

Case Report

Wilms' tumor in 3 years old female patient - A case report

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Abstract

Nephroblastoma or Wilms' tumor is an embryonic tumor derived from primitive renal epithelial and mesenchymal components. It is the most common abdominal malignant tumour of young children. Overall, Wilms' tumor incidence is 7.8 cases per million children. Peak age of incidence is 2 to 3 years of age, or 99% occurring less than six years of age. Here, we are presenting a case of 3 years old female patient with diagnosis of Nephroblastoma.

Key words

Nephroblastoma, Wilms' tumor, Abdominal malignant tumor, Young children.

Introduction

Nephroblastoma or Wilms' tumor is common among children with renal cancer. Overall, Wilms' tumor incidence is 7.8 cases per million children [1]. Peak age of incidence is 2 to 3 years of age, or 99% occurring less than six years of age [2-4]. Approximately 50-85% of children with Wilms' tumor present with a palpable abdominal mass, usually found incidentally [5, 6]. Here, we are presenting a case of 3 years old female patient with diagnosis of Nephroblastoma where we are able to find and document the typical features of Nephroblastoma.

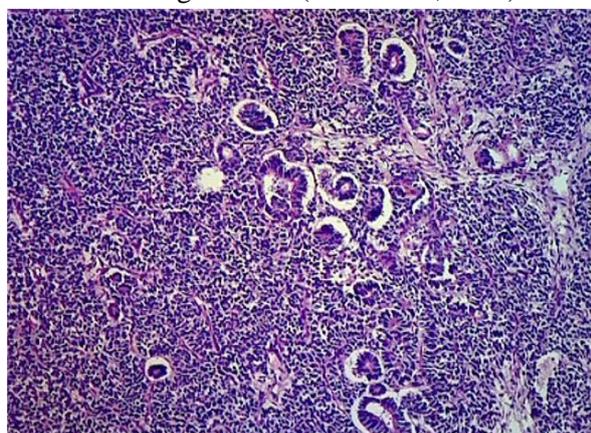
Case report

A 3 year old female patient came to our hospital with swelling in left side of abdomen for 1 month noticed by mother. Swelling was gradually progressive in nature and associated with hypertension. There was no history of fever, abdominal pain, vomiting, hematuria and constipation as per her mother, the historian. Our patient's past medical history was unremarkable. There was no recurrent sinus, middle-ear, urinary tract infections or any congenital problems. Birth history revealed an uncomplicated full-term vaginal delivery.

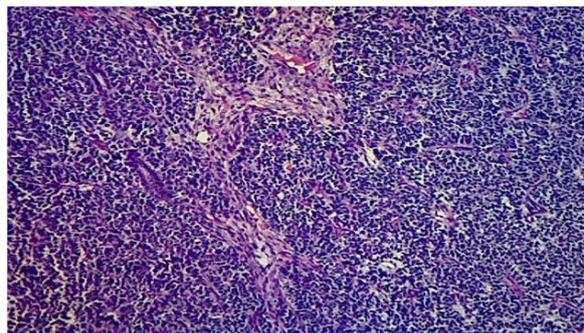
Her development was normal, immunizations were up-to-date, and her health had been good since the swelling had been noticed. There was no family history of neoplasms, gastrointestinal problems, seizure, congenital heart problems, hemoglobinopathy or other major illnesses.

On physical examination our patient appeared sick. Her axillary temperature was 98.6 degrees Fahrenheit, pulse regular 110 beats per minute, and respirations 28 breaths per minute. Blood pressure was 130/90 mmHg. On abdominal exam a mass of 5× 4 cm with regularly shaped margins was palpated with light depth and verified with percussion in the left upper quadrant and did not cross the midline. The child denied pain during the exam, but guarded and panted during palpation. All other systemic examination was normal. Laboratory data revealed a slightly low hematocrit and haemoglobin (Hb-8.3gm/dl) with microcytic, hypochromic red cells. All other serological and biochemical parameters were within normal limit. The ultrasound revealed a Heterogeneous echogenic mass of size 7.5x 5.3 cm seen in left renal fossa suggestive of Wilms' tumor. The CT scan showed a solid mass with cystic areas in the lower and upper quadrant arising from the left kidney. Nephrectomy specimen was sent for histopathological examination and diagnosis of Wilms' tumor Stage I was given (**Photomicrograph - 1, 2, 3, 4**).

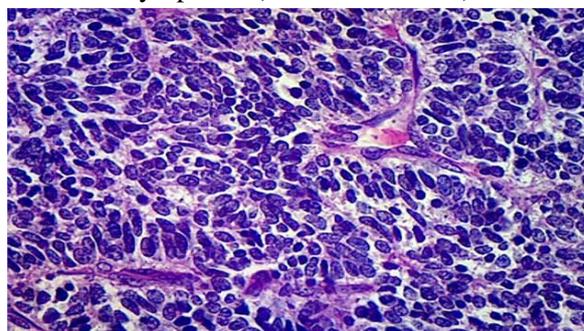
Photograph – 1: showed a combination of blastema, stroma, epithelial tubular formations, and immature glomeruli (H&E stain, 10 X).



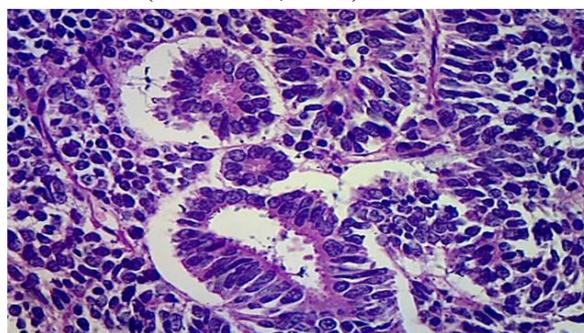
Photograph – 2: showed extremely cellular *blastematous* areas (H&E stain, 10 X).



Photograph – 3: showed *blastematous* areas composed of small round-to-oval primitive cells and scant cytoplasm (H&E stain, 40 X).



Photograph – 4: showed immature tubular formations (H&E stain, 40 X).



Discussion

Nephroblastoma or Wilms' tumor is an embryonic tumor derived from primitive renal epithelial and mesenchymal components. It is the most common abdominal malignant tumour of young children. It accounted for 6% of all malignancies [7]. It almost always occurs in children less than five years of age -90% of cases are diagnosed before the age of three [7, 8], the peak incidence is in the age range of 2-5 years [9].

Wilms' tumor is highly associated with other congenital anomalies such as aniridia, hemihypertrophy, hypospadias, and kidney malformations such as fused, horseshoe or polycystic kidneys in as many as 12% of patients [10, 11]. Thirty-three percent of children with aniridia develop Wilms' tumor [10]. Our patient did not possess any of these anomalies.

Wilms' tumors may be inherited in an autosomal dominant fashion with the dysfunction or deletion of the Wilms' tumor suppression gene along chromosome 11 possibly causing nephrogenic rests [12]. Wilms' tumor can grow for a long time without any characteristic symptoms, causing only fever, abdominal pain, nausea, or vomiting, which is the reason why it is often discovered accidentally [13]. In this case abdominal swelling was noticed accidentally by patient's mother.

Wilms' tumor arises anywhere in the kidney as embryologic precursors to renal cells. These cells mimic normal development of the kidney and consist of three components: blastema, epithelium, and stroma [1]. The *blastematous* areas are extremely cellular and composed of small round-to-oval primitive cells; the cytoplasm is usually very scanty, but sometimes is more abundant and exhibits an oncocytoid appearance. The *epithelial* component is characterized by the formation of embryonic tubular (and sometimes glomerular) structures that closely recapitulate the appearance of normal developing metanephric tubules.

Treatment is chemotherapy and nephrectomy for single kidney involvement with excision of tumor cells and preservation of normal cell functioning for bilateral kidney involvement [1]. Combination chemotherapy and radiation are given as a pretreatment in high-risk cases such as the bilateral renal involvement, diffuse anaplasia, or metastases [1]. Our patient had taken treatment in the form of nephrectomy and post-operative chemotherapy.

Conclusion

Screening for Wilms' tumor is very important for children who have syndromes or birth defects known to be linked to this disease. For these children physical exams by a specialist and ultrasound exams on a regular basis are recommended (for example, about every 3 or 4 months at least until the age of 8) to find any kidney tumours when they are still small and have not yet spread to other organs.

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