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Abdulrahaman Al Zhrani

Abdullah Baothman

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Abdulrahaman Al Zhrani
King Saud bin Abdul-Aziz University for Health Sciences, Jeddah, Saudi Arabia.
Abdullah Baothman
King Saud bin Abdul-Aziz University for Health Sciences, Jeddah, Saudi Arabia.
Princess Norah Oncology Center, King Abdul-Aziz Medical City-Jeddah, Saudi Arabia
Email: baothmanaa@ngha.med.sa

Abstract

It is common for Wilms tumor to invade the inferior vena cava and right atrium, but a massive pulmonary embolism is rare. This case report describes an 8-year-old male patient who presented to an outpatient clinic with mild pain associated with subjective fever for 1 week. A left renal mass with extension of an inferior vena cava thrombus was seen on abdominal computed tomography, suggesting a Wilms tumor. An exploratory laparotomy biopsy was scheduled, but not completed because the patient's health suddenly deteriorated, and he died despite high-quality cardiopulmonary resuscitation. A massive pulmonary embolism was found to cause a sudden and fatal cardiac arrest.

Keywords: *Nephroblastoma, pulmonary embolism, Wilms' tumor.*

Introduction

Nephroblastoma is the most common abdominal tumor in pediatrics (Shamberger et al., 2001; Davidoff, 2009). According to the Saudi Cancer Registry in 2016, 25 nephroblastoma cases were reported, representing an incidence of 4% (National Health Information Center, 2019). Wilms tumor (WT) commonly presented as an asymptomatic abdominal mass; it tends to occupy the renal vein, causing thrombi in the inferior vena cava (IVC), and it can progress further, entering the right atrium (Ceelen et al., 1997; Fukuda et al., 2019). Nearly 6% of Wilms' tumors have intravascular extensions, and about 82% of these are particular to the IVC. Intravascular extension of WT is more commonly seen in right-sided disease (Dome et al., 2020; Shamberger et al., 2001). Late discovery of intravascular extension can have fatal complications such as pulmonary embolism (PE) (Mohammadi, 2011; Zakowski, MF et al., 1990). The current study describes a case of nephroblastoma that was complicated by a massive fatal PE.

Case Report

An 8-year-old male patient, medically free, presented to the clinic complaining of localized left hypochondrial abdominal pain with a one-week history of subjective fever. There was no positive history for nausea, vomiting, shortness of breath, coughing, bleeding from orifices, nor hematuria. Regarding the family history, the patient's older sister was diagnosed with leukemia. On admission, the patient was vitally stable: blood pressure was 108/66 mm Hg, heart rate was 75 bpm, and respiration rate was 22 breaths per minute. Abdominal inspection revealed bruises and hematomas on the left side, and deep palpation revealed a mass in the left hypochondrial area. A complete blood count and an initial coagulation profile were normal. The von Willebrand factor was 167 IU per dL (reference range, 50-150 IU), D-dimer was high at 6.15 µg/mL (reference range 0.0-0.5 µg/mL), and fibrinogen level was high at 6.39 g/L (reference range 1.8-4 g/L). Ultrasound revealed a large left renal heterogeneous echogenic mass with a tumoral thrombus within the IVC, suggesting WT. Abdominal computed tomography showed progressive enlargement of the tumoral thrombus: At diagnosis, it was at the infrahepatic IVC (Figures 1,2), and a few days later it extended to the retrohepatic IVC, just below the confluence of the hepatic veins (Figure 3). An exploratory laparotomy biopsy revealed non-specific necrotic tissue with spindle cell proliferation likely representing fibrotic tissue with granulation adjacent to the necrotic tumor. The patient was scheduled for another biopsy to confirm the diagnosis, but he suddenly deteriorated and became hypoxemic. A bedside ultrasound revealed an unfilled IVC, signifying thrombus migration to the lungs. In spite of high-quality CPR, the patient remained unresponsive and died.

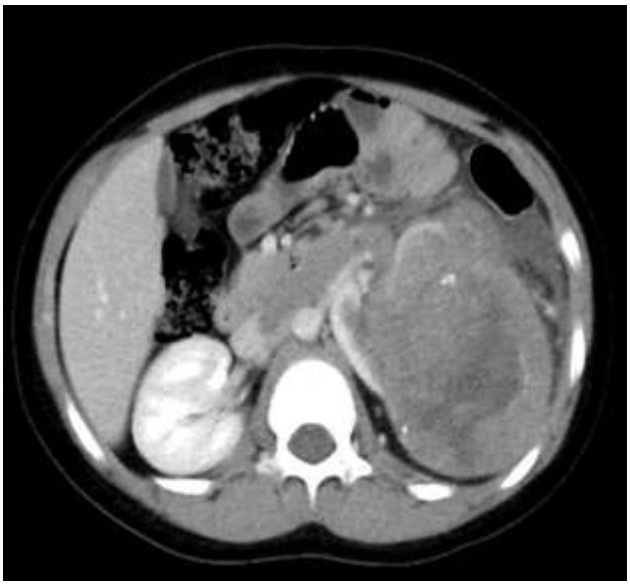

Figure 1: Axial CT scan of the abdomen at the time of diagnosis shows large left renal mass and left renal vein (curved arrows) thrombosis extends to the IVC (arrow).

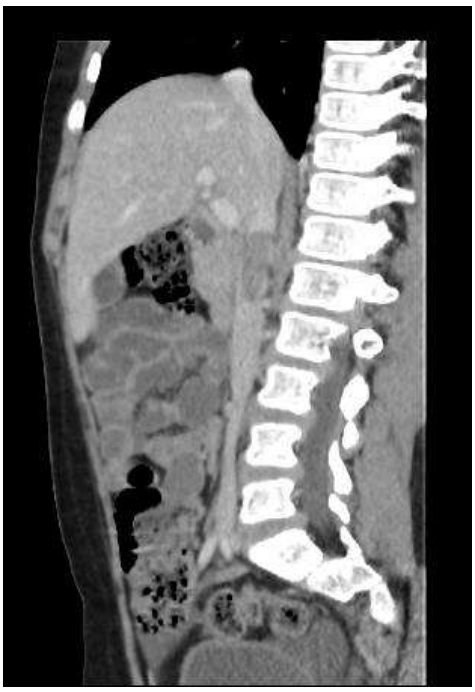

Figure 2: Sagittal CT scan of the abdomen at the time of diagnosis shows IVC thrombus at the infra hepatic level (arrow).

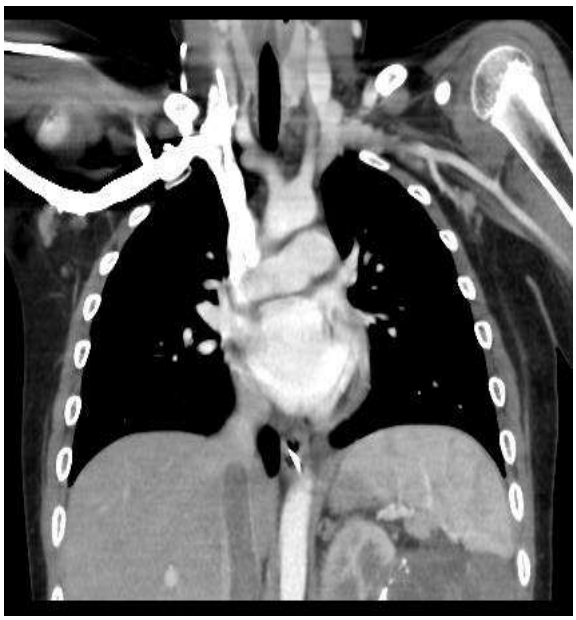

Figure 3: Coronal CT scan of the chest shows progression of the IVC thrombus (arrow) to the retro hepatic IVC below the confluence of the hepatic veins. The main pulmonary arteries are patent (dash arrows).

Discussion

Nephroblastoma, also known as WT, is an embryonic mass originating from epithelial and mesenchymal cells in the kidneys; approximately 6% of the cases show intravenous tumor extension, of which 86% involve the IVC, rarely extending to the right atrium (Mohammadi, 2011; Zakowski, MF et al., 1990). Massive PE as a complication of Wilms' tumor is quite rare, but fatal. Several studies suggest preoperative chemotherapy to decrease tumoral thrombus size, limiting surgical complications such as inadvertent tumor rupture and spillage (Ritchey Michael L. et al., 1988; Szavay et al., 2004). Shamberger et al showed that 165 of 2731 patients (6%) from the National Wilms Tumor Group (NWTSG4) had local intravascular infiltration of the tumor (Shamberger et al., 2001). Of those, 134 involved the IVC, and 31 (1%) involved the right atrium. Surgical complications occurred in 17% of those with IVC extensions and over 36% of those with atrial involvement. Of those receiving neoadjuvant chemotherapy, significant tumor regression was achieved in over 85% of those IVC extensions and 58% of those with atrial extensions; complications were rare (Shamberger et al., 2001).

The UKW3 trial reported that 59 of 730 patients (8.1%) with WT had IVC infiltration, and 10 of those (17%) had intracardial thrombus extension. Fifty-two (88%) received neoadjuvant chemotherapy and showed thrombotic regression. Sudden death due to PE was not reported (Lall et al., 2006), and associated typical histological background was not found in patients who suddenly died (Shamberger et al., 2001; Szavay et al., 2004). Intravascular WT was not found to affect survival rate (Mushtaq et al., 1996; Ritchey Michael L. et al., 1988; Shamberger et al., 2001; Szavay et al., 2004; Xu et al., 2019). Neoadjuvant chemotherapy is associated with shorter

operative times and shorter hospital stays compared to surgical intervention. It also decreased the need for CPB, thus reducing CPB-associated complications such as bleeding, thrombosis, and infection (Giannoulia-Karadana et al., 2000; Xu et al., 2019). However, CPB is favored in some cases where urgent surgery is necessary, particularly when cardiac tumoral extension is found or the patient is at high risk of sudden cardiorespiratory instability (Giannoulia-Karadana et al., 2000). A multidisciplinary team including all related specialties should be formed to discuss risks versus benefits and treatment options. Temporary IVC filters are always a choice when intravascular invasion can avoid venous clots migrating to the lung. Empiric anticoagulation in patients with intravascular tumor extensions is not helpful; it potentially increases the risk of tumor thrombus spread.

However, anticoagulation should be considered if a plain thrombus is found in addition to a tumor thrombus or if distal thrombosis develops without collateral venous pathway complicated with deep venous thrombosis (Quencer et al., 2017; Ratajczyk et al., 2018). Continuously monitoring vital signs will detect early signs of PE such as desaturation and tachycardia. Doppler ultrasonography should be used as well. In our case, the thrombus in the IVC could have been poorly attached to the vessel walls, and with increasing size, it dislodged, migrated to the lungs, and caused a massive PE. Pathologic examinations showed fibrosis of the thrombus was absent in patients undergoing primary nephrectomies; however, the incidence was 43% among those treated with preoperative chemotherapy. Vessel wall adherence was noted in 43.7% of those treated with primary surgery versus 61.7% of those treated with preoperative therapy (Shamberger et al., 2001).

Conclusion

Despite good overall prognoses for children with WTs, distinct cases continue to have poor overall survival rates, a high risk for late effects, or severe complications such as PE. Serial computed tomography scans of the abdomen and the pelvis to monitor tumor and thrombus size can promote early identification of thrombus migration and shorten the time needed to formulate an appropriate management plan. Closer attention to cases presenting with intravascular tumor extensions, which can have fatal outcomes such as PEs, is necessary. Older patients with large primary tumors and extensive vascular invasions, unless invading the right atrium, must be identified. The best management for those might be preoperative chemotherapy and delayed tumor resection to minimize the risk of surgical complications.

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