Pediatrics
Wilms Tumor with dilated hypertensive cardiomyopathy, acute myocarditis, pulmonary edema, and heart failure

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ABSTRACT

An 11-month-old boy with unilateral Wilms tumor had an unusual presentation, with dilated hypertensive cardiomyopathy, acute myocarditis, pulmonary edema, and congestive heart failure secondary to the Wilms tumor and requiring intensive care. After surgery, all of the findings were normalized. According to the literature review there were reported previously in only seven cases with dilated cardiomyopathy. Wilms tumor should be included in the differential diagnosis of any child with dilated cardiomyopathy and an abdominal mass, regardless of the presence of hypertension, especially less than one year old. Also, vasoactive mediators including renin should be evaluated in the case of hypertension.

Introduction

Wilms tumor is accounting for approximately 90% of all pediatric tumors of the kidney, and 6% of all pediatric tumors.1 The typical presentation is an asymptomatic abdominal mass.1 Other clinical manifestations are hematuria, hypertension, dilated cardiomyopathy, and congestive heart failure (CHF). According to the literature review, although dilated cardiomyopathy and CHF are very unusual presentations there were reported previously in only seven cases.2–5

An 11-month-old boy presented herein, the first in Korea, had an unusual presentation, with dilated hypertensive cardiomyopathy, acute myocarditis, pulmonary edema, and CHF secondary to the Wilms tumor.

Case presentation

An 11-month-old boy presented with a history of irritability, fever, vomiting, diarrhea, oral candidiasis, tachycardia, and respiratory distress has been transferred to our hospital. He had a mild fever, dyspnea, tachycardia, and hypertension (systolic 140–150, diastolic blood pressure 95–100 mmHg). On the physical examination, there was a firm, non-tender mass at the left upper abdomen. He had oral candidiasis 1 week ago, high fever 4 days ago, vomiting and diarrhea every 3–4 hours 3 days ago, and dyspnea/tachycardia 1 day ago.

His serum troponin I, B-type natriuretic peptide, myoglobin, CK and CK-MB levels were elevated. Serum PT time, PT INR, aPTT, AST, LDH, creatinine, and uric acid levels were also increased. Serum Ca, and Na levels were decreased. Arterial blood gas analysis demonstrated increased PH, decreased pCO2, pO2, pHCO3, and increased base excess.

A chest radiograph revealed mild cardiomegaly, diffusely increased opacity in both lungs, and bilateral pleural effusion (Fig. 1 A). Computed tomography of the chest demonstrated consolidation and ground-glass opacity in both lungs accompanied by bilateral pleural effusion and cardiomegaly suggesting to cardiogenic pulmonary edema (Fig. 1 B, C). Echocardiogram and echocardiography demonstrated a sinus tachycardia, dilated hypokinetic left ventricle with an ejection fraction of 29.4%, grade II mitral regurgitation, and trivial aortic regurgitation. Ultrasound and computed tomography of the abdomen revealed a left solid renal mass (6.7 × 7.8 × 8.1 cm) without internal necrotic portion or calcification, a significant amount of ascites, and bilateral pleural effusion (Fig. 1 D, E, F). There was no distant metastasis on the metastasis work-up.

The patient received emergency intensive care, including inotropic...
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agent, antihypertensive drugs and diuretics, in intensive care unit. Despite the intensive treatment, the patient’s condition did not improve, the radical nephrectomy was performed after 6 days. There was no thrombus of renal vein in preoperative radiologic and intraoperative finding. After surgery, all of the preoperative pulmonary edema, pleural effusion, blood pressure, clinical symptoms, and blood chemistry studies were normalized (Fig. 2 A, B, C), and echocardiogram at postoperative 10 days demonstrated improved ventricular contractility, with an ejection fraction of 39% (before surgery 29.3%). The inotropic agent could be stopped 2 days after surgery. The histology examination demonstrated favorable histology composing of blastemal (50%), stromal (25%), epithelial component (25%). There were a negative adjacent organ, resection margin, Gerota’s fascia, lymph node invasion, and no multipolar atypical mitotic figures/anaplasia, but the positive lymph-vascular invasion and renal sinus invasion by tumor (Fig. 2 D, E). The final stage of Wilms tumor was stage 2 by the Children’s Oncology Group staging system. The adjuvant chemotherapy without radiotherapy was continued in his hometown (New Zealand).

Discussion

Wilms tumor occurs primarily between the ages of 2 and 5 years, which is rare during the first year of life.1–3 Interestingly, four out of eight cases including this case were less than 1 year old [Table 1]. Also, four cases showed hyperreninemia with hypertension and CHF.1–3 Agarwala et al.5 suggested that the CHF was caused by severe hypertension secondary to hyperreninemia. Regrettfully, we did not check vasoactive mediators such renin in this case. However, Parry et al.2 and Trebo et al.4 reported two cases and one case of Wilms tumor with dilated cardiomyopathy and absence of hyperreninemia, respectively. They postulated that vasoactive mediators other than renin being produced from the Wilms tumor as the cause of the dilated cardiomyopathy. It seemed related to a vasoactive mediator’s secretion such as renin, catecholamine, or other vasoactive mediators.3

All seven previously reported cases completely resolved CHF/hypertension from 2 months to 3 years after nephrectomy and chemotherapy [Table 1]. Based on clinical outcomes in eight cases including this case, the decrease in mass by chemotherapy or nephrectomy resulted in a hemodynamic stabilization, and then by the progressive recovery of myocardial function.

Conclusion

In conclusion, Wilms tumor should be included in the differential diagnosis of any child with dilated cardiomyopathy and an abdominal mass, regardless of the presence of hypertension, especially less than one year old. Also, hyperreninemia and/or serum catecholamine levels should be evaluated in the case of hypertension. Lastly Wilms tumor...
Fig. 2. Chest radiographs, computed tomography of chest after radical nephrectomy, and gross and histologic findings.
A: chest x-ray demonstrated no cardiomegaly, pleural effusion, and pulmonary infiltration in both lungs.
B (axial view), C (coronal view): After radical nephrectomy 7 days, there was very much improved consolidation and ground-glass opacity in both lungs, and bilateral pleural effusion and cardiomegaly.
D: Cross-sectional gross findings demonstrated tumor thrombus is unremarkable, grossly on opening along the renal vein. The superior to the inferior aspect of the kidney is remarkable for an irregular mass, which has lobulated edges with dimensions of 9.6 × 6.5 × 5.8 cm.
E: Tumor showed favorable triphasic histology composing of blastemal (50%), stromal (25%), epithelial component (25%) (H&E × 40).

Table 1
Clinical characteristics, laboratory findings, treatment and outcome in patients with Wilms’ tumor and dilated cardiomyopathy

| Ref.          | Age (Sex) | Primary tumor Size: cm | Stage | Clinical presentations | Laboratory findings | Treatment                                      | Outcomes                                      |
|--------------|-----------|-------------------------|-------|-----------------------|---------------------|------------------------------------------------|-----------------------------------------------|
| Stine et al., 1986 | 9 mo (M)  | Bilateral kidneys (right 11 × 11 × 9) (left 13 × 13 × 9) | V     | Abdominal distension, CHF, HT | Increased renin     | Chemotherapy, partial nephrectomy (no CHF/HT after 3 mo) |                                              |
| Agarwala et al., 1997 | 2 yr (F)  | Right kidney (10 × 15 × 8) | II    | CHF, DCM, pulmonary edema, HT | Increased renin     | Right nephrectomy, chemotherapy (Alleviated CHF/no HT at discharge and no CHF/HT 1 yr after surgery) |                                              |
| Trebo et al., 2003 | 2.5 yr (F) | Right kidney (NA) (NA) | IV    | DCM, no HT | Normal renin | Right nephrectomy, chemotherapy, radiotherapy (Alleviated CHF at early postoperative period and resolved CHF/HT after 3 yr) | Right nephrectomy (Alleviated CHF/HT at 12 days after surgery and resolved CHF/HT after 2 mo) |
| Chalavon et al., 2017 | 7 mo (F)  | Right kidney (8.5 × 10 × 8) | I     | CHF, DCM, Pulmonary edema, no HT | Increased renin     | Right nephrectomy, chemotherapy (Alleviated CHF/HT after surgery and resolved CHF/HT after 10 mo) |                                              |
| Sethasathien et al., 2019 | 3 yr (M) | Left kidney (9.7 × 9.7 × 9.5) | IV    | CHF, DCM, HT | Increased renin     | Left nephrectomy, chemotherapy, radiotherapy (Resolved CHF/HT after 3 mo) |                                              |
| Parry et al., 2019 | 2 yr (M)  | Right kidney | I     | Abdominal distension, | Increased renin     | Right nephrectomy, chemotherapy (Resolved CHF/HT, NA time) |                                              |
| Present case | 11 mo (M) | Left kidney (6.7 × 7.8 × 8.1) | II    | DCM, no HT | /angiotension II | Right nephrectomy, chemotherapy (×1) (Alleviated CHF/no HT after surgery, 1 wk) |                                              |

CHF: Congestive heart failure, DCM: Dilated cardiomyopathy, HT: Hypertension, NA: not available.
with dilated cardiomyopathy is life-threatening. Therefore prompt oncologic therapies along with intensive care are necessary.

CRediT authorship contribution statement

Jae Min Chung: Supervision, Writing - review & editing. Sang Don Lee: Conceptualization, Data curation, Writing - original draft, Writing - review & editing.

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