Metastatic Wilms tumor and dilated cardiomyopathy

Sir,

Wilms tumor (WT) is the most common pediatric kidney tumor and typically presents as an asymptomatic abdominal mass.[1] WT-associated cardiomyopathy is a rare complication that is thought to be either secondary to mechanical compression of the renal artery or the production of renin by the tumor.[2] This report describes a unique case of metastatic WT-associated cardiomyopathy with treatment complicated by pulsus alternans.

A 13-year-old female presented with abdominal pain and weight loss for 1 year. Physical examination demonstrated a tender distended abdomen, a palpable abdominal mass (heart rate: 86, blood pressure: 167/92, respiratory rate: 18, O_2 saturation 97%). Computed tomography revealed a left renal mass with multiple pulmonary nodules. Follow-up magnetic resonance imaging demonstrated a 2.4 cm × 12.9 cm × 14.3 cm mass in the left renal fossa [Figure 1a]. The patient underwent a left radical nephrectomy and removal of the retroperitoneal mass with pathology demonstrating Stage IV WT with favorable histology.

The patient was started on adjuvant chemotherapy. Before adding doxorubicin to her regimen, an echocardiogram demonstrated a reduced left ventricular ejection fraction (LVEF) of 31% and a global longitudinal strain of 13.4% [Figure 1b]. Doxorubicin was excluded from her treatment regimen to avoid cardiotoxicity. The patient was started on oral heart failure therapy, which was complicated by pulsus alternans after the patient was switched from carvedilol to metoprolol [Figure 1c]. The patient was changed back to carvedilol with no further evidence of pulsus alternans.

After 6 weeks of chemotherapy, her lung metastases were reduced but not eliminated. At this time, her cardiomyopathy had not improved (LVEF: 30%). Chest radiation was considered but not pursued to avoid the risk of additional cardiotoxicity. It was decided that she would receive an additional 6 weeks of chemotherapy followed by a 12-week taper. This led to complete resolution of pulmonary metastasis and modest improvement of her LVEF (43%). She continued to be asymptomatic, and after completion of adjuvant chemotherapy, an echocardiogram demonstrated an LVEF of 61.8% and global longitudinal strain of 19.2% [Figure 1b].

The association between dilated cardiomyopathy and WT has only been reported in 6 previous cases. After resection of the WT, improvement of cardiac function despite receiving some potentially cardiotoxic chemotherapy drugs would suggest the WT is responsible for the cardiomyopathy.[3] The etiology of LV dysfunction has been speculated to be caused by prolonged hypertension,[2,4] however, our patient's blood pressure was largely normal after beginning a heart failure regimen but continued to have LV dysfunction. Reports of patients having WT-associated cardiomyopathy without hypertension were secondary to vasoactive mediators other than renin and catecholamines. Our hypothesis is that our patient's LV dysfunction was likely a combination of both mechanisms. Pulsus alternans is also a rare phenomenon our patient experienced. The beat-to-beat variation in systolic flow typically indicates left ventricular dysfunction and is associated with a poor prognosis.[4] This phenomenon was observed when the patient was switched from carvedilol to metoprolol, which has not been documented previously.

Only one prior case of WT-associated cardiomyopathy had metastases at diagnosis. This patient's metastatic disease resolved after 3 months of chemotherapy and had an LVEF increase of 29%–50%. After surgical resection and adjuvant chemoradiation, the patient's LVEF increased to 58%.[5] In our patient, surgical resection had little effect on cardiac function. She required a prolonged course of adjuvant therapy to resolve her

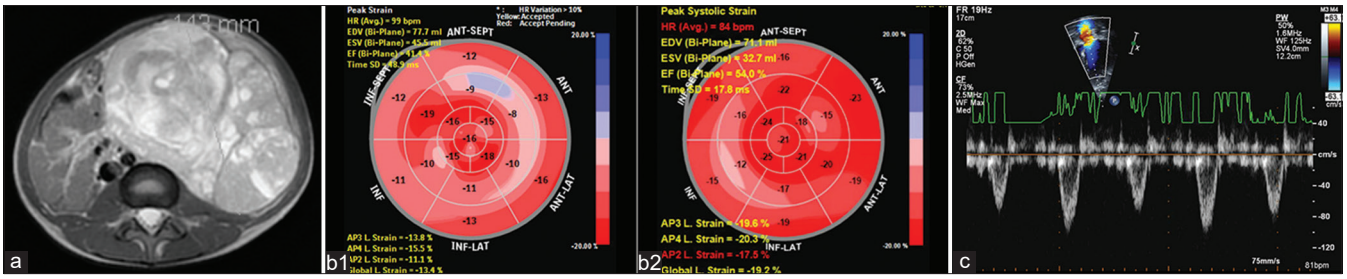

Figure 1: (a) Abdominal magnetic resonance imaging showing the mass (2.4 cm × 12.9 cm × 14.3 cm). (b) Cardiac strain demonstrating global longitudinal strain of 13.4% (1). One year after shows a global longitudinal strain of 19.2% (2). (c) Transthoracic echocardiogram shows beat variations in the left ventricular outflow tract

metastases and full recovery of her LVEF (61.8%). The timing of LVEF improvement would suggest that the remaining metastatic disease was responsible for the slow LVEF recovery.

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Conflicts of interest

There are no conflicts of interest.

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