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Case Report:

Wilms Tumour with Intracardiac Extension.

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Abstract: Wilms tumour or nephroblastoma is the most common renal tumour of in children. It accounts for 6% of all pediatric tumours and is the second most frequent intrabdominal solid organ tumour in children. Inferior vena cava (IVC) involvement by Wilms tumour occurs in 4-10% of patients and right atrium thrombus extension in less than 1%. Using a multidisciplinary approach, overall survival is excellent in Wilms tumour. We are presenting this case because Wilms tumour with right atrial thrombus is relatively rare and continues to remain a challenge for treating physicians.

Keywords: Wilms tumour; Intracardiac extension; Nephroblastoma

Case Report

A 6 year old girl was admitted with history of fever and generalized weakness for 3 weeks and abdominal distension and hematuria for 5 days.

On examination, she had tachycardia and tachypnea. Her blood pressure was 117/ 80 mm Hg. A short systolic murmur was heard at the lower left sternal border and mitral area. Breath sounds were reduced in both lung bases. The abdomen was distended and a firm mass was present in the right lumbar region.

Hemogram was normal. Renal and liver function tests were normal.

Chest radiograph showed mild cardiomegaly with obliteration of left cardiophrenic angle. Abdominal ultrasound revealed a right renal mass with tumour thrombus extending into right renal vein and inferior venal cava.

CT abdomen showed a large heterogeneously enhancing soft tissue mass arising from the right renal fossa – 11cmX10cmX11cm with paraaortic lymphadenopathy. Increased luminal caliber of the inferior vena cava (2.5cm) was seen, which was not enhancing on contrast, extending up to the level of the right atrium.

Echocardiogram confirmed a large mass in the right atrium causing obstruction to the tricuspid valve.

A diagnosis of Wilms tumour arising from the right kidney with extensive tumour thrombus involving the inferior vena cava up to the right atrium was made.

The child received 4 weeks of neoadjuvant chemotherapy. A cardiac team removed the intravascular tumour and a surgical oncology team performed the nephrectomy. Recovery was uneventful.

Histopathological examination of the resected tumour was consistent with Wilms tumour - favourable histology - arising from the right kidney. Tumour showed chemotherapy induced necrosis (80%). Residual tumour showed epithelial and mesenchymal components. Lumen of the hilar blood vessel showed no residual tumour, Ureteric cut margin, renal capsule, perinephric adipose tissue and suprarenal gland were free of tumour.

Post operative chest x ray was normal. Repeat echocardiogram revealed no residual tumour, trivial mitral and triscuspid regurgitation with normal biventricular function.

The child was then given external beam radiotherapy to the right flank (10G/16Fractions) followed by adjuvant chemotherapy according to NWTS IV protocol.

Reassessment at the end of treatment was within normal limits. She has been on follow up for 30 months.

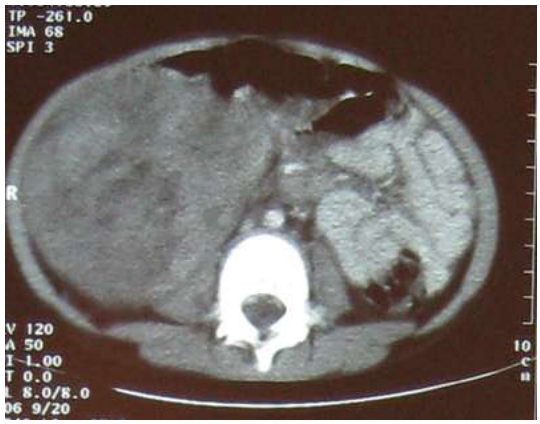

Fig 1: CT abdomen showing a large heterogeneously enhancing soft tissue mass arising from the right renal fossa with paraaortic lymphadenopathy.

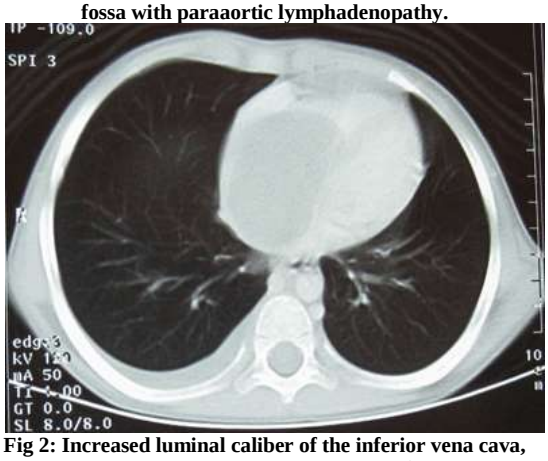

Fig 2: Increased luminal caliber of the inferior vena cava, not enhancing, extending upto the level of the right atrium

Discussion

Wilm's tumour is the most common pediatric renal tumour.[1] Despite the successes in treatment of Wilm's tumour, challenges remain – one of them being the treatment of intra atrial thrombus.[2]

There are two primary treatment strategies to treating a child with Wilms tumour. The first utilizes upfront nephrectomy followed by chemotherapy, the second employs pre-nephrectomy chemotherapy.[3] To date no randomized studies have been conducted to guide definitive therapy in a patient with caval or atrial extension. The benefits for preoperative chemotherapy include possibility of resolution of the thrombus, thereby decreasing the need for cardiopulmonary bypass surgery. The drawbacks include tumour emboli, tumour progression, and the recognized increased difficulty of removing a tumour from the venal cava or atria following chemotherapy.[4]

Three large series provide insight to managing a child with vascular extension. In the SIOP 93-01 GPOH study, 33 of 1151 patients had vascular extension. In nine, there was extension into the atrium. Twenty-nine received preoperative chemotherapy. Twenty (69%) responded to chemotherapy including one of those with extension into the atria. Nine required cardiopulmonary bypass to remove the tumour. There were no surgical deaths.[5]

In the UKW3 trial 59 patients had vascular extension with 10 extending into the atria. Fifty-two received preoperative chemotherapy with 35 (67%) responding. 5/52 (10%) died due to uncontrolled bleeding during surgery.[6]

The National Wilms Tumour Study Group IV trial reported outcomes on 134 patients with vena cava or vascular extension. 69 received preoperative chemotherapy. Seventy-one percent had some response to therapy. Five cases developed tumour embolism and progression was noted in three patients. Although the overall complications were similar in those receiving preoperative chemotherapy and those who underwent primary resection, the majority of children responded to chemotherapy. The compelling response rate of the thrombus to chemotherapy has lead the Children's Oncology Group to recommend preoperative chemotherapy for all patients with tumour extension above the hepatic cava.[7]

In conclusion, preoperative chemotherapy is warranted but tumour embolism may occur. Although the tumour may shrink, morbidity and mortality are significant as noted in the UKW3 and NWTS4 reports. These cases require a multidisciplinary approach to ensure a good outcome.

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