

## Case Report

## Diffuse neonatal hemangiomatosis presenting as congestive cardiac failure - A case report

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## ABSTRACT

Infantile hepatic hemangioma has substantial arteriovenous shunting which may lead to cardiovascular compromise and hydrops fetalis. It may present as hepatomegaly since the entire liver is involved in most cases. As mortality is very high, a high index of suspicion is required to make a diagnosis and common complications arising out of it, especially in the presence of cutaneous hemangioma. We present a 2-month-old baby born at term presented with features suggestive of sepsis with multiple cutaneous hemangiomas, and on evaluation, there was congestive cardiac failure, which was initially thought of cardiac origin but subsequently came out to be arteriovenous shunting of blood in liver.

**Key words:** Diffuse hepatic hemangiomatosis, Hemangioma, Hydrops fetalis

**C**ongestive cardiac failure in late neonatal period, the usual practice is to evaluate for heart disease and related hyperdynamic state, leading to decompensation. Although diffuse hepatic hemangiomatosis (DHH) is a rare disorder, an uncommon disease in the neonate can present from skin lesion to grave complications. In our case, the infant presented with unexplained cardiac failure, which was difficult to explain in a setting of the absence of other features. A high index of suspicion is required in such multiple and midline hemangiomas, subjecting to search for internal organ involvement and association of high-output heart failure which can result in early intervention and decrease morbidity because, if left untreated or delayed recognition can result in high mortality.

## CASE REPORT

A 2 months female infant came to pediatric emergency with complaints of excessive crying and poor feeding for 9 days and cough and rapid breathing for 4 days duration. There was no history of fever, vomiting, jaundice, cyanosis, or symptoms suggestive of urosepsis. The baby was born to mother G1P1, at 37 weeks 3 days gestation by assisted vaginal delivery with breech presentation in hospital. The baby was exclusively breastfed since birth. The baby had multiple cutaneous hemangiomas all over the body including face, back, lower limbs, and abdomen. They were 20–30 in number, size ranging from pinpoint to 2.5 cm in diameter, largest being on the right thigh (Fig. 1). Mother also noticed increase in number of hemangioma since birth. The baby was completely asymptomatic till the present episode. On admission, examination reveals that the baby had respiratory

distress with features suggestive of congestive cardiac failure in the form of tachypnea (RR–60/min), tachycardia (165/min), gallop rhythm and tender hepatomegaly, liver 6.5 cm below the right costal margin (liver span 9 cm), capillary refill time was 2 s, and SpO<sub>2</sub> 94–96% in room air. Clinically, no added sound heard in precordium except gallop rhythm.

In view of multiple cutaneous hemangiomas, possibility of visceral hemangioma especially hemangioma of liver was kept, which might have predisposed the patient to congenital cervical fusion. Ultrasonography abdomen and contrast-enhanced computed tomography (CECT) abdomen showed multiple hypoattenuating liver masses in both lobes of liver showing enhancement paralleling the vascular pool suggestive of multiple hemangiomas in liver, maximum diameter being 3.1 cm in both lobes of liver (Fig. 2). Other investigations (CECT chest and magnetic resonance imaging brain, X-ray of lumbosacral region) and ophthalmologic examination were done to rule out the presence of hemangioma(s) in other organs, but we could not find any such lesion in the specified area under scanner. Echocardiography was also normal which ruled out any structural malformation of the heart.

Pediatrics surgery opinion was taken and the baby was put on decongestive measures in the form of furosemide (2 mg/day), digoxin (40 mics/kg), and prednisolone (1 mg/kg), and propranolol (2 mg/kg) in conventional doses along with other supportive treatments. The patient improved clinically as regard to decrease in respiratory distress and liver size. Attendants requested discharge on request on the 7<sup>th</sup> day because of personal problems, and the baby was discharged on prednisolone, propranolol, diuretics, and digoxin orally on standard



Figure 1: Child with diffuse hemangiomatosis

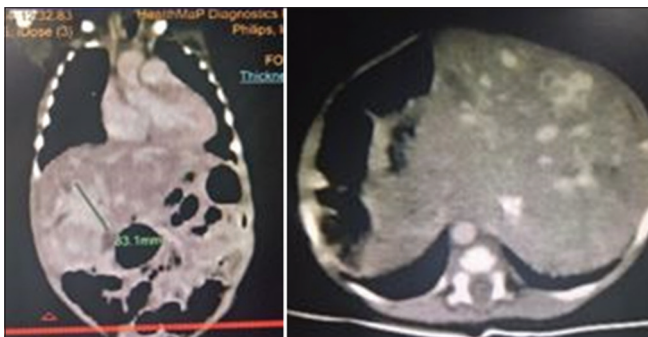


Figure 2: Contrast-enhanced computed tomography abdomen showing multiple hepatic hemangiomas

recommended doses. Mother was advised to come for follow-up ultrasound of liver to look for decrease in size of hemangioma and hepatic embolization if no response could be observed. The patient was lost to follow-up. The baby died at home after 10 days of discharge as informed by attendants telephonically when they were called up for follow-up.

## DISCUSSION

DHH is not a common disorder in the neonate, characterized by extensive replacement of liver parenchyma with hemangiomatous lesions [1]. It is frequently associated with a palpable abdominal mass and high-output heart failure, with high mortality rate in neonates. Diffuse neonatal hemangiomatosis (DNH) is a fatal disease characterized by multiple hemangiomas affecting the skin and visceral organs, although malignant transformation is rare. Typically, these hemangiomas have their onset at birth or during the 1<sup>st</sup> month of life [2]. Liver hemangioma is the most common hepatic vascular anomalies in infancy accounting for 90% of these lesions. Multiple numbers may be found in 10–20% cases [3]. Horii *et al.* found in a prospective multicentric study involving 201 patients with cutaneous hemangiomas an incidence of 16% of hepatic lesions in infants with five or more cutaneous infantile hemangiomas versus 0% in those with fewer than five [4]. In another study by Dickie *et al.* in 26 patients with hepatic hemangioma found that none was having none, single, or three cutaneous hemangiomas. That is the reason

screening is important in patient with any number of cutaneous hemangioma that presents with hepatomegaly with an abdominal ultrasonography [5].

It is very important to differentiate this from the more common multiple cutaneous hemangiomatosis in which internal organs are spared [6]. The liver is the most common extracutaneous site (64%), but virtually any organ may be affected including brain (52%), gastrointestinal (GI) tract (52%), lungs (52%), and eyes [7]. Like isolated hemangioma, DNH proliferates, and then involutes, but extracutaneous involvement may lead to life-threatening complications. Screening with abdominal ultrasonography and other radio imaging is often recommended in patients with more than five cutaneous hemangiomas. These infants carry a greater frequency of hepatic hemangioma in comparison with those with fewer than five cutaneous hemangiomas [4].

Death usually occurs within the first 10 weeks of life as a result of AV shunting in the liver causing increased cardiac output and congestive heart failure as occurred in our case [2]. In addition, other complications include hemorrhage, obstructive jaundice, and consumptive coagulopathy [8]. Without any treatment, the mortality rate is as high as 77%, but with appropriate treatment, the mortality can be reduced to 27% [8]. Diffuse liver hemangiomas, which may occur in the absence of skin hemangiomas, causes massive liver enlargement with compartment syndrome of abdomen, impaired ventilation, decreased venous return, and resulting in higher mortality [9]. Kasabach–Merritt phenomenon is again a condition associated with cutaneous vascular tumor with thrombocytopenia and/or coagulopathy. This condition is more commonly associated with Kaposiform hemangioendothelioma and tufted angioma and truly not associated with infantile hemangioma [10]. However, diffuse hepatic hemangioma can develop even in patients with single cutaneous hemangioma [11]. This highlights the importance of physical examination and screening ultrasound in these patients.

Emergent treatment is required in such condition as otherwise disease is fatal. Multiple treatment modalities are available and corticosteroids are the mainstay of treatment, especially in life-threatening conditions. Corticosteroid stops the proliferation of blood vessels and induces early involution of hemangiomas. Usual dose is 2–3 mg/kg/day which can be increased to 5 mg/kg/day, although associated with more adverse effects [12]. Other medical therapies include interferon 2b alpha, vincristine/vinblastine, and cyclophosphamide can be given, which works by inhibiting angiogenesis but used rarely due to adverse effects.

A non-selective beta blocker, propranolol (1–3 mg/kg/day) has been successfully used for cutaneous hemangioma, has also shown good results in symptomatic hepatic hemangioma. Exact mechanism of action of propranolol is unknown, probably acts by decreasing renin production and causing decreased vascular endothelial growth factor production and also possibly related to vasoconstriction [7,13]. A recent meta-analysis done by Lou *et al.*, in 2013, involving 324 infantile hemangiomas showed that the efficacy of propranolol was greater than other therapies such as steroids, interferon, and vincristine in treating this condition with less severe side effects [14]. The striking effect of propranolol

on infantile hemangioma can be attributed to three molecular mechanisms: Vasoconstriction, inhibition of angiogenesis, and induction of apoptosis [15]. Hypotension, bradycardia, and hypoglycemia are well-recognized side effects of propranolol and need to be monitored closely. In our patient, propranolol treatment was well tolerated and was associated with dramatic improvement in the cardiac function. Hepatic embolization or surgical resection is recommended for focal lesions with direct shunt as a means of controlling heart failure refractory to medical treatment [16].

In our case, the patient had multiple hepatic hemangiomas in addition to cutaneous hemangioma. Most hepatic hemangiomas are asymptomatic and can become symptomatic with (1) high-output heart failure due to AV shunting, (2) associated hypothyroidism leading to overproduction of iodothyronine deiodinase, and (3) Kasabach merit syndrome presenting with coagulopathy and thrombocytopenia. Our patient had high-output cardiac failure for which furosemide, digoxin, and propranolol were started in addition to prednisolone. The patient did not come for follow-up and we could have considered upgrading medical therapy, hepatic artery embolization, or surgical resection in view of poor response to medical therapy.

## CONCLUSION

Diffuse hemangiomatosis of liver with coexisting congestive heart failure, an evidence of AV shunting of blood without extrahepatic involvement, is not uncommon in infancy. Health awareness of the existence of this uncommon entity and supported by screening radio imaging in every case of any number of cutaneous hemangioma can help reach an accurate and early diagnosis, and therefore, early management of these patients can reduce morbidity to a large extent.

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