

A 32 DAYS OLD BABY WITH NEPHROBLASTOMA- A CASE REPORT

Md. Mokhles Uddin¹, Faizunnesa Bhuiya², Sadia Sharmin³, Swapan Bandyopadhyaa⁴,
Md. Khorshed Alam⁵

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Introduction

Nephroblastoma synonym(s)-nephroma, renal adenocarcinoma, renal cancer, Wilms' tumor and hypernephroma. Nephroblastoma is an embryonal paediatric tumor of the kidney which also be seen rarely in adults¹. Microscopically it is composed of a mixture of cellular elements (blastema, stromal and epithelial) in varying proportions. The proportion of the different components has prognostic significance. It is most common solid abdominal tumor in childhood and usually appears as a painless mass in an otherwise well child. Frequently the paediatrician on routine examination or the parents while playing with the child, will notice a large, firm mass below the rib cage on either the right or left side of the abdomen. The peak incidence of nephroblastoma is between the second and fifth year of life².

In the United States, two-thirds of cases are diagnosed before five years of age, and 95 percent before 10 years of age. In patients with unilateral involvement, the median age at diagnosis is 43 months in girls and 37 months in boys³. Children with bilateral disease are diagnosed at an earlier age (median age, girls at 31 months and boys at 24 months)³. Patients with associated congenital anomalies, such as aniridia or genitourinary abnormalities, are also diagnosed at an earlier age.

The incidence of nephroblastoma is slightly higher in girls than in boys. The female to male

ratio is 1.1 to 1 for unilateral tumor and 1.7 for bilateral involvement³.

The risk of developing nephroblastoma varies among ethnic groups. In the United States, the risk of nephroblastoma is greater in African-American children than in non-Hispanic white children. The rate of nephroblastoma is lower in Asian compared to Caucasian children living in the same geographic location⁴.

Case History:

A 32 days old boy named Shaheen, child of Kohinor Begum hailing from 16/b, P.O: Bashabo, Thana: Shabujbag, Dist: Dhaka, referred to Radiation and oncology Department of Dhaka Medical College Hospital (DMCH) from Paediatric Surgery unit with the complaints of (1) Swelling in the abdomen since birth (2) Fever for 10 days (3) Passage of blood mixed urine for 5 days.

According to the statement of the patient's mother, the baby was alright except a bit swollen in the abdomen since birth. Then the swelling was gradually increased in size. She also complains of fever which was low grade, continuous in nature for 10 days. She also noticed occasional passage of blood mixed urine for last 5 days.

For the above complaints she consulted with a local general physician. On physical examination the physician got a palpable abdominal mass in the right lumbar region. Then he advised to do Ultrasonography of the whole abdomen on March'08. Ultrasonography report revealed a mass (10x8 cms in size) in the abdomen occupying the right lumbar and umbilical region. Then she was referred to the Paediatric Surgery Unit of Dhaka Medical College Hospital (DMCH).

On 20.03.08 the patient was admitted in Paediatric Surgery Unit of Dhaka Medical

1. Professor, Radiotherapy, Department of Chittagong Medical College
2. HMO, Radiation and Oncology, Department of Dhaka Medical College
3. HMO, Radiation and Oncology, Department of Dhaka Medical College
4. Registrar, Radiation and Oncology, Department of Dhaka Medical College
5. Assistant Professor, Radiation Oncology, NICRH, Mohakhali, Dhaka

Address of Correspondence: Prof. Md. Mokhles Uddin Professor, Radiotherapy, Department of Chittagong Medical College

College Hospital. FNAC was done from the right abdominal mass and report revealed nephroblastoma. Then patient was referred to Radiation and Oncology department of DMCH on 29.03.08. On general examination, patient was ill looking, toxic, irritable, mildly anaemic, nonicteric and there was no lymphadenopathy. On systemic examination, a lump was palpated on the abdomen occupying the right lumbar and umbilical region which was firm in consistency, nontender and approximately 8x6 cms. On Investigation, urine R/M/E showed puscell 3 to 5/HPF, RBC 0 -2/HPF, epithelial cell 3 to 5/HPF and liver function test showed serum bilirubin 1.6 mg/dl, serum alkaline phosphatase 42 U/L and SGPT 20 U/L.

Patient was planned for neoadjuvant chemotherapy as per schedule: Injection Dactinomycin 15mcg/kg, IV D1-D3 at first and third week and Injection Vincristine 1.5mg/m²-D1 at first, second, third and fourth week. After chemotherapy the swelling decreased in size that was clinically by 50%. Again ultrasonography was done and report revealed a mass in right lumbar region that was 5x3 cms. Fever subsided, than the patient was referred to paediatric surgery department of Dhaka Medical College Hospital for surgical management Since than patient is missing.

Discussion

Nephroblastoma is the most common malignant renal tumor of childhood⁴. It occurs with an annual incidence of 7 cases per million children younger than 15 years of age. Approximately 450 new cases are diagnosed each year in North America⁴. It is difficult to define the exact picture of nephroblastoma in our country due to lack of both population based and hospital based cancer registry. Although the peak incidence is between 3 to 4 years of age, in that case patient was only 32 days old baby, which is extremely rare.

Nephroblastoma arise as sporadic or hereditary tumors, or in the genetic disorders. Most tumors are solitary lesions, multifocal in a single kidney in 12% and bilateral in 7%⁵. The clinical syndromes associated with nephroblastoma include WAGR syndrome (Wilms' tumor, aniridia, genitourinary malformations, mental

retardation), Denysh Dash syndrome (pseudohermaphroditism, mesangial sclerosis, renal failure and Wilms' tumor) and overgrowth syndromes like Beckwith-wiedeman syndrome (somatic gigantism, omphalocele, macroglossia, genitourinary abnormalities, ear creases, hypoglycemia, hemihypertrophy, and a predisposition to nephroblastoma and other malignancies) and Simpson Golabi Behmel syndrome and other syndromes include Perlman syndrome and Sotos syndrome⁶. In that case, tumor arise as sporadic origin and solitary lesion in a single kidney.

The classic presentation of nephroblastoma is that of a healthy child in whom abdominal swelling is discovered by the child's mother, or a physician during physical examination. A smooth, firm, nontender mass is in the lumbar region in one side or bilaterally. Gross haematuria present in as much as 25% cases. The child may be hypertensive or have nonspecific symptoms, such as malaise or fever⁵. In our case, mass was also discovered by patient's mother which was smooth, nontender, firm in consistency in one side occupying the right lumbar and umbilical region and patient also presents with low grade continuous fever and occasional haematuria.

In diagnostic purpose Plain X-ray of the abdomen may play a great role to differentiate neuroblastoma and nephroblastoma by demonstrating the calcifications, which occur in 60% to 70% of neuroblastoma but in only 5% to 10% of nephroblastoma. Urinary vinyl mandelic acid also done to differentiate of these two. Now a days intravenous pyelography is replaced by ultrasonography and CT scan which is very useful to diagnose. Abdominal CT scans can demonstrate gross extrarenal spread, lymphnode involvement, liver metastases and the status of opposite kidney. MRI is superior to CT scan to identifying renal origin and vascular extension of the tumor. Plain chest radiography is also essential because asymptomatic pulmonary metastases are common. CT scan is more sensitive in detecting pulmonary metastases⁸.

In this case, ultrasonography of the whole abdomen revealed a mass in the abdomen,

urinary vinyl mandelic acid was negative, chest X-ray revealed normal findings, CT scan and MRI was not possible due to poor financial condition. FNAC of the mass was done which revealed nephroblastoma.

The diagnosis is usually made before surgery and confirmed by laparotomy. The contralateral kidney should be palpated and visualized to rule out bilateral nephroblastoma before nephrectomy. The recommended chemotherapy and radiotherapy depends upon stage and histologic subtype⁷.

In our case, surgery was not possible due to huge mass and patient's poor general condition. So, he was supposed to receive neoadjuvant chemotherapy to reduce the bulk of the tumor to a resectable status. The patient received neoadjuvant chemotherapy as per standard schedule and the tumour regress from 10x -8 cm to 5 x 3 cm but unfortunately patient has been absconded and result could not be evaluated.

Conclusion:

Nephroblastoma is a highly curable neoplasm. The prognosis of children has considerably improved from a very high mortality rate at the beginning of the 20th century to the current cure rate of over 90%(8). 90% nephroblastoma is a paediatric tumor and most of the cases occur within 2 to 15 years of age, within 1 year it is rare. So far we have known, this is an extremely rare case of 32 days old baby who was borne with nephroblastoma that is why it is being reported.

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