A 1-year-old boy with hepatosplenomegaly

History:

A 1-year-old boy was referred from Chiang Rai Hospital due to hepatosplenomegaly. At the age of 7 months, it was incidentally found during a medical attention of respiratory illness. There had been no history of jaundice, bleeding tendency, or fever. Stool and urine color were normal. Initially, he was investigated at the secondary care setting, in which an ultrasonography showed markedly enlarged liver and spleen; the VDRL and anti-HIV were both negative. During follow up, gradual progression of the hepatosplenomegaly was observed.

Past history:

He was born at the GA of 39 weeks with a birth weight of 4300 g. The patient is the second child in this family. At birth, there was a history of hypoglycemia and jaundice. PDA was detected, which subsequently closed. No apparent family history suggestive of significant genetic diseases. Development: Normal for age; No delayed or regression of the development documented

Feeding: Appropriate solid diet 3 meals/day with formula supplement

Physical examination:

General appearance- Thai boy, active, no dysmorphic facies
Body weight 9.3 kg, height 73 cm
Vital signs: T 37 C, PR 120/min, RR 28/min, BP 100/80 mmHg
HEENT: mild pale, no jaundice, no generalized lymphadenopathy
Heart: regular rhythm, no murmur
Lungs: normal breath sound, no adventitious sound
Abdomen: Moderate to marked distention, No superficial vein dilatation,
No abnormal mass, Not tender, normal bowel sound, Ascites-negative
Liver: Size 4 cm below RCM, soft consistency, smooth surface, sharp edge, not tender
Spleen: Size 9 cm below LCM, firm to hard consistency, not tender
No signs of chronic liver disease
Ext: no pitting edema, no rash
CNS: intact

Basic investigations:

CBC : Hb 11.6 g/dl, Hct 37%, WBC 10,100/cumm, N19, L68, Mono5, Plt 101,000/cumm
PBS: NCNC, MCV 74.3%, MCH 23.3%, MCHC 31.4%
UA : pH 5, sp gr. 1.015, protein-negative, sugar-negative, rbc-0, wbc- 0-1/HPF
BUN 11 mg/dl, Cr 0.3 mg/dl, Na 137 mEq/L, K 3.8 mEq/L, Cl 103 mEq/L, CO2CP 19 mEq/L
LFT: Albumin/globulin 4/2.1 g/dl, AP 299 IU/L, cholesterol 144 mg/dl, AST/ALT 53/61 IU/L, TB/DB 0.33/0.12
PT 14.9 sec (12.8) PTT 37.6 sec (33.4)

Metabolic work ups:

FBS 69 mg/dl
Lactate 2.68 mmol/L (0.56-2.25)
Lipid profile
TG 123, Cholesterol 144, HDL 31, LDL 80, VLDL 25
Urine metabolic screening- negative all
AFP 7.51 IU/ml (normal)
LDH 411 IU/ml

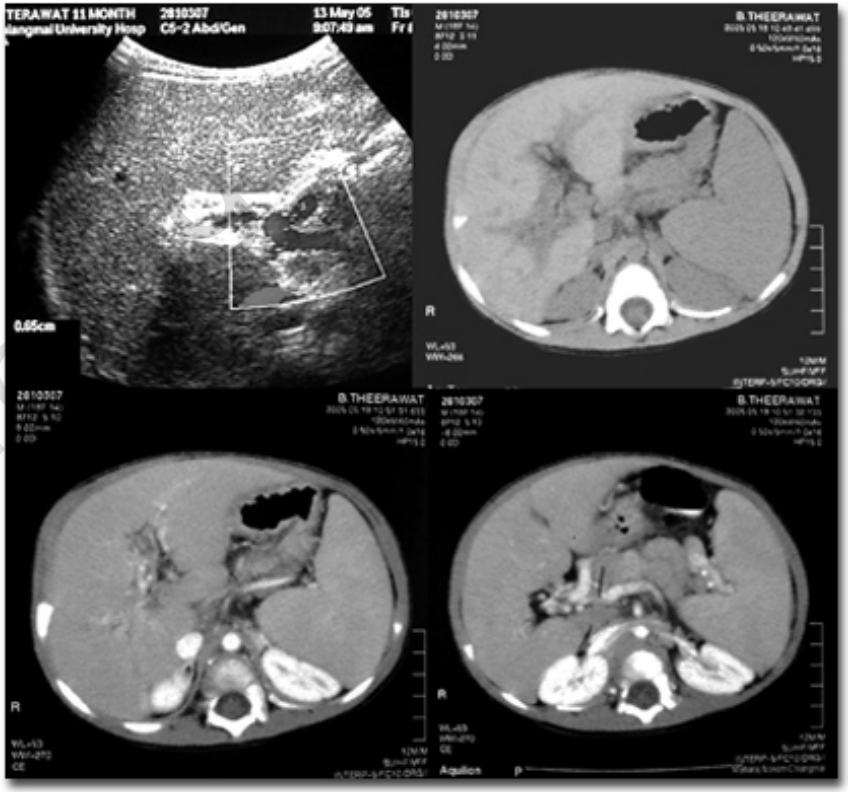

Figure 1: US shows periportal hyperechogenicity with portal vein branches obstruction. The CT scan shows portal vein branches obstruction with the evidence of cavernous transformation at the hepatic hilar. Intraabdominal lymphadenopathy are noted.

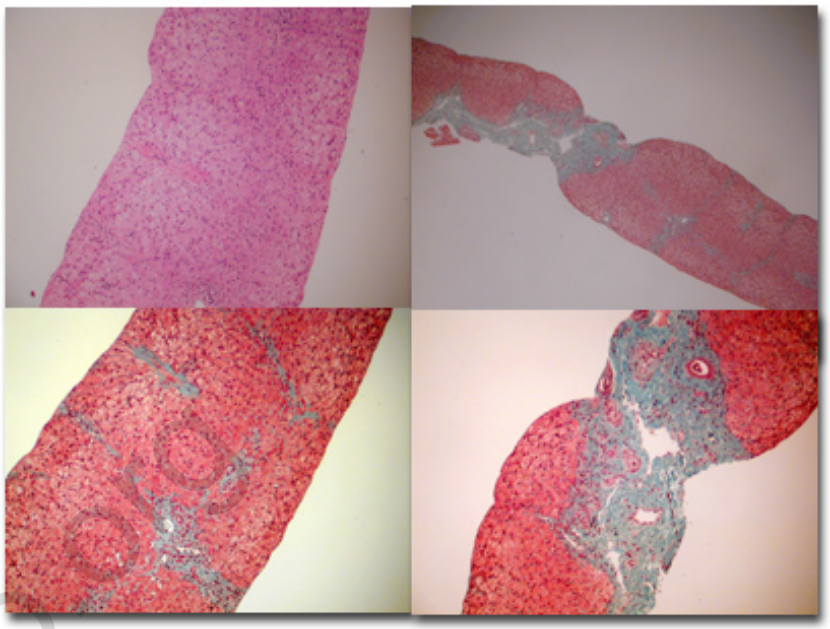

Figure 2: Liver biopsy shows peri-central vein, peri-portal vein fibrosis. There is also evidence of fibrosis in the portal tract. Neither regenerative nodule nor bile duct anomaly is seen.

Diagnosis :

>Hepatoportal sclerosis
>Intraabdominal lymphadenopathy
Abdominal infection as a cause of HPS ??

Treatment :

Propranolol administration and consideration for surgical shunt.

Hepatoportal sclerosis--- Go to topic review page for details