CASE REPORT
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Bilateral Wilms Tumor in Female Child of Nine Months

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ABSTRACT

Among malignant neoplastic lesion of childhood, Wilms tumor is the most common malignant and approximately five to seven percent of these tumour present as bilateral disease. This bilateral tumour usually affects more often female patient. At present, bilateral disease is managed with pre-operative chemotherapy at the time of diagnosis followed by nephron-sparing surgery. The recent literatures revealed no case with bilateral involvement. For this reason, we discussed this case with bilateral Wilms tumour (BWT) in a nine months old female patient as a rare one. Management of bilateral Wilms tumor needs experienced radiologist who are familiar with imaging of nephrogenic cells, histopathologist who critically analyze cell line, pediatric surgeon dedicated for nephron-sparing surgery and oncologist with clear knowledge to prevent recurrence.

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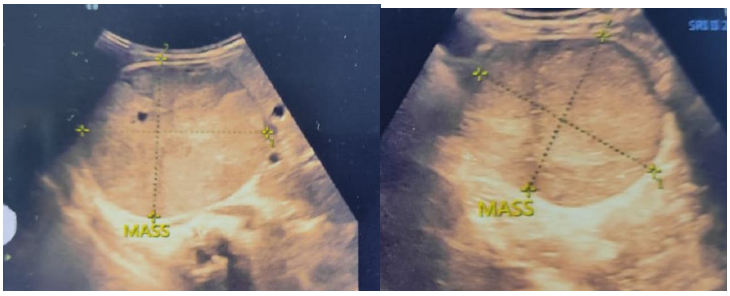
INTRODUCTION

Wilms tumour is one of the commonest malignant renal tumour of childhood. It was estimated in USA that eight cases of this tumour per million children less than fifteen years of age per year. Recent management includes unilateral nephrectomy with systemic chemotherapy and associated ionizing radiation.[1,2] Due to improved surgical techniques, prognosis of this lethal tumour improved. The overall survival rate of localized disease is currently greater than 90%. About five to seven percent of tumour present with bilateral disease either synchronously or metachronously.[3] Management of a child with bilateral Wilms tumour (BWT) is very challenging. Preservation of the maximum amount of renal parenchyma is needed to prevent renal failure, but complete resection is required to optimize the chances for cure of the malignancy. This tumour arises from mesodermal precursors of the renal parenchyma, increasingly gene loci are being implicated on chromosome 11 (WT1: 11p13 and WT2: 11p15) as well as WTX on chromosome X, B-catenin on chromosome 3 or TP53 on chromosome 17.[4] On gross inspection, these tumours are usually well-circumscribed or macrolobulated. Haemorrhage and central necrosis are common findings. Abdominal X-ray reveals a large soft tissue opacity displacing bowel. Ultrasound is a very much helpful procedure and will be the primary investigation of choice. It is helpful to localize the mass to the kidney and also distinguish from other causes of renal masses. Doppler

examination can be performed to examine the renal vein and Inferior vena cava (IVC) to assess for the presence of tumour thrombus.[5,6] Computed tomography (CT) scan is useful for characterizing renal tumours in children. CT scan done in portal venous phase is sufficient to characterize Wilms tumour. These tumours are heterogeneous soft-tissue density masses with infrequent areas of calcification and fat-density regions. Enhancement is also patchy and allows for better delineation of the relationship between the mass and kidney. Ten to twenty percent (10-20%) of cases have lung metastasis at the time of diagnosis.[7,8] Where MRI is available, it is the investigation of choice for staging since it does not involve ionizing radiation. It is also one of the accurate modality in assessing for IVC involvement where protocols have been optimized.[1] These tumours appear heterogeneous on all sequences and frequently contain blood products.[1-3] Due to late metastasis in the bones, its scans are not consider as routine investigation. F-18 FDG PET-CT is increasingly used as a problem-solving tool and to distinguish scar tissue from residual active tumour.[6,8] In case of unilateral Wilms tumour, surgery or preoperative chemotherapy followed by surgery is done. In local extensive disease or bilateral involvement, chemotherapy is choice of treatment. Radiotherapy has a limited role, but may be employed in cases of peritoneal spread or incomplete resection.[8] Cure is now possible in ~90% of cases. Recurrence is seen both within the tumour bed, as well as distally within the lungs or liver.[6,8] Response depends on rhabdomyoblastic differentiation, even though it is a favourable histological subtype.[7] Advanced functional imaging using apparent diffusion coefficient is now applied which has the potential to make distinction of tumour. Patients with bilateral disease need to maintain maximal renal function to ensure longevity requiring advanced imaging and surgical techniques.[2,8,9]

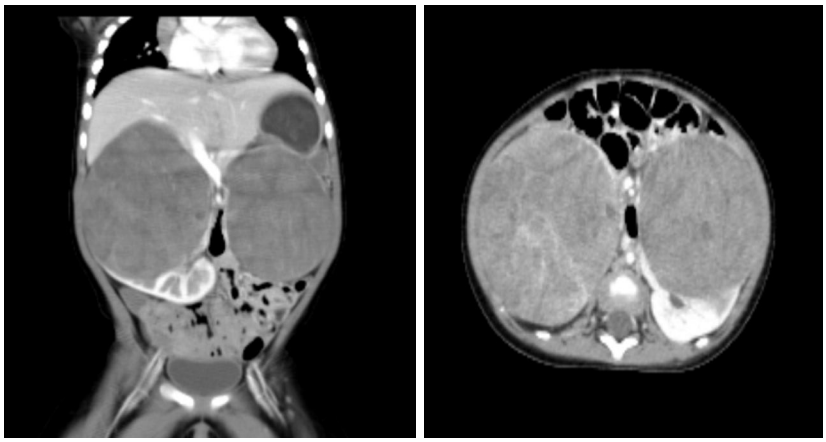
The Case

A nine months old female child hailing from Sirajganj, Bangladesh attended in the Department of paediatric outdoor of Rafatullah Community Hospital (TMC and RCH), Bogura, Bangladesh with huge abdominal distension and flank mass. On examination patient was not anaemic and hypertensive. Per abdominal examination found upper abdominal mass but its lower margin was not reachable. Ultrasonography (USG) of whole abdomen revealed bilateral renal mass measures about 71x63 mm on right and about 80x63 mm on left side (Figure1). Sonographic differentials were bilateral adrenal mass, favoring medulloblastoma. USG guided FNAC from both kidneys revealed round blue cell tumour suggesting Wilms tumour. Unfortunately, slides were lost from patient's parent during travelling from Sirajganj to Bogura. Only cytological report was found. Contrast CT scan of whole abdomen showed lobulated mildly enhancing mass arising from upper-mid pole of right kidney (about 78x69 mm) and anterior aspect of mid pole of left kidney (about 85x67 mm). Adrenal glands and liver were separated from the lesion (Figure 2). No pulmonary lesion was found. From above findings, it was concluded that this was a case of bilateral Wilms tumour. We consulted with oncologist and peadiatric surgeon for academic interest.



(A) (B)

Figure 1: Ultrasonography showing right (A) and left (B) renal solid mass



(A) (B)

Figure 2: (A) Contrast enhance coronal reformatted CT scan of whole abdomen showing mildly enhancing bilateral renal mass. (B) Contrast enhances axial CT scan of whole abdomen showing mildly enhancing mass involving both kidneys.

DISCUSSION

Wilms tumours are largely heterogeneous solid masses which displace adjacent structures. Occasionally they may be mostly cystic or have calcifications.[5] In this case; tumours were solid with no local invasion or evidence of metastasis. It typically occurs in early childhood (1 to 11 years) with peak incidence between three and four years of age[1] but the age of this case was nine months. There is no recognized gender predilection, but in female the presentation is a bit later.[3] Clinical presentation is typically with a painless upper quadrant abdominal mass. Haematuria is seen in ~20% of cases[3] and pain is uncommon. On examination, hypertension due to excessive renin production is found in up to 25% of patients.[1] This case was presented with huge abdominal distension with flank mass. Patient was non-hypertensive and not anaemic. Metastasis are most commonly to lung (85%), liver and local lymph nodes.[7] Similar to renal cell carcinoma, thrombus into the renal vein, IVC and right atrium are also characteristic of advanced disease. In this case, no metastasis is visualized in lung fields or evidence of renal venous thrombosis. Unilateral Wilms tumours are treated by a combination of nephrectomy and chemotherapy. Occasionally, chemotherapy can be administered prior to surgery to downstage the tumour.[5] This is especially useful when tumours are bilateral. As per consultation with oncologist, chemotherapy was advised for this case.

CONCLUSION

Wilms tumour is a common childhood malignancy. Bilateral involvement of this tumour is very rare and treatment is challenging. So, collaboration of concerned specialists is required for its suitable management.

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Conflict of interest: Not declared.

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