Infantile Hemangioendothelioma - Case Report

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Abstract: A girl infant aged two months from Aswan complaining of abdominal distention and colic. Repeated attacks of vomiting and diarrhea .Multiple skin rashes distributed over the skull and the body, which are vivid superficial tiny masses known as capillary hemangioma. Also, there was heard murmur over the heart. Laboratory investigation and abdominal ultrasonography were done .There was anemia. Normal liver enzymes and liver functions: normal PT hypoalbuminemia. Abdominal ultrasound revealed hepatomegaly with multiple hypo echoic masses of variable sizes at both right and left lobes of liver. Doppler study and Echocardiography revealed secondum ASD 6 mm in diameter and small restrictive PDA for follow up. Also, CT abdomen was done there was enlarged liver which studied by innumerable focal masses in the hepatic parenchyma with tendency to confluence. These masses display hpodense attenuation on the precontrast series .This imaging are classic for infantile Hemangioendothelioma Alpha fetoprotein was done. CT brain was normal. It was normal. Medical therapy with steroid and liver support together with follows up by clinical examination and ultrasonography.

Introduction

A hemangioma is a benign (non-cancerous tumor consisting of dilated blood vessels. When a hemangioma occurs in the liver it is called a hepatic hemangioma. Alternative names are Hemangioma of the liver, cavernous hepatic hemangioma, infantile hemangioendothelioma, and Multinodular hepatic hemangiomatosis. This rare, non-cancerous tumor has been linked to high rates of heart failure and death in infants **[1].** It usually diagnosed at six months old hemangioendotheliomas (HAEs) are the most frequently encountered in infants. The lesions may be solitary or multiple.

Histologically: They are divided into two histological types **[2].Type I** lesions the most common consist of vascular channels of varying size lined by immature endothelial cells and separated by a fibrous stroma which may contain biliary ductules. **Type II** lesions are less common, appear more aggressive, and demonstrate variability in endothelial cell size and shape with ill-formed vascular spaces. There is no stromal bile ductless. There is a degree of overlap between type I and II, and both may show areas of fibrosis and calcification. The lack of vascular invasion and absence of mitotic activity usually makes the differentiation between malignant angiosarcoma more easy .The clinical presentation is variable and depends on the size of the lesions **[2].**

HAEs are characterized by rapid arteriovenous shunting through the vascular channels leading to a raised cardiac output and a vascular (steal syndrome).There is Enlargement of the hepatic arteries and a recirculation of blood back to the right heart, which leads to overt cardiac failure. While, hepatomegaly may contributes to respiratory problems and may need ventilatory support. Sumping of blood within the abnormal vascular channels may responsible for thrombocytopenia due to platelet trapping,

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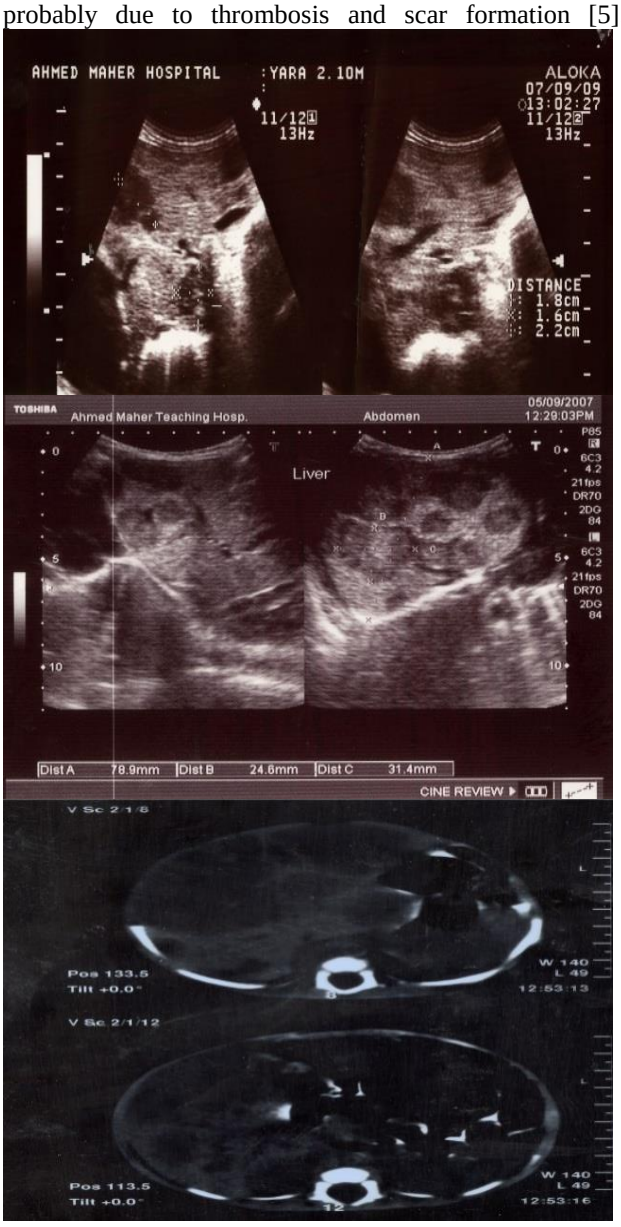
If accompanied by anemia and a consumption coagulopathy (Kasbach-Marritt syndrome **[3**]. Also hypothyroidism is a potential complication of hemangiomas **[4]** particularly for hepatic hemangioma. The mechanism is believed to be tumor production of type 3 idiothyronine deiodinase, causing peripheral inactivation of thyroid hormone. Also, intracranial hemangiomas as well as associated malformative anomalies including absence, stenosis or aneurysm formation of the cervical and intracranial arteries, Cerebral infarction, Cerebral defect such as posterior fossa cystic anomalies, absent corpus callosum, cortical dysplasia and hydrocephalus. So brain imaging should be done. Also, if coetaneous hemangioma overlying the spine is recognized MRI of the spine with and without intravenous gadolinium to rule out intraspinal pathology, as intraspinal extension and dystrophic lesions including spinal dermoid, tethered cord and lipomyeloschisis.

Case Report:

A girl infant aged two months from Aswan complaining of abdominal distention, multiple skin rashes, repeated chest infection and sometimes vomiting and diarrhea. She is the first baby, delivered by CS at 32 weeks gestational age due to premature rupture of membrane. She developed physiological jaundice at age 3 days. Then at 10 days age, rashes appear first on the skull then distributed on the body. They are very small in size, vivid in color and superficial known as capillary hemangioma. After physical, and laboratory investigations, there are hepatomegaly, anemia, moderate thrombocytopenia, Normal liver enzymes and functions. Heart murmurs, for echocardiography, there were situs solitus, secondom ASD 6 mm in diameter causes increasing in pulmonary flow, with systolic gradient across pulmonary artery equals 24 mmhg. Small restrictive PDA monodirectional flow for follow up. After one month, ASD became 3mm

in diameter with mild tricuspid regurgitation, and mildly dilated pulmonary artery. Tinny patent ductus arteriosus detected by color doppler. Abdominal Ultrasound was done; there was hepatomegaly with multiple hypo echoic masses of variable sizes at right and left lobe, the largest 2.4x3.3cm, with tendency to confluent. Alpha fetoprotein was normal. Post contrast CT scan of abdomen Revealed enlarged liver Which is studded by Innumerable, variable Sized focal lesions Affecting the hepatic parenchyma with tendency to confluence These masses display hypo dense attenuation On the precontrast series and exhibit nodular enhancement in the arterial phase with the development of fairly homogenous density in the delayed phase. These are classic for infantile hemangioendothelioma treated by steroid, liver support. She is also closely followed up clinically for serious condition like congestive heart failure or coagulopathy. Also, follow up by ultrasound either for continue to grow or spontaneously regress, probably due to thrombosis and scar formation [5].



Discussion:

Infantile hemangioendothelioma is the most common vascular tumor of the liver in infancy, during the first six months of life[4].The lesions may be single or multiple, calcification are seen at histopathology analysis in 50% of cases[6]. Although it is usually benign tumor but malignant sarcoma has been reported to arise in existing tumor [7] the clinical manifestation of HE is variable. The tumor, as in our case may be asymptomatic and discovered incidentally. The tumor is large and manifests as hepatomegaly, abdominal distention, with multiple cutaneous hemangioma (disseminated hemangiomatosis) which appear early in life at 10 days, months, usually at risk of having hepatic hemangioma. Which are documented by laboratory investigations, anemia, normal alpha fetoprotein and abdominal ultrasound, and CT imaging hemangiomas usually are small, measuring only a quarter of inch in diameter, but they can be several inches in diameter or even larger. The vast majority of hemangioma of the liver never causes symptoms or health problems. Most hepatic hemangiomas are discovered incidentally at the time of testing for unrelated medical problems, most commonly with ultrasound imaging or CT scanning of abdomen. Very large hemangiomas can cause symptoms, esp. if they are positioned near other organs. Pain, nausea, or enlargement of the liver can occur. Rarely, larger hemangiomas can rupture, causing severe pain and bleeding into the abdomen that may be severe or even life threatening. For the physician is to be sure that it is in fact a hemangioma and not another type of tumor, particularly a malignant one, with specialized tests, may include scintigraphy using a tiny amount of a radioactive substance to identify the hemangiomas, CT scanning, or MRI **[8].** In general a biopsy of suspected hemangioma is avoided because of their benign nature and the potential risk of bleeding from the biopsy. Angiography is performed only when endovascular therapy is necessary. Embolization is indicated rarely, generally to control high cardiac output failure or bleeding that is not responsive to pharmacotherapy. Brain imaging should be carried to detect intracranial hemangiomas as well as associated malformative anomalies **[9**]. Hemangiomas go through 3 stages of development and decay. First; is proliferative stage, a hemangioma grow very quickly. This stage can last up to twelve months. In the second stage; there is a very little change in a hemangiomas appearance, this usually lasts until the infant is one to two years old. The third is the involution phase; a hemangioma finally begins to diminish in size **[11**]. Fifty percent of lesions will have disappeared by age five with the vast majority gone by puberty.

Differential Diagnosis:

Three other liver tumors should be considered; Hepatoblastoma, Mesenchymal hamartomas, and metastatic neuroblastoma [**10**]. Hepatoblastoma is rarely seen in patients less than one year of age. The alpha-fetoprotein level is usually markedly elevated, whereas in cases of infantile hemangioendothelioma, it is usually normal or only mildly to moderately increased. By CT,

hepatoblastoma is more heterogeneous than infantile HE, esp. after contrast administration. Mesenchymal hamartomas; usually occur in the infant, and are not associated with elevated alpha fetoprotein levels. Mesenchymal hamartomas; typically appear as multilobular cystic masses, rarely as a solid lesion. When solid mesenchymal hamartomas appear as a vascular or hypovascular masses on angiography, scintigraphy and doppler ultrasound. Metastatic Neuroblastoma; may be confused with the multicentric form of infantile hemangioendothelioma with metastatic neuroblastoma, urinary levels of catecholamine are almost always elevated While, the primary tumor, often an adrenal Mass, can be seen. On contrast enhanced CT scan, marked enhancement is usually noted in cases of infantile HD. While with metastatic neuroblastoma enhancing areas present normal residual liver. Most patients of infantile Hemangioendothelioma present between one and six months of age, although the lesion can be identified with prenatal ultrasound. One-third of cases are discovered in the first month of life, while, less than 5% of the cases are detected beyond one year of age **.**

Treatment:

The treatment depends upon the signs and symptoms. In asymptomatic child:

No treatment may be necessary, although follow-up imaging is recommended in order to demonstrate tumor regression [**13**]. Patients with CHF are treated with prednisone, digoxin, and furosemide. Prednisone appears to accelerate regression of the lesion **[9]**. Most recently, alpha-interferon which has a role in the treatment of life threatening hemangiomas [**10] .** Radiotherapy and Chemotherapy with cyclophosphamide most clinicians tend to adopt an approach to therapy initially with steroid progressing to chemotherapy if no response is seen **[9].** Hepatic artery embolization may be effective [**13],** particularly to stabilize patients prior to elective hepatic resection or as an alternative to hepatic artery ligation. Surgical intervention is required for life threatening lesions [**8**] and involves either hepatic lobectomy if the lesion is unilobar or ligation of the hepatic artery if the lesion is bilobal and multiple. Ligation causes a selective ischemia which reduces the vascular shunt and aids spontaneous involution. Liver Transplantation [**14]** is an option in cases with diffuse tumor involvement or uncontrollable CHF.

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