VASCULAR MALFORMATION OF THE LIVER IN NEONATE: CHALLENGES IN DIAGNOSIS AND MANAGEMENT.REPORT 4 CASES WITH LITERATURE REVIEW.

HAZLINA MOHD KHALID[1], SHUNMUGAM RAJAH[1], ZAKARIA ZAHARI[2] ,ASHOK KRISHNAN[1], SITI HAJAR OMAR[3]
[1]DEPARTMENT OF PAEDIATRIC SURGERY, SABAH WOMEN AND CHILDRENS' HOSPITAL,MALAYSIA
[2]DEPARTMENT OF PAEDIATRIC SURGERY, HOSPITAL KUALA LUMPUR, MALAYSIA
[3]DEPARTMENT OF RADIOLOGY, SABAH WOMEN AND CHILDREN'S HOSPITAL, MALAYSIA.

INTRODUCTION

In children, liver tumours account for about 5-6% of all intrabdominal tumours. Usually primary pediatric liver tumours are malignant. Haemangioma are the primary benign vascular liver tumours which occur in infancy. The clinical presentation may vary from the tumour being silent, noticed while 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.
Some of them may require medical and/or surgical intervention. Therefore, a clinical evaluation is very important to achive a proper diagnosis and treatment to avoid any morbidity and mortality.
We report 4 cases of neonate with liver haemangioma who were treated in our centre. This study aims to evaluate the clinical presentation, imaging modalities, management and outcome of these patients.

MATERIAL AND METHOD

Retrospective study of case record of four neonates who was diagnosed to have liver hemangioma in Department of Paediatric Surgery, Sabah Women Children Hospital in period of January 2010 till December 2016.

RESULT

As shown in Table.1,four neonates with age ranged from newborn to 30 days were treated for liver hemangioma in our department. Three of them were male who presented within first week of life. All of them noted to have abdominal mass at birth. Two cases were referred as hepatoblastoma (CASE 1&3) and suspicious of tumour bleeding in one of the case (CASE 1). Two had symptoms of heart failure. Abdominal ultrasonography were done in all patients showed heterogenous hyperechoic liver mass. Three cases had contrasted abdominal computed tomography (CT) which one patient (CASE 1) had acute renal failure due to contrast nephropathy .His renal function deteriorated with urea 18.0 nmol/l, creatinine 280 nmol/l which later was resolved. CT finding showed multifocal heterogenous enhancing liver lesion in one case [FIG 1] and the other two had focal liver lesion involving right lobe [FIG 2 & 3]. Two cases (CASE 1&3) had liver failure, anaemia, thrombocytopaenia and coagulopathy which required multiple blood transfusion in the first two weeks. As in CASE 1,liver function test showed total bilirubin level was 450mg/dl,(direct:320mg/dl),alkaline phosphatase 925 u/l, alanine transaminase 82 u/l, aspartate transaminase 130 u/l and albumin 15g/l.
Two patients had congestive cardiac failure which echocardiography showed Patent Ductus Arteriosus (PDA) and one had arterioventricular septal defect (AVSD) required diuretics. Three patients (CASE 1,3 & 4) were treated with propranolol starting dose 0.5mg/kg day but one had undergone right hepatectomy for uncontrolled symptoms and failure to thrive. One patient not required any treatment. All patients were well during follow up and serial USG abdomen showed regression of the liver lesion. One dead occur in a child who had hepatectomy due to sepsis.

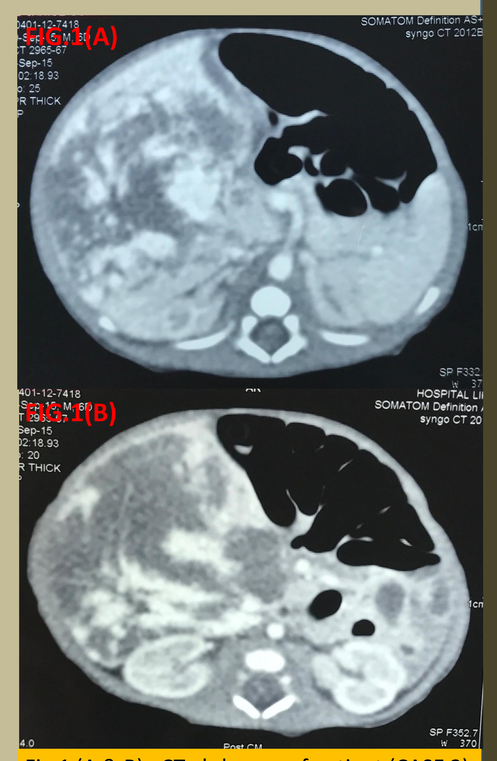

Fig.1 (A & B) : CT abdomen of patient (CASE 2) showing multiple heterogenous enhancing lesion involving both lobes of liver.

	CASE 1	CASE 2	CASE 3	CASE 4
1.AGE	DAY 5	DAY 1	DAY 3	DAY 14
2.SEX	MALE	MALE	MALE	FEMALE
3.CLINICAL PRESENTATION	Abdominal mass, jaundice	Abdominal mass	Abdominal mass, congestive heart failure	Abdominal mass, congestiive heart failure
4.IMAGING MODALITIES				
ULTRASONOGRAPHY	√	√	√	√
COMPUTED TOMOGRAPHY	Focal lesion at right lobe	Multifocal	Focal lesion at right lobe	-
5.ASSOCIATED CONDITION	Acute renal failure ?contrast nephropathy	Nil	Large PDA on double diuretics Failure to thrive ,Coagulopathy	PDA, Small AVSD on single diuretic.
6.LIVER FUNCTION TEST	Elevated liver enzymes, Direct hyperbilirubinaemia,TB:450g/dl	Normal	Elevated liver enzymes and serum bilirubin.	Normal
7.BLOOD TRANSFUSION	Multiples	None	Multiples	None
8.TREATMENT				
PROPANOLOL	Yes	No	Yes	No
SURGERY	-	-	Right hepatectomy	-
9.FOLLOW-UP AND OUTCOME	1 year,size smaller	2 years,size smaller	Died at 2 months old	5 years ,small size static

TABLE 1 : **Summary of clinical symptoms, radiological finding and treatment of four neonates who diagnosed to have liver haemangioma**

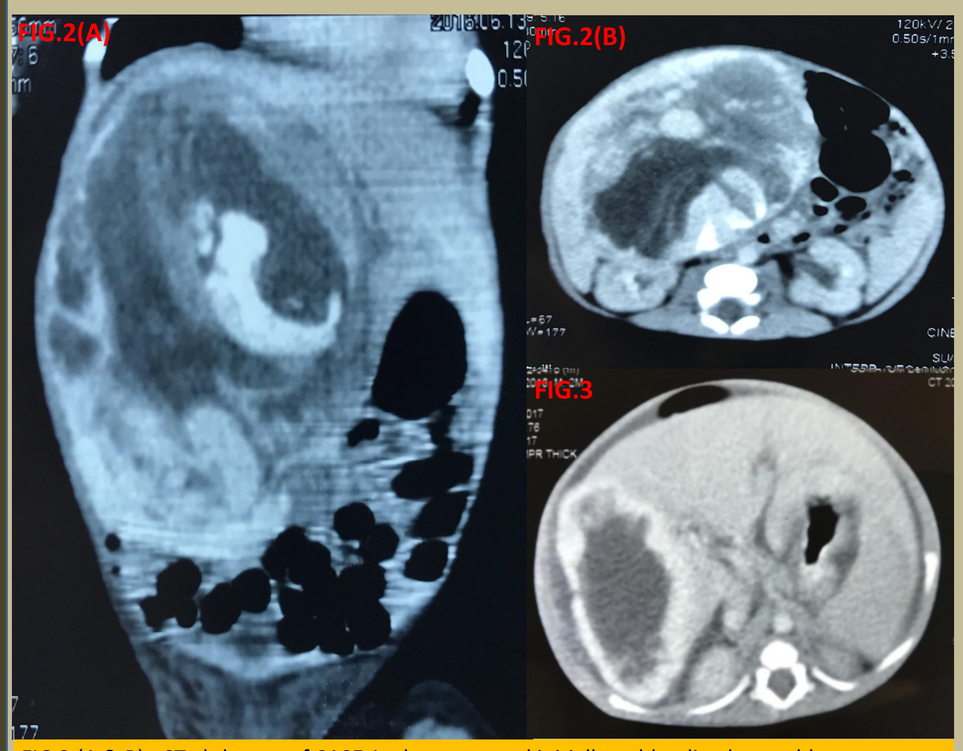

FIG 2.(A & B) : CT abdomen of CASE 1 who suspected initially as bleeding hepatoblastoma showed heterogenously enhancing lesion arising from segment V/VI of liver measuring 6.9x7.3x8.1cm with central necrosis and hemorrhage within
FIG.3.: CT abdomen of CASE 3 showed large well define lobulated hypodense mass with avid peripheral nodular enhancement in segment V/VI/VII of liver measuring 7.3x5.2x8.8cm.

DISCUSSION.

Infant with liver haemangioma (IHH) usually present with one or more of the following ; an abdominal mass, a high output cardiac state, or in frank congestive heart failure. Once a liver mass is identified, the differential diagnosis ranges from vascular malformations to benign and malignant tumours including mesenchymal hamartoma, hepatoblastoma, metastatic neuroblastoma so careful physical examination, imaging studies and if indicated tumor markers and biopsy are important to ascertain the diagnosis. The other associated problems include thrombocytopenia
with platelet sequestration, consumptive coagulopathy, and disorders of hepatic function. Radiological imaging can prove extremely helpful to further characterize the lesion and define the anatomic extent of liver involvement. Hepatoblastomas tend to appear heterogeneous on a T2 weighted MRI sequence and enhance heterogeneosly as opposed to the homogeneous and rapid enhancement of IHHs. The diagnosis is indicated by the clinical features and is supported by the ultrasonographic and computed tomographic appearance.
A variety of approaches to treatment have been tried, with mixed results. Diuretics and digitalis, steroids, radiation therapy, surgical resection, and hepatic artery embolization and ligation have each been claimed to be important in the management of this condition.
In 2008 propranolol, a non-selective ß-blocker was discovered as the treatment of hemangiomas by accelerating IH involution compared to other therapies[20]. IHHs have been shown to successfully respond to the propranolol, as well as the cutaneous counterpart[21-23]. Corticosteroids were considered the gold-standard treatment for before propranolol established. However failure rate was as high as 20%-30% and in 40% of cases there was only a stabilization of the lesion growth more than acceleration in the involution[26]. Moreover corticosteroids lead to significant side effects. These include growth retardation, hyperglycemia, Cushinoid syndrome, hypertension and immunosuppression[27]. It has to be mentioned, however, that even propranolol is not free from side effects but are much less severe than the above medications[22]. Before pharmacotherapy, surgical resection and embolization was considered the mainstay treatment[28]. Surgery for hepatic hemangiomas, mainly only in cases that are refractory to medical management cases. Though IHH is a benign tumor, it can present as a spectrum of disease due to its complication. The rarity of hepatic hemangiomas, the great variability in extent, severity, and complications of these lesions, and the occurrence of spontaneous regression make it difficult to assess the efficacy of the various forms of treatment.

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There is no COI to be disclosed

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