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Case Report

Infantile Hepatic Hemangioma: Report of a New Case

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Abstract

Infantile hepatic hemangioma (IHH) can have asymptomatic presentation or life threatening complications. They can manifest as solitary or multi-centric lesions. We hereby, report a 10 days old male who presented thrombocytopenia, anemia and was accidentally found to have a liver mass, which eventually was diagnosed as infantile hepatic hemangioma and successfully treated with left hepatic lobectomy in our hospital.

Keywords: *Liver Mass; Infantile Hepatic Hemangioma*

Introduction

Primary liver tumors constitute less than 3% of tumors seen in the pediatric population, and only one third of those tumors are benign [1]. Once a liver mass is identified in an infant, the differential diagnosis ranges from vascular malformations to benign and malignant tumors like hepatoblastoma, mesenchymal hamartoma or metastatic neuroblastoma. Careful physical examination, imaging studies, tumor markers and liver biopsy are crucial to determine the final diagnosis [2].

The clinical presentation may vary from the tumor being clinically silent, noticed while a routine prenatal or postnatal imaging for some other cause, or as life threatening complications such as congestive cardiac failure, fulminant hepatic failure, hypothyroidism or abdominal compartment syndrome [1,3].

Case Report

A 4- day-old Chinese newborn boy was admitted to a neonatal intensive care unit (NICU) of our hospital because of abdominal distention, thrombocytopenia and anemia.

The patient was born by spontaneous vaginal delivery at our hospital after an uncomplicated gestation of 37 weeks 4 days. Parents are young and unrelated. His mother had received prenatal care in Health Centre and obstetrician ultrasonography in our hospital at 22 weeks 6 days of gestation was normal. The antepartum screening tests for human immunodeficiency virus (HIV), hepatitis B virus, and syphilis were negative. A maternal test for IgG antibiotic to rubella virus, CMV was in reference range for immunity.

At delivery, the mother had a rupture of membranes 10 - 24 hours but the amniotic fluid was clear. The baby had Apgar scores of 10/10/10 at 1/5/10-minute, respectively. The birth weight was 2.8 kg (25^{th} - 50^{th} percentile), the length 46 cm (25^{th} percentile), and the head circumference 33 cm (50^{th} percentile).

On examination of the newborn the following 48 hours, the vital signs were normal. No scattered petechiae was present on the body. The platelet count was 60 X 10E9/L (reference range 100 - 400) and hemoglobin level decreased after birth (Hb 10.7 g/dL→9.6 g/dL→8.5 g/dL). At 4 days of age, the patient was transferred to the NICU because of abdominal distension, thrombocytopenia and anemia. In NICU, the newborn was awakened and appeared jaundice and mild dehydration, afebrile with a normal blood pressure.

On examination, the abdomen revealed hepatomegaly measuring 5 cm below the right costal margin, hard in consistency, moving with respiration, regular borders, and a palpate left lobe of the liver. Spleen wasn't palpable.

Laboratory test results are shown in table 1. Congenital TORCH Infections blood test and the hepatic virus serology test are negative, respectively and a blood sample was obtained for culture was negative.

Variable	Reference Range	Pre operation	Post operation (5 days)	Post operation (1 month)	Post operation (5 months)	Post operation (10 months)
WBC X10E9/L		12.5	17.4		9.3	6.1
Differential count (%)						
Neutrophils		48.1%	43.9%		29.6%	22.6%
Lymphocytes		27.3%	32.8%		50.0%	650%
Monocytes		21.8%	20.4%		12.1%	8.4%
Eosinophils		1.7%	0.6%		7.6%	3/5%
Platelet count X 10E9/L		86	267		291	247
RBC X 10E12/L		2.7	4.9		4.8	5.1
Hb		8.5 g/dL	13.2		12.2	12.7
HCT		25.8%	41.4%		36.9%	38.8%
Bilirubin (mmol/L)						
Total	(0 - 24 mmol/L)	208	19	4	2	4
Direct		14	9.1			
AST	(<=40 U/L)	39	29	28	26	32
ALT	(<=41 U/L)	15	70	21	25	20
G-GT	(<60U/L)		173			10
Alk-P	(<462 U/L)		186			260
CRP			7.4			
AFP	(< 7.0 ng/mL)	33659.00		713.10	64.78	13.17
Total Beta-HCG	(< 2.6 mIU/mL)	0.1				
FT3	(2.00 - 5.20 pg/mL)	3.34				
FT4	(0.83 - 3.09 ng/dL)	2.02				
TSH	(0.43 - 16.10 mIU/L)	3.66				
PT/INR/APTT	(11.1/ -- /29.5)	9.7/0.9/32.4				10.7/0.9/34.1
Fibrinogen	(288 mg/dL)	363				
D-Dimer		5.58 ug/mL				
Body weight	2.8Kg			4.5 Kg	7.6 Kg	8.78 Kg

Table 1: *Laboratory Data.*

Chest radiography (Figure 1) revealed a normal heart and mediastinum and clear lungs. Abdominal ultrasonography revealed a heterogeneously isoechoic mass in left lobe of liver, measuring about 5.4 cm x 3.9 cm x 5.4 cm and left portal vein is not seen well (Figure 2A, 2B).

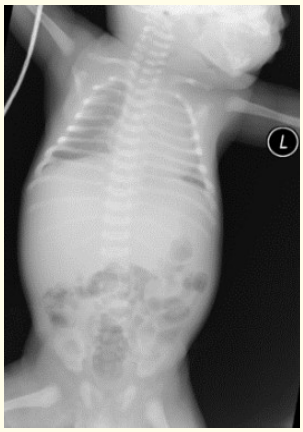

Figure 1: *Normal heart and clear lungs in Chest X-ray.*

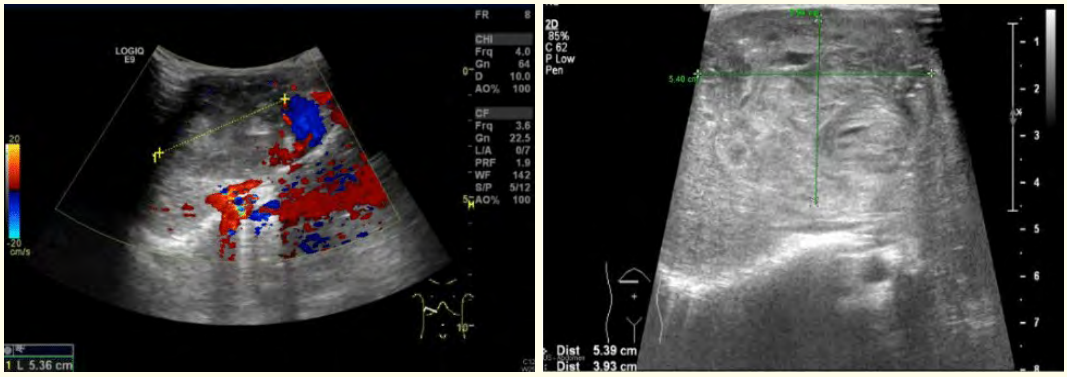

Figure 2A ***Figure 2B***

Figure 2A, 2B: *There is a heterogeneous isoechoic mass in left lobe of liver, about 5.4 cm X 3.9 cm X 5.4 cm.*

Based on these finding we planned for computed tomography (CT) of the abdomen. D it revealed a huge circumscribed heterogeneous hypo-attenuating mass with mild lobular margin about 6.7 cm x 4.5 cm x 5.8 cm, is found in the liver mainly in enhancement in homogeneously after intravenous contrast. CT reported Left hepatic lobe huge mass, hepatoblastoma? Undifferentiated embryonal sarcoma? The differential diagnosis should include mesenchymal hamartoma (Figure 3A-3D).

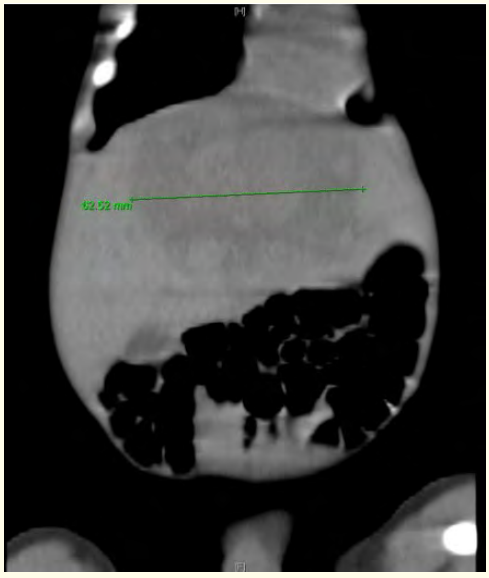

Figure 3A

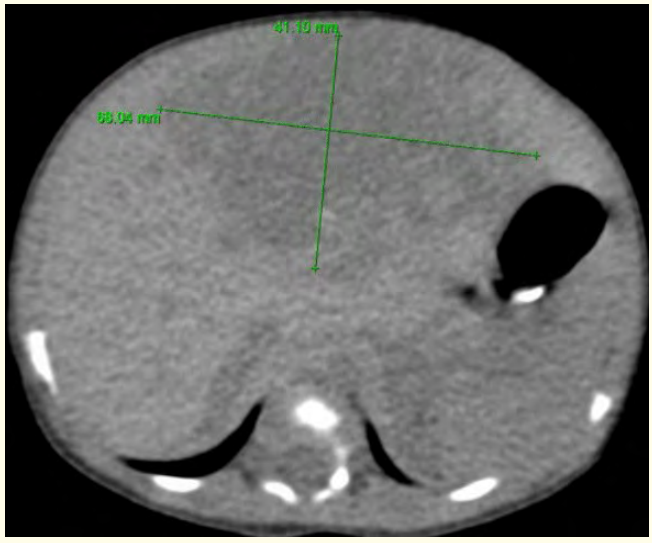

Figure 3B: *Liver CT in non-contrast phase.*

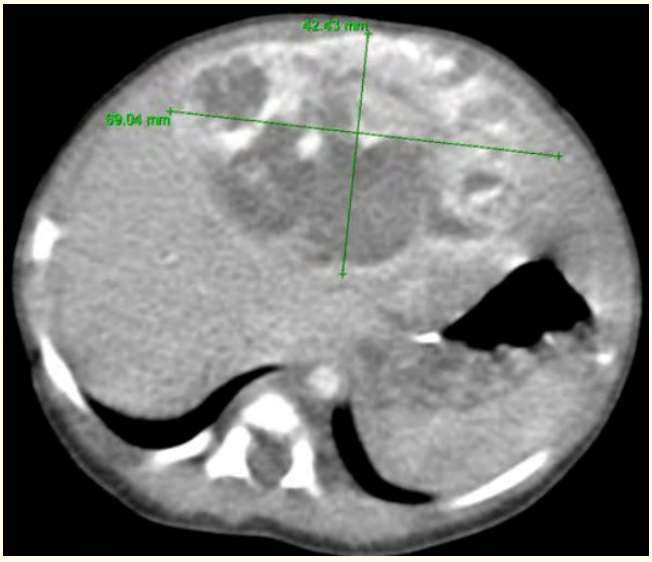

Figure 3C: *Liver CT in arteria phase.*

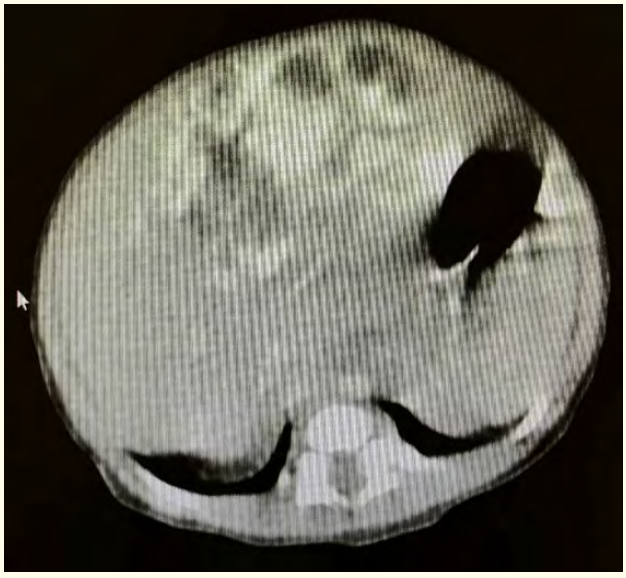

Figure 3D: *Liver CT in portal venous phase.*

Figure 3A-3D: *Marked motion artifacts are noted. A huge circumscribed heterogeneous hypo-attenuating mass with mild lobular margin, measuring about 6.7 cm X 4.5 cm X 5.8 cm, is found in the liver mainly in the left hepatic lobe. Small calcification noted in the mass.*

As the result of these two radiological investigations were that the hepatic mass was a malignant tumor, based on these findings, we planned for a Left hepatic lobectomy undergo a laparotomy.

A histopathological diagnosis was made, the resection of left hepatic lobe (Segment II, III, IV) was a 75g, the endothelial cells do not show obvious atypia. Immunohistochemically, the vascular spaces showed reactivity to CD 34 and CD 31 (done in our hospital lab), but is negative for GLUT 1 (done in on Queen Elizabeth Hospital Hong Kong).

Overall, the tumor is a benign vascular lesion that may be classified as infantile haemangioendothelioma in the 2000 WHO classification. But according to more recent studies on immunostaining of vascular lesions of pediatric patients based on immunostaining for GLUT1, this case should now be classified as congenital hepatic vascular malformation with capillary proliferation with associated capillary proliferation (HVMCP), in view of its negative GLUT 1 staining [4].

After surgical resection, the thrombocyte count was sufficiently raised. On following up the child is asymptomatic and gaining weight with controlled Ultrasonography of the liver was noted the residual liver has been normal. A marked improvement in the hematological data seemed to be obtained by the surgical resection.

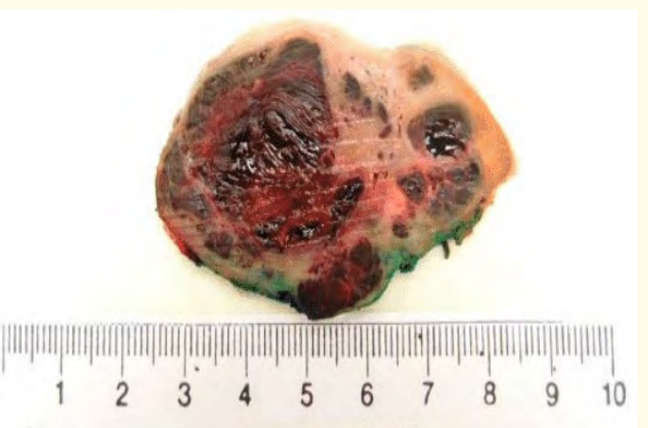

Figure 4A: *The pathological resulted of the patient with infantile congenital hepatic vascular malformation with associated capillary proliferation (HVCMP), measuring 6 x 5.1 x 4 cm, with a grayish brown color in cut surface and have multiple area of extensive hemorrhagic necrosis.*

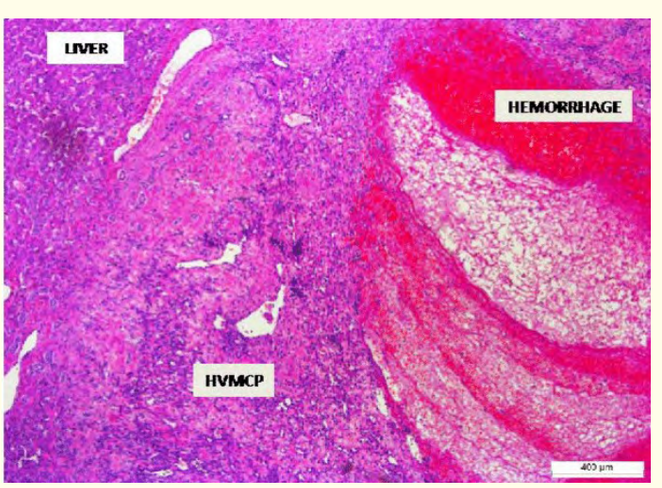

Figure 4B: *HVMCP with central hemorrhage and peripheral reactive fibrovascular stroma containing large tortuous abnormal vessels. (Hematoxylin and eosin 5x).*

Discussion and Conclusion

Hepatic Hemangioma has got several nomenclatures such as cavernous hepatic hemangioma, infantile hemangioendothelioma, multinodular hepatic hemagiomatosis. The various clinical manifestation of IHH, depend upon the tumor size, localization and complication. In

our case, the newborn had abdominal distension, thrombocytopenia and anemia then the liver mass was noted on abdominal ultrasound examination, it reported neoplasm which prompted us to go ahead with a CT scan abdomen being reported there was a huge mass in left hepatic lobe as hepatoblastoma, undifferentiated embryonal sarcoma and mesenchymal hamartoma. In hepatoblastoma, the alpha-fetoprotein (AFP) levels are persistently, markedly elevated and in undifferentiated embryonal sarcoma which clinical and radiological diagnosis is often difficult and it is based on its histology and immunophenotype. Hamartoma is usually a congenital malformation which appears to be cystic or multi-cystic.

It is sometimes impossible to distinguish between these three tumors as was evident in our case. The IHH in our case presented as an extensive hepatic involvement, nearly replacing most of the hepatic parenchyma. It is therefore, very crucial to treat and monitor these patients, as they have the tendency for a stormy clinical course.

In relation to the International Society for the Study of Vascular Anomalies (ISSVA) classification, it is clinically very important to determine whether the hepatic lesions are neoplastic hemangioma or vascular malformations.

This classification has a significant clinical impact, because the former may be sensitive to steroids through the down regulation of proliferating neoplastic cells of endothelial origin, whereas the latter is unaffected. The expression of glucose transporter protein 1 (GLUT-1) is a well-known marker for infantile hemangioma and can be used to distinguish neoplastic hemangioma from vascular malformation [5].

The ISSVA classification categorizes IHH under vasoproliferative vascular tumor and names it as a type of infantile or hepatic hemangioma as it exhibits typical triphasic course (proliferative, plateau, and involution phase) just as in infantile hemangiomas [6].

The newer ISSVA 2015 classification [7] names it under benign vascular tumor category. Based on the presentation, IHH can be categorized as focal, multifocal, and diffuse types, with focal being the most common type [8,9]. Most of the affected infants present by 6 months of age [9], with early onset symptoms indicating significant disease severity, where a large multifocal IHH contributed to significant hemodynamic consequences. The majority of untreated IHH progresses to develop high-output congestive heart failure if not recognized and treated early.

The current Japanese survey clarified that not only patients with diffuse lesion, but also those with huge solitary lesion may be at risk of fatal outcome. Furthermore, the current observation also suggested that any subtype of hemangioendothelioma might be potentially associated with high risk. For those critical IHH patients who have steroid-resistant thrombocytopenia and PT prolongation novel therapeutic options, including beta-blocker therapy, surgical or radiological intervention, and liver transplantation, should be urgently considered as alterative treatment [5].

In summary, although IHH is a benign tumor, it can present as a diagnostic dilemma for the treating doctor. We have reported a case of early symptomatic IHH in a term newborn infant presenting with thrombocytopenia and anemia, which was managed successfully with hepatic lobectomy. After surgical resection, the thrombocyte and hemoglobin count were sufficiently raised. A marked improvement in the hematological data seemed to be obtained by the surgical resection of IHH, but the indications for surgery may have been biased. Surgical patients were considered to be in a stable general condition and therefore capable of undergoing a laparotomy.

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