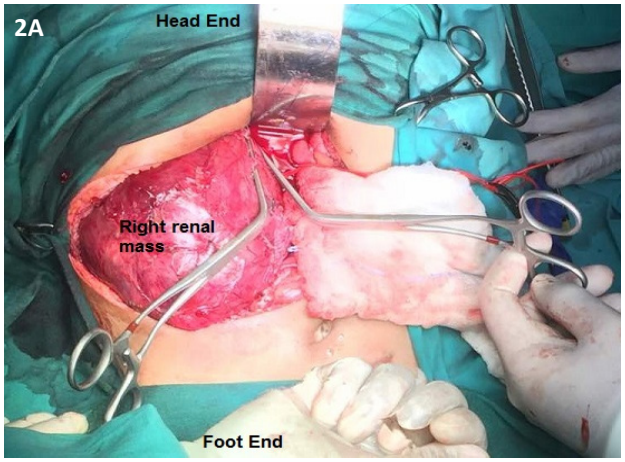
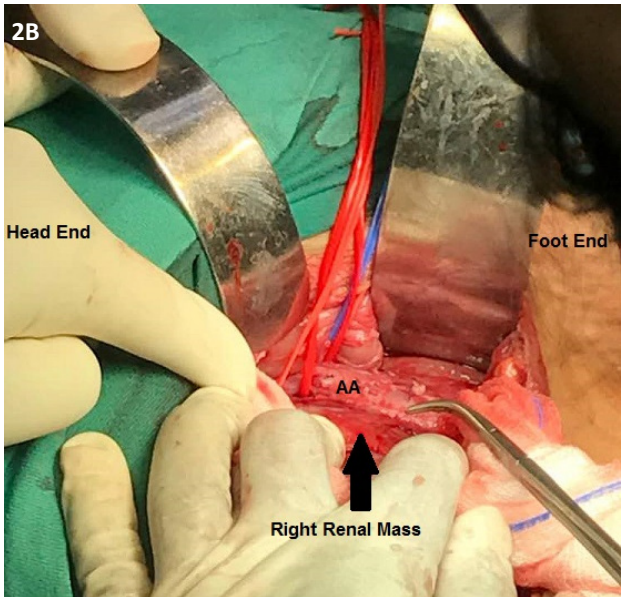


Figure 1: Contrast enhanced CT abdomen showing a right renal mass





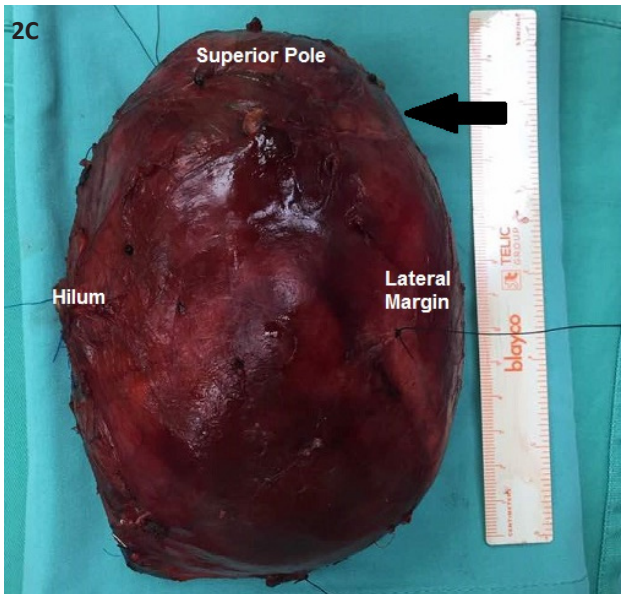
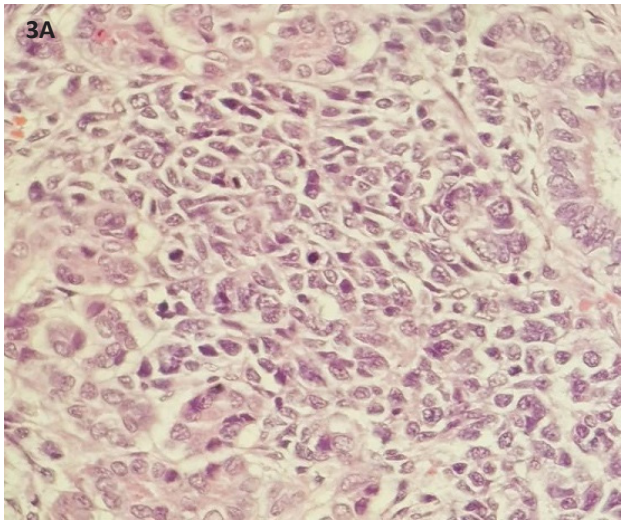
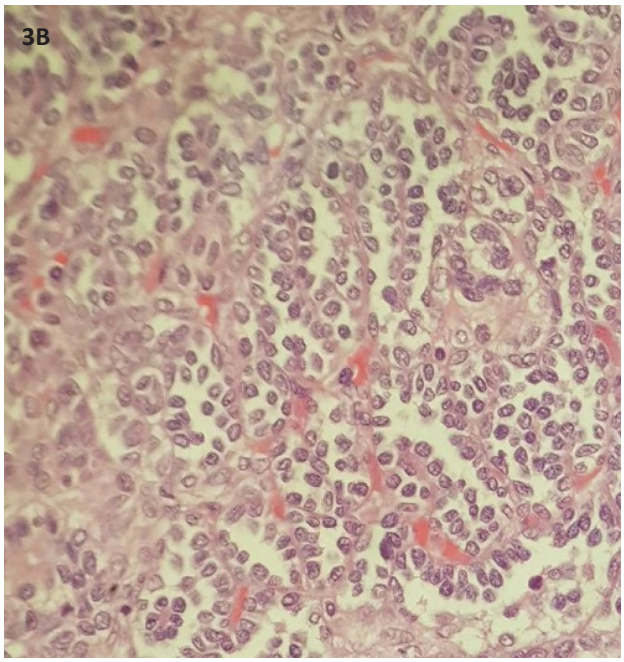
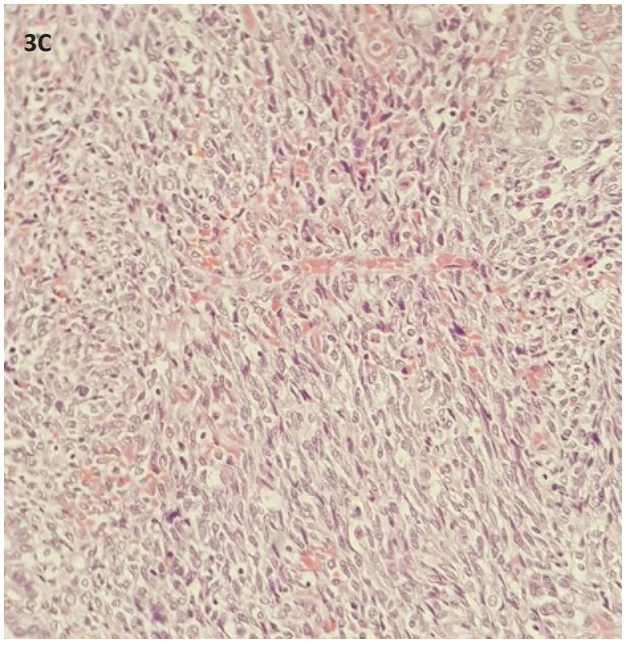


Figure 2: A. Intra-operatively, there is a large solid cystic mass occupying the right side of the abdomen. B. The right renal mass is densely adhered the abdominal aorta (AA). C. Resected specimen with arrow pointing towards site of rupture intra-operatively superio lateral margin







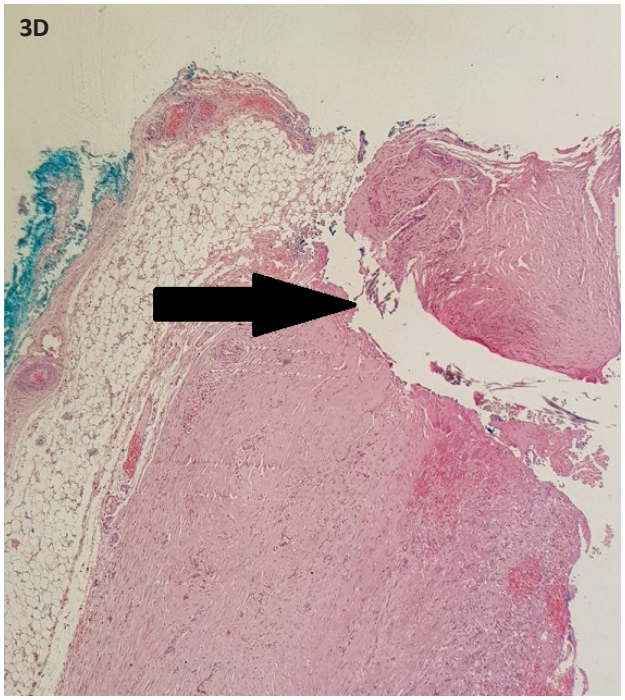


Figure 3: Histological slides of the resected specimen showing A. Blastemal cells. B. Epithelial Cells. C. Stromal cells. D. Microscopic evidence of focal disruption of capsular (black arrow) at the superior lateral site of tumor rupture

Discussion

Malaysian Cancer Registry Report (2012-2016) only recorded a total of eight renal cancers amongst young males aged 10-15 years old as described in this case presentation. This data highlights the limited number of cases diagnosed locally especially in Wilms' renal tumor which is diagnosed within the pediatric age group (2). Patients are generally asymptomatic until a mass is palpable in the abdomen. Initial presentation of abdominal pain due to tumor rupture is uncommonly seen, usually occurring after minor abdominal trauma over a large renal mass (4). In our case, the presenting symptoms were acute abdominal pain over the right iliac fossa, raised white cell count and pyrexia, which led to the initial diagnosis of acute appendicitis. An ultrasound of the abdomen was not performed in this case due to the acute presentation which mimics appendicitis with localized right iliac fossa guarding and a raised white cell count. In the presence of abdominal guarding eluded to the positive findings of a right sided abdominal mass where an ultrasound abdomen may be beneficial in pre-operative planning. There were no history of trauma and urine for microscopic examination tested negative for blood, which did raise to the suspicion of a renal pathology. Brisse et al. reported that abdominal pain as the commonest clinical sign of tumor rupture but, this is with a positive history of abdominal trauma (5).

The treatment for tumor rupture is emergency surgery. However, surgery is decided only after continuous monitoring guided by serial imaging and a failed

conservative management. Common criteria for emergency surgery include the presence of a large expanding hematoma, urinoma > 4 cm and intense abdominal pain (4). This patient responded to analgesics and diagnosis was confirmed via an ultrasound guided percutaneous biopsy.

The gold standard to determine WT tumor rupture is based on gross macroscopic intraoperatively or microscopic visualization (5). Diagnosis of tumor rupture is crucial as it influences the subsequent treatment and prognosis (6). According to the International Society of Pediatric Oncology (SIOP) group, the stage of disease changes to IIIC if there is any evidence of tumor rupture before or intra-operatively. Oncology treatment involves 28 weeks of chemotherapy followed by abdominal radiotherapy (7, 8). In a retrospective study performed on a population of patients with preoperative WT rupture, 26 patients in the stage III group (including 15 with incomplete resection) were subjected to flank radiotherapy. From the 26 patients, only one patient had peritoneal relapse, five patients progressed to metastatic disease and the majority of 20 patients were in complete remission within one year after completion of treatment. From this study, the majority with stage III disease will respond to oncologic treatment with a good outcome of one year after complete remission (5).

Although this child is in a stage IIIc with incomplete resection, prognosis remains good with chemotherapy and radiotherapy. The four-year relapse-free survival at this stage is 90 % with a 95 % 5 year-overall survival (9). In general, the role of pre-operative imaging in pediatric patients with typical signs and symptoms of appendicitis after evaluation by pediatric surgeon is almost 99 % sensitive for its diagnosis. Preoperative imaging does not only delay the subsequent management and rarely changes the course of treatment which is appendectomy in the majority of cases. However, pre-operative imaging has been found to be useful in situations where there are equivocal findings of appendicitis and obese patients with thick abdomen. The conservative treatment for the majority of patients with atypical findings and low likelihood of appendicitis is bowel rest and antibiotics. In the event of such cases, ultrasonography is the preferred choice due to its non-invasive nature and eliminates radiation risk in childhood (10). This case is important to highlight the uncommon presentation of Wilms' tumor which may mimic acute appendicitis.

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Competing Interests

The authors declare that they have no competing interest.

Informed Consent

Written informed consent was obtained from patient who participated in this study.

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