

## Case Report

# Massive hepatomegaly in a neonate: an unusual cause

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### ABSTRACT

A female neonate presented with progressive abdominal distension. Liver was palpable, 11 cm below costal margin and a spleen of 10 cm. USG abdomen revealed massive hepatomegaly with multiple nodules in liver, kidney size and texture being normal. Contrast enhanced CT scan of the abdomen was suggestive of enlarged multinodular liver with splenomegaly. Liver biopsy revealed replacement of liver architecture by small rounded cells, which were positive for synaptophysin and S100 stains suggestive of neuroblastoma. A repeat abdominal radiological evaluation revealed a small 2 x 1 cm mass in left suprarenal area. Neuroblastoma stage 4S presenting as hepatomegaly is rare.

**Keywords:** Hepatomegaly, Neonate, Neuroblastoma 4S

### INTRODUCTION

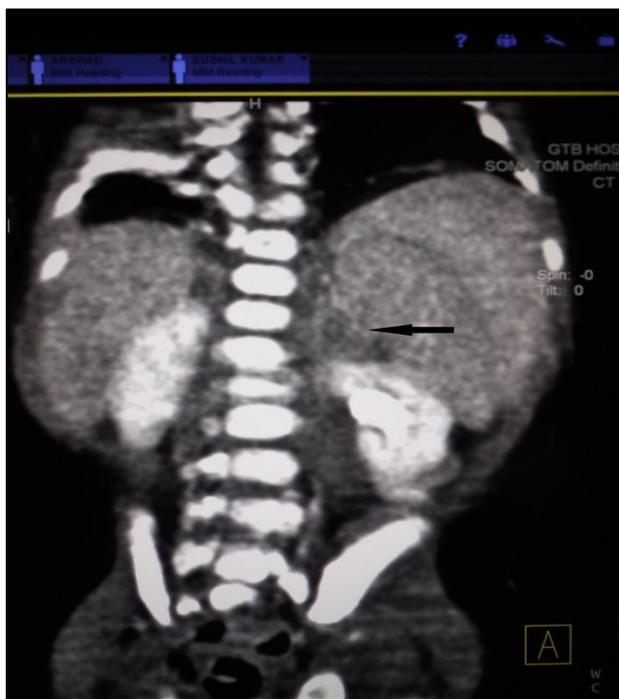
Hepatomegaly in infancy has varied causes. Neuroblastoma presenting as hepatomegaly is rare. Neuroblastoma usually presents as an abdominal mass arising from adrenal glands or posterior mediastinum.<sup>1</sup> About 7-10% cases present with a small primary tumor with metastasis to skin, bone marrow or liver; classified as stage 4S.<sup>2</sup>

### CASE REPORT

A term infant, 3 kg at birth, presented at 1 month with progressive distension of abdomen since 10 days of age. The distension was gradual, painless, not associated with vomiting, feed intolerance, and jaundice or bleeding from any site. On examination, the child weighed 5 kg, there was no pallor, jaundice or cataracts. Abdomen was grossly distended, liver was markedly enlarged, and 11 cm below costal margin with a liver span of 20cm and a

spleen of 10 cm. Cardiovascular and respiratory system did not reveal any abnormality. Haemoglobin was 9.5 gm/dl, total leukocyte count 9000/microlitre, serum bilirubin of 0.6 mg/dl, SGOT of 12U/L and SGPT of 24 U/L. USG abdomen revealed massive hepatomegaly with multiple nodules in liver, kidney size and texture being normal. Serology for intrauterine infections and radiographs of pelvis, spine and skull were also normal.

Contrast enhanced CT scan of the abdomen was suggestive of enlarged multinodular liver with splenomegaly. Liver biopsy revealed replacement of liver architecture by small rounded cells, which were positive for synaptophysin and S100 stains suggestive of neuroblastoma. A repeat abdominal radiological evaluation revealed a small 2 x 1 cm mass in left suprarenal area. The diagnosis was confirmed to be as neuroblastoma stage 4S and referred to the oncology department for initiation of chemotherapy with oncovin, cisplatin and etoposide.



**Figure 1: Primary tumor in the left suprarenal region.**

## DISCUSSION

Neuroblastoma is embryonic malignancy of the sympathetic nervous system arising from the pluripotent sympathetic cells, ie. neuroblasts. The primary tumor can be in the adrenals, paraspinal region or in the posterior mediastinum.<sup>1</sup>

Neuroblastoma is the most common extra cranial tumor in children.<sup>2</sup> According to International Neuroblastoma Staging System (INSS), stage 4S is defined as localized primitive tumor (stage 1 or 2) with metastasis to liver, skin or bone marrow, with most children presenting in infancy.<sup>3</sup> It is one of the rare causes of hepatomegaly in infancy as in our child. A case of stage 4S neuroblastoma with bilateral adrenal tumors has been reported at day 1 of life.<sup>2</sup> In another case report a 6 week old infant presented with enlarging liver. Diagnosis was not possible on initial radiological evaluation though a repeat ultrasound examination revealed cyst in the superior pole of kidney.<sup>4</sup> Our case had a similar course of events as the diagnosis was missed on initial clinical and radiological evaluation. At times a neonate may have abdominal mass detected in the prenatal period.<sup>4</sup>

Several treatment modalities have been tried in stage 4S neuroblastoma. Low to moderate dose chemotherapy has been found useful in some cases specially under 2 months of age with massive hepatomegaly and organ compromise resulting in rapid regression of the tumor.<sup>5</sup> Surgical resection is usually of not much benefit except some cases of complicated massive hepatomegaly can be considered for abdominal decompression surgery.<sup>6</sup>

Hepatic artery embolization has been tried as a safe and feasible treatment modality in neuroblastoma cases with massive hepatomegaly.<sup>7</sup>

In spite of being a metastatic tumor it has a very good progression with majority of the tumors undergoing spontaneous regression. However, in some infants, especially those less than 2 months may suffer rapid disease progression with a mortality of 10-20%.<sup>8</sup>

Possibility of neuroblastoma should be considered in infants presenting as hepatomegaly. Appropriate radiological investigation should be performed with emphasis to find any primary lesion in suprarenal area.

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