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Case Report
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Congenital neuroblastoma presented with cholestasis and huge hepatomegaly

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Abstract

This case report is about a 28-day-old neonate that had abdominal distention and massive hepatomegaly since 15th days of birth. First, he was normal, and he had not any sign and symptom of sepsis. At admission, he was about twelve days, he had a rapid progressive presentation of sepsis and unfortunately, he passed away. His liver biopsies manifested the neuroblastoma.

Keywords: Neuroblastoma, Neonatal malignancy, Abdominal distention

Background

Neuroblastomas are embryonal cancers of the peripheral sympathetic nervous system with heterogeneous clinical presentation and course, ranging from tumors that undergo spontaneous regression to very aggressive tumors unresponsive to very intensive multimodal therapy. Causes of most cases remain unknown, and although significant advances have been made in the treatment of children with these tumors, the outcomes for aggressive forms of neuroblastoma remain poor.

Neuroblastoma is the most common extracranial solid tumor in children and the most commonly diagnosed malignancy in infants. Approximately 600 new cases are diagnosed each year in the United States, accounting for 8-10% of childhood malignancies and one third of cancers in infants. Neuroblastoma accounts for >15% of the mortality from cancer in children. Median age of children at diagnosis of neuroblastoma is 22 mo, and 90% of cases are diagnosed by 5 yr of age. Incidence is slightly higher in boys and in whites.

Presenting concerns

In our case report, we had a 28 days boy that he was term and delivered by C-section with birthweight of 3200 grams. Since 15th day of birth, his mother noticed about the abnormal abdominal distention. In the 28th day of birth, he was admitted to Rasool Akram Teaching Hospital. He had not any sign of sepsis such as fever, nausea, vomiting, diarrhea and poor feeding. He was breastfed by his mother.

Clinical finding

In family history, he was the second child of his parents. The first was healthy. His parents had not contagious diseases. There was not any past medical history in this family. In the first day of admission, the general condition was good except for some jaundice and some tachypnea. His weight was 4300 gr, head circumference: 37.5 cm, and length: 54cm. In physical exam, respiratory rate was 65, in the heart he had a systolic murmur about 2/6. The sound of lung was clear; the abdomen was distended, the liver was extended to the pelvic and was firm, but the spleen was normal. Capput medoosa was seen. Scrotal edema in two sides was seen. There was no edema in the limbs. In echocardiography, moderate MR was reported. Sonography of abdomen showed the massive hepatomegaly, more enhancement in echo of liver. Ophthalmology consultation was normal. There was an abnormal coagulation test. Although, according to gastrointestinal

Table 1. Laboratory data

WBC	8.9	17	23.3	17.8
HB	8	11.1	11.6	8.7
HCT	25.2	35.3	32.4	27.4
MCV	100	94.4	89	93.5
PLT	209	316	308	258
PT	30.6	23.4	25.4	12
PTT	>120	65	66	53
INR	4.2	4.13	1	1
BS	69	75	60	
BUN	4	4	14	21
CR	0.4	0.5	0.6	0.5
TG	57			
Cholesterol	53			
NA	125	125	123	108
K	3.3	3.6	6.9	4.6
CA	9.6	9		
Bilirubin total	22.5			
Bilirubin direct	10.5			
Ferritin	895			
AFP	>1000			
GGT	183			
AST	87			
ALT	22			
Total PRO	4.7			
ALB	3.4			
Ammoniac	112.4			
Lactate	19.3			
Torch study	negative			

consult, liver biopsy was needed. All the metabolic tests were normal. Feritin was normal, Torch study was done and it was normal. During the hospitalization, he was icteric and was more and more weak. Fever and leukocytosis were appeared after 5 days of admission. After infection consultation, the appropriate anti-microbial agents were started. Hemoglobin decreased at 7th day of admission and respiratory distress was appeared. The patient transferred to the PICU and intubated. After injection of neoseven, under the cardiopulmonary monitoring, biopsy of liver with success was done. Two days after biopsy, his respiratory distress were exaggerated. Unfortunately, he had cardiac arrest in 12th day of admission, and did not respond to CPR and expired.

Diagnostic focuses and assessment

In our differential diagnosis, glycogen storage type 4, Tyrosinemia, Niemanpick type C, Alpha one antitrypsin deficiency, CMV infection, neonatal iron storage disease were the most important. However, in the specimen received in formalin from the liver, we had found the filiform fragment of creamy soft tissue measuring 1*1*0.1. Section showed neoplastic tissue composed of sheets of small neoplastic cells characterized by hyperchromatic high N/C ratio nuclei with molding effect and scant pale eosinophilic cytoplasm. In some foci tumoral cells form rosette formation. IHC stained sections are positive for chromogranin in some tumoral cells and negative for CK, CD99, and CEA.

Diagnosis

Small round cell tumor suggestive for neuroblastoma with negative CK in tumoral cell, negative in CEA, negative in CD99 and positive chromogranin in tumoral cell.

Discussion

We had found that presentation of Neuroblastoma is vary from patient to patient. Neuroblastoma may develop at any site of sympathetic nervous system tissue. About half of neuroblastoma tumors arise in the adrenal glands, and most of them originate in the paraspinal sympathetic ganglia. Metastatic spread, which is more common in children older than 1 yr of age at diagnosis, occurs via local invasion or distant hematogenous or lymphatic routes. Most common sites of metastasis are the regional or distant lymph nodes, long bones and skull, bone marrow, liver, and skin. Lung and brain metastases are rare, occurring in >3% of cases. Prognosis of children with neuroblastoma varies with the histologic of the tumor. When the tumor extended to bilateral, N-Myc is positive, bone marrow metastases prognosis is very poor. We should be aware of any related sign and symptom in children and in neonates especially in abdominal distention.

Conflicts of interest: None declared.

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