Case Report: Wilms' Tumor

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Background

Wilms' tumor is an embryonal tumor of childhood occurring in 1 out of 1,00,000 children less than 15 years of age. An incidental case was found in the Paediatric surgery ward of Dhaka Medical College Hospital on 20th June 2012. A 3 years old girl complained of right sided painless abdominal mass for one month with occasional hematuria for 15 days. Detailed history and investigation confirmed the diagnosis as Wilms' tumor. In developing countries most of such cases present at advanced stages. Having no scope for specific treatment, multimodal approach i.e., neoadjuvant chemotherapy followed by surgical excision has been the best choice to reduce the morbidity and mortality of these cases.

Wilms' tumor or Nephroblastoma is a cancer of kidneys that typically occurs in children. Its common name is an eponym, referring to Dr. Max Wilms, the German surgeon (1867–1918) who first described this kind of a tumor. It is the most common renal malignancy of childhood containing metanephric blastema, stromal and epithelial derivatives. It tends to be encapsulated and vascularized tumor that does not cross the midline of the abdomen. It usually metastesize to the lungs and liver. It is usually unilateral and presents with microscopic haematuria , malaise, weight loss, anaemia and sometimes with varicocele or thromboembolic disorder in the heart. A rupture of Wilms' tumor puts the patient at risk of hemorrhage and peritoneal dissemination of the tumor. In such cases, surgical intervention by a surgeon who is experienced in the removal of such a fragile tumor is imperative.

Case

A girl aged 3 years, hailing from Zurain, Dhaka was admitted into Dhaka Medical College Hospital with the complaints of right sided abdominal mass for one month with occasional hematuria for 15 days. The mass had been gradually increasing in size which was painless. There was no history of pain or burning sensation during micturation. She had no history of fever, anorexia, weight loss, vomiting & cough. On examination she was apparently healthy, mildly anaemic, afebrile and normotensive. The mass was palpable occupying right lumbar, right hypochondriac and umbilical region, measuring 14 cm x 11 cm. It was non tender, surface was smooth, firm in consistency and its movement with respiration could not be elicited. Her bladder and bowel habits were normal. USG of whole abdomen and contrast CT Scan of abdomen suggested renal carcinoma. Diagnosis was confirmed by USG guided FNAC and histopathology. She had undergone preoperative neoadjuvant chemotherapy according to SIOP protocol after consulting with the radiotherapy department. All the doses of chemotherapy were administered in the ward, so that she might not miss the sequential chemotherapy. After that right sided nephroureterectomy was done followed by postoperative chemotherapy.

Results

She attended the first follow up visit after 3 months and was uneventful.

Conclusions

Mutations of the WT1 gene on chromosome 11 p 13 are observed in approximately 20% of Wilms' tumors. But rest of the cases have not identified genetic association or any other significant risk factors. As it is usually painless and relatively asymptomatic, it is difficult to notice for the children on their own. Moreover due to poverty and ignorance many children in developing countries present with advanced stages of Wilms' tumor with higher chance of metastasis. As a result they suffer from a bad prognosis. Except preconceptional counseling in case of positive family history, there is no specific way of prevention so far. That's why early diagnosis and effective intervention are direly needed. The multimodal treatment especially neoadjuvant chemotherapy may reduce the extension of the disease that may help further surgical excision – total or partial. This demands for multidisciplinary collaboration of Paediatric surgery, Pathology, Paediatric oncology and Radiotherapy department along with social councelling. Only then we can reduce the morbidity and mortality of the disease.

The possibility of a Wilms' tumor should be considered when a children presents with hematuria and a renal mass. Although the treatment modality is yet to be standardized, the outcome for patients diagnosed with Wilms' tumor is steadily improving.