PST | PEDIATRIC SURGERY IN TROPICS

HEPATIC ATERIOVENOUS MALFORMATION WITH HIGH OUTPUT CARDIAC FAILURE: A CASE REPORT

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Keywords

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Abstract

Introduction: Hepatic arteriovenous malformation (HAVM) is a rare vascular anomaly characterized by an abnormal fistulous connection between the hepatic arterial supply and the hepatic venous system. Although uncommon, HAVM carries significant morbidity and mortality if left untreated.

Case Report: We report a rare case of congenital HAVM presenting at the age of 5 years, complicated by high-output cardiac failure. The patient was managed by coil embolization of the feeding vessels resulting in resolution of symptoms.

Conclusion: Early recognition of HAVM in children is crucial due to the associated mortality and morbidity if left untreated. While endovascular therapy offers excellent outcomes timely multidisciplinary approach is essential in settings where resources are limited.

Abbreviations

HAVM: Hepatic arteriovenous malformation
Hb: Hemoglobin
MCHC: Mean Corpuscular Hemoglobin Concentration
INR: International Normalized Ratio
ECHO: Echocardiography
CT: Computed tomography

INTRODUCTION

Hepatic arteriovenous malformation (HAVM) is a rare vascular anomaly with an incidence of less than 1 in 100 000 in children. HAVM is characterized by a direct fistulous connection between an arterial supply and the portal or hepatic venous systems. Patients may present with microangiopathic hemolytic anemia, portal hypertension, consumptive coagulopathy, and chronic pulmonary hypertension of infancy. In children the median age of presentation is 2.2 months. [1,2] We highlight the diagnostic challenges and management considerations for this rare condition.

CASE REPORT

A five-year-old female patient with no significant past medical, surgical or trauma history presented with malaise, abdominal distention, loss of appetite, and weight loss. The duration of symptoms is unknown as the patient had multiple caregivers, however the patient appeared chronically ill, and was underweight for age. Examination revealed massive ascites, a hepatic bruit and thrill, significant hepatomegaly, a displaced cardiac apex beat, a grade 3 systolic ejection murmur, and a split-second heart sound. Biochemistry revealed a normal liver function test, a microcytic hypochromic anemia (Hb = 7.4 g/dL, MCHC =27.3 g/dL), and a slightly elevated International Normalized Ratio (INR =1.3). Echocardiography (ECHO) revealed mild tricuspid regurgitation and a dilated right ventricle with preserved systolic function. At ECHO, a feeder artery arising from the descending aorta and connecting with a large intrahepatic vascular lesion was noted.

Doppler abdominal ultrasound revealed splenomegaly, ascites, portal hypertension, and hepatomegaly with a large intrahepatic cystic lesion that displayed a Yin-Yang sign suggestive of a hepatic pseudoaneurysm. The Yin-Yang sign is indicative of bidirectional flow and helps to distinguish between vascular and non-vascular hepatic lesions. Triple phase abdominal Computed tomography (CT) scan confirmed a large HAVM involving an anomalous arterial supply from the coeliac trunk, the left portal vein and the left hepatic vein. (Fig 1) An additional feeding vessel originating from the left hepatic artery was also noted.

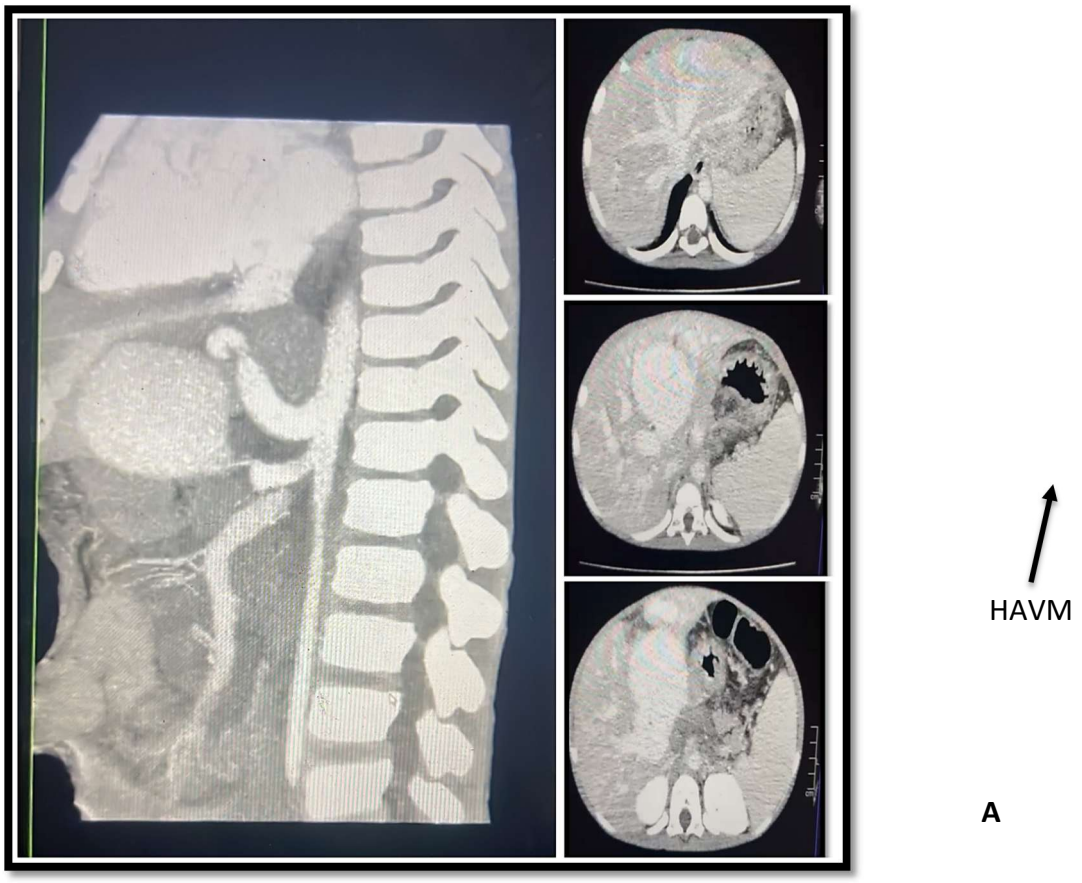

Fig 1. Triple phase abdominal CT scan demonstrating large HAVM (arrow) (A): Sagittal view demonstrating a cluster of abnormal enhancing feeders from branches of the celiac trunk supplying the aneurysm. (B-D) Sequential axial view of the aneurysm (arrow) at different levels of the liver

Formal angiography demonstrated a dominant fistulous communication between an anomalous artery originating from the coeliac trunk, the left portal vein and the left hepatic vein. A second non-dominant feeding vessel originating distal to the origin of the left hepatic artery was also noted. The patient underwent successful embolization of the dominant feeder vessel, though the non-dominant vessel was not addressed due to concern of iatrogenic injury to the left hepatic artery. Post-embolization imaging demonstrated occlusion of the upper coeliac feeder vessel, but the lower hepatic arterial feeder remained patent and continued to supply the HAVM. (Fig 2)

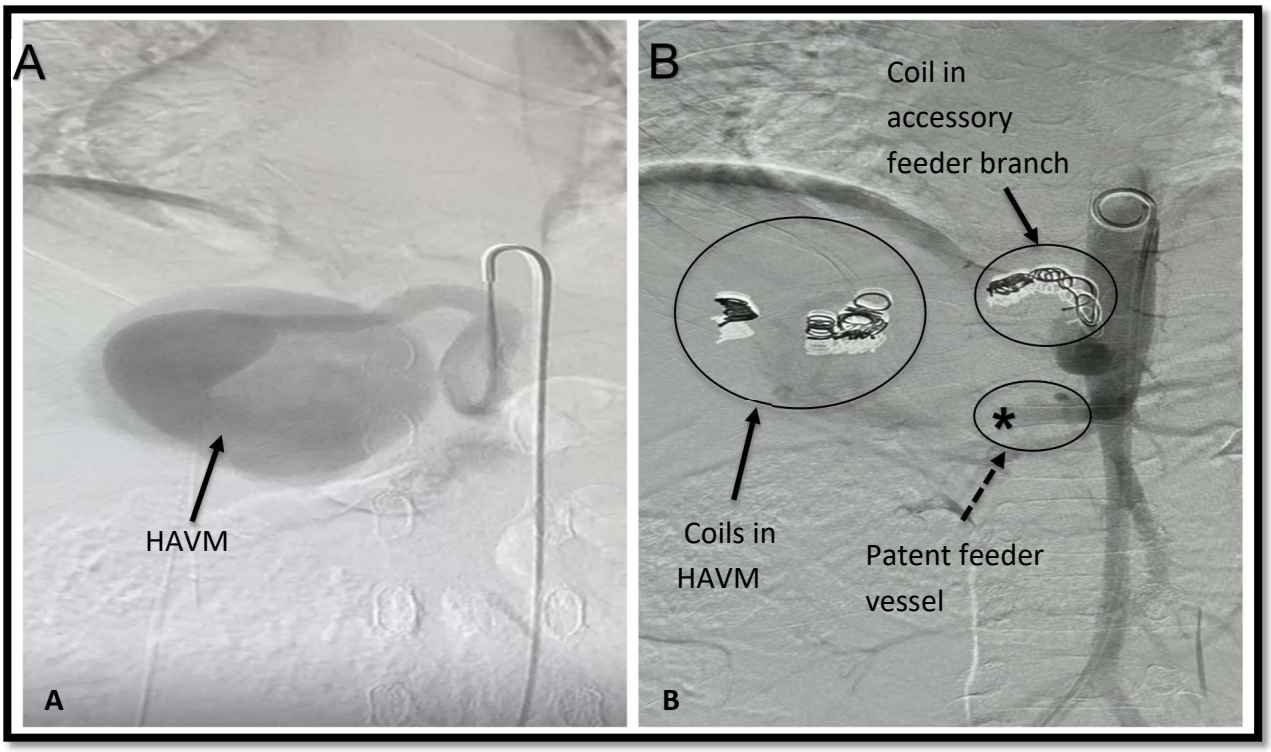

Fig 2. Hepatic selective arteriography before (A) and after (B) coil embolization of a HAVM. (A) Pre-embolization image showing the HAVM nidus (arrow). (B) Post embolization image showing multiple coils (arrows) in primary feeding vessel and an accessory feeder branch with remaining patent feeder vessel (dashed arrow) supplying the lesion

At two-month follow-up, the patient showed dramatic improvement with clinical resolution of ascites and abdominal distension, weight gain, and significantly improved effort tolerance. Given the clinical improvement and proximity of the left hepatic artery to the patent aberrant feeding vessel, the decision was made to repeat imaging in 6 months and then decide on the need for- and safety of, repeat embolization of the remaining feeding vessel.

DISCUSSION

Hepatic arteriovenous malformation (HAVM) is a rare vascular anomaly whose etiology and natural history remain poorly understood. In children, HAVM frequently manifests at birth, indicating a developmental origin. HAVM is hypothesized to result from abnormalities in vascular development between the fourth and tenth weeks of gestation. [(3)] These malformations are attributed to predominantly somatic genetic mutations, which disrupt intracellular signaling pathways essential for normal vascular formation. [(3)] Histologically,

these anomalies are characterized by dysplastic vessels with quiescent endothelial lining that create direct arterial connections to a fistula-like venous drainage system, bypassing the normal capillary bed. [4] In certain cases, HAVM may result iatrogenically from trauma and interventions such as livery biopsy, percutaneous transhepatic biliary drainage, and surgery. [5]

The incidence of congenital HAVM is reported to be less than 1 in 100 000, with a median age of presentation at 2.2 months. [1,2] Our patient presented at the age of 5-years without any risk factors for an acquired HAVM. The presence of abnormal feeders from the celiac trunk further supports the likelihood of a congenital HAVM rather than an acquired one. The delayed presentation is likely due to the child's poor social circumstances.
HAVM often presents a diagnostic and therapeutic challenge and is associated with complications such as high-output cardiac failure, persistent pulmonary hypertension, hepatomegaly, microangiopathic hemolytic anemia, consumption coagulopathy, portal hypertension and hydrops fetalis. [2, 6] A high index of suspicion is therefore essential in children presenting with unexplained hepatomegaly, signs of high-output cardiac failure or abnormal hepatic vasculature on imaging. An early multidisciplinary approach may assist in expediting the diagnostic process and facilitating early intervention.

High-output cardiac failure in HAVM occurs when one or more arterial feeder vessels drain into the malformation, causing progressive shunting of blood from the high-pressure arterial side to the venous circulation. [1] This increased blood flow through the low-resistance system leads to elevated cardiac output leading to a high-output cardiac failure. At birth, the systemic vascular resistance rises resulting in a corresponding increase in blood flow through HAVM. This hemodynamic change is thought to be the key factor in the early postnatal manifestation of HAVM, often presenting heart failure in the neonatal period. [6] If left untreated, complications of HAVM can lead to significant morbidity and mortality with mortality rates ranging from 50-90% with deaths occurring prenatally and during the neonatal period. [6] Due to rarity of this condition, there is limited data on age specific mortality. It is likely that our patient's late presentation was due to her poor social circumstances, lack of access to specialist care, and possibly due to a slow worsening of the patients' shunt as she grew.

Coil and catheter embolization and conservative management with antifailure treatment have been documented in case studies of patients with HAVM, with coil embolization being the preferred method, particularly in cases involving a single arteriovenous fistula. [1,7,8] Additionally, surgical ligation of feeding vessels, liver resection and liver transplantation have been reported in literature. [9] In our case, the patient's clinical condition showed a notable improvement following coil embolization performed at Chris Hani Baragwanath Academic Hospital, a major tertiary referral center in South Africa. While in high-income settings such procedures may be undertaken by a dedicated pediatric interventional radiologist, such subspecialists are scarce in our context. At our institution these technically demanding interventions in pediatric patients are undertaken by experienced general interventional radiologists who adapt their skills and expertise to meet the needs of this population.

Despite this, a residual feeding vessel could not be embolized during the initial procedure, however the patient remained asymptomatic at follow-up. This said, we acknowledge the potential need for further intervention. Given the morbidity associated with surgical intervention, endovascular intervention is the treatment of choice to achieve symptom relief and improve prognosis for patients with HAVM. [8] However, we recognize that in other settings such expertise and resources may not be readily available therefore a decision regarding the best management should be made through a collaborative, multidisciplinary approach considering the skills, experience and infrastructure at hand.

CONCLUSION

HAVM are rare lesions that are usually presented early in life in children and are associated with significant morbidity and mortality. Traditional surgical management has been surpassed by endovascular intervention. In low- and middle-income countries the expertise to perform such intervention, particularly in children, may be limited therefore an early multidisciplinary approach is essential to assess the available skills and resources and plan the most effective intervention for optimal outcomes. Follow-up should be tailored to

monitor disease and symptom progression and determine the need for further therapeutic intervention.

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