Unorthodox Display of Nephroblastoma with Abdominal Pain and Distention

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INTRODUCTION

Wilms' tumour is a renal malignancy of childhood with incidence between 2 to 5 years of age group accounting approximately 95% cases of childhood renal malignancies. We report a case of 11-year-old female child presented with flank pain and abdominal lump. Abdominal CT (Computed Tomography) scan showed a right renal mass suggestive of malignancy. Wilms' tumour was confirmed by CT scan when physicians think that child may have a tumour, imaging tests like ultrasound, X-rays, CT scan and MRI (Magnetic Resonance Imaging) are used to consider differential diagnosis. These tests are necessary and will enable the doctors not only locate but also to diagnose the type and size of tumour and in addition to this they also help to explain how much the tumour has extended in the kidney or other parts of body and in accordingly therapy is done.

Nephroblastoma or Wilms' tumor accounts for 6% of all malignancies and is common among children with renal cancer. It mostly occurs in children less than five years of age and 90% of cases are diagnosed before age of three. 1 Wilms tumour is associated with majority of anomalies and syndromes which involve genitourinary tract including renal ectopia, renal hypoplasia, ureteral duplication, hypospadias, cryptorchidism and disorder of sexual development. 2 Peak incidence is 2 - 5 years of age group. Early diagnosis and adequate treatment can easily help the patient with benefiting results. The diagnosis can be made on various imaging modalities. Radiography and ultrasound is the basic early investigations carried out in a rural setup like our hospital followed by a contrast enhanced CT scan. Here in this case report, we present a case of 11-year-old female with Wilms' tumour, which is an unorthodox age of presentation, its ultrasonographic and CT scan features help in the surgical management.

PRESENTATION OF CASE

An 11-year-old female child presented in emergency department of Acharya Vinoba Bhave Rural Hospital, Sawangi, Meghe with chief complains of abdominal distention and pain in abdomen since 4 days. As narrated by father the child was alright till 4 days back after which she complained of pain in abdomen, which was insidious in onset and non-specific in nature, it was associated with distention of abdomen. Child had history of hospitalization 3 months back for fever, cough, cold where reports were suggestive of raised WBC count. There was no significant family history and child was immunised till date. She attained all milestones as per age. On physical examination heart rate was 100 beats per minute with respiratory rate of 24 per minute.
The child's skin looked normal, well-nourished. Blood pressure was normal and ranged from 108 / 70 to 120 / 75 mmHg. The child weighed 30 kilograms and her height was 143 cm. Heart sounds were normal, S1 and S2 present with no evidence of murmur. The gastrointestinal tract and genitourinary tract were normal. The child bowel habits were normal. On auscultation, breath sounds were decreased on right side. On per abdomen examination, abdomen was tense, tender and flank fullness were present. There was evidence of a mass present extending from right hypochondrium and extending across the umbilicus. The initial investigation was radiographs and ultrasound examination. The chest radiograph revealed homogenous opacity in the right hemithorax with obliteration of the right costophrenic and right diaphragmatic line suggestive of massive pleural effusion. And X-ray abdomen showed soft tissue density mass on right side of abdomen causing extrinsic compression to the bowel and no calcification was seen in the mass. On ultrasound, right kidney shows a heterogeneous lesion measuring (20 x 15 x 13 cms) arising from upper and middle pole with multiple tiny cystic spaces with tiny echogenic foci without posterior acoustic shadowing possibility of tiny calcifications. The colour Doppler of the lesion showed vascularity only along the walls. No central vascularity was noted in the mass. On contrast enhanced computed tomography of abdomen-pelvis and thorax there is large well-defined multi-lobulated heterogeneously enhancing mass lesion with non-enhancing necrotic areas within arising from the posterior aspect of mid and upper of the right kidney extending from T8 to L3 measuring approximately 22 x 16 x 14 cms in size. The lesion is displacing the liver antero-superiorly and towards the left. Rest of the renal parenchyma is displaced antero-inferiorly and laterally by the mass causing stretching of renal vessels anteriorly without any evidence of invasion of renal vein. The IVC (Inferior Vena Cava) is compressed and displaced towards left. Cortical phase, nephrogenic phase and parenchymal phase appear normal with normal contrast excretion.

Figure 1. Gray Scale Ultrasound Showing a Heterogeneous Mass with Many Cystic Spaces within. It was Arising from the Mid and Upper Pole of Right Kidney

Figure 2. Gray Scale Ultrasound Showing a Heterogeneous Mass with the Lower Pole of Kidney Being Spared

Figure 3. Colour Doppler Ultrasound Showing That the Mass is Not Very Vascular and Renal Vein is Supplying the Lower Pole of the Kidney

Figure 4. CT Scan Arterial Phase Showing a Large Well Defined Multi-Lobulated Heterogeneously Enhancing Mass Lesion

Figure 5. CT Scan Arterial Phase Showing Normal Left Kidney. Note Massive Pleural Effusion on the Side of Affected Kidney - Right

**DISCUSSION**

Wilms' tumour is most common primary renal tumour in children. Wilms' tumour is comparable or simulates teratomas...
as they have a large diversity in its cell types and stage of differentiation. In clinical practice monophasic and biphasic types found but it is observed that most cases show triphasic appearance with blastemal, stromal and epithelial elements within.

Wilm’s tumour arises from abnormal proliferation of metanephric blastema, although very rarely it is found to affect the adult kidneys as well in our case, the patient age was 11 year old. Interestingly only 3% of Wilm’s tumour occurs in adults and in such cases the age of patients ranges from 16 to 62 years (with a median age of 25.4 years). Radiographically, Wilm’s tumour when compared to the normal parenchyma can present as an inhomogeneous mass with low density and mild contrast enhancement on post-contrast scans. Some of the masses may present as complex, cystic masses with solid components and calcifications and so can mimic renal tumours other than Wilm’s.

Abnormal renal development is hypothesized in pathogenesis of Wilm’s tumour. It has been reported that WT1 gene, a type of tumour suppressor gene, located at 11p13 encodes zinc finger transcription factor which plays an important role in renal and gonadal development, generally wild type allele is somatically inactivated in tumors with constitutional WT1 mutations or deletions. Wilm’s tumour can also be associated with other syndromes like WAGR (Wilms-Aniridia-Genitourinary-Mental Retardation) syndrome, Denys-Drash syndrome (WT1 mutation), Beckwith-Wiedemann syndrome is associated with at 11p15 and other congenital anomalies. So lung computed tomography scan, ultrasound scan involving genitourinary tract analysis and gene testing should be carried out.

Symptoms of Wilm’s tumour may vary, and mostly children come with complaint of abdominal pain and a palpable mass per abdomen. Patients with no symptoms pose a challenge to the doctor for correct diagnosis. In our case patient initially came with abdominal pain, distention and a palpable mass; after various imaging techniques and by careful examination on ultrasound and computed tomography she was finally diagnosed as having Wilm’s tumour. Abdominal imaging techniques including ultrasound, computed tomography scans and magnetic resonance imaging can be applied for evaluation of abdominal mass, as it is the most common clinical presentation of paediatric renal malignancy. Wilm’s tumour can be differentiated from other types of cancers, such as clear cell sarcoma, rhabdoid tumour, congenital mesoblastic nephroma, renal cell carcinoma, and renal medullary carcinoma on the basis of histology. Surgery to remove tumour is main treatment for Wilm’s tumour. Adjuvant chemotherapy and radiation therapy can be considered according to stage of cancer. Treatment for Wilm’s tumour is of great success in paediatric oncology, with long-term survival rate of > 90% when disease was localized to the abdomen and > 70% when there is associated metastasis of the disease. If child is having raised blood pressure, it can be considered another sign of a kidney tumour and so proper monitoring of child during hospitalization should be done if needed. Blood and urine samples should be routinely collected for testing. Although it is rare, Wilm’s tumour can run in families. When a child is diagnosed as Wilm’s tumour other investigations and tests can be done on parents to evaluate for a familial origin of such tumours.

CONCLUSIONS

Whenever child presents with complaint of abdominal pain and palpable mass per abdomen Wilm’s tumour should always be kept in mind. A complete medical history needs to be taken to see all symptoms and to know how long these symptoms have existed. When physicians are looking at a possibility the child may have a tumour, various imaging tests needs to be considered. Ultrasound, X-rays, MRI are one of the routinely done investigations needed to get a differential diagnosis. These tests are of immense importance and will assist doctors to locate the lesion, diagnose the type and know the size of tumour and in addition they also help to explain how much the tumour has extended in kidney or other parts of the body with this knowledge therapy is planned. Follow up of cases and finding out prognosis of the tumour is also done by the imaging techniques. Age is considered very important when we are approaching such patients because older children have higher risk of misleading diagnosis. More attention should be paid in children who are asymptomatic with anaemia, abdominal pain and abdominal mass because it can be sign of Wilm’s tumour.

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