

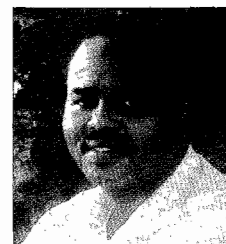
## Case Report: Neonatal Multiple Hepatic Haemangiomas:- A rare presentation

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### Abstract

Hepatic haemangiomas are vascular malformations that rarely affect infants. They have no characteristic presentation. Clinical diagnosis is difficult and various imaging techniques may be required. Different modalities of treatment were tried but in vain. When ruptured, the outcome is grave. They remain a challenge to pediatric surgeons. We are presenting our experience with one infant who presented with a ruptured hepatic haemangioma

**Key words:** Hepatic haemangioma, neonates



### Case Report

**History:** A five days old neonate, outcome of normal spontaneous vaginal delivery, presented with excessive crying, vomiting and right inguino-scrotal swelling for two days associated with low grade fever and slight abdominal distention. There was no constipation, however, the mother noticed recurrent inguinal swelling since birth and a trial of manual reduction was performed at a hospital six hours prior to his presentation.

**General examination:** revealed a very ill, irritable, tachypnic [RR: 64/m], pale and febrile [T39c] neonate, with no jaundice or cyanosis. His pulse was 140 beats/m. Cardiovascular and Chest examination revealed no abnormalities. Abdomen was distended, tympanic, soft, with absent bowel sound, no palpable organs or masses, and normal per rectum examination.

**Local examination:** showed swollen, hot, tense, and tender right inguino-scrotal region with bluish discoloration of the skin over it [Fig 1]. The testicle cannot be felt separately. Fluctuation and transillumination tests were negative.

**Investigations:** Hb 8.8 g/dl, TWBCs: 9,800. Urine analysis, blood urea, serum creatinine, sodium and potassium were normal. Erect X-ray film of the abdomen showed moderately distended small and large bowel loops with no air fluid levels.

**Diagnosis and Management:** Differential diagnosis of strangulated right inguinal hernia with sepsis, haematocele and testicular torsion was made. Nasogastric tube and urethral catheter were inserted and the patient was put on IV Ringer's lactate, metronidazole, ceftriaxone and preoperative oxygen mask. One pint of cross matched blood was prepared. His parents were informed and consented for emergency surgery.

**Operative procedure and findings:** Under well monitored general anesthesia. Through right transverse inguinal incision with good

haemostasis using bipolar diathermy, the subcutaneous tissue, and spermatic cord were found black together with the anterior abdominal muscles [Fig 2].



Fig. (1): Right Inguinal Region and scrotum show colour changes.



Fig.(2) Right transverse Inguinal Incision, showing Subcutaneous and spermatic cord stained bluish to black.

There was black altered blood in the right inguinal region as well as in the right scrotum. The cord was delivered out and the altered blood was evacuated. The cord was examined and process vaginalis was found to be patent. It was opened and the testicle was found intact and viable.

There was a gush of altered blood from the internal inguinal ring. The right inguinal region was covered with sterile gauze and the decision of exploratory laparotomy through a right supra-umbilical transverse incision was

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made. The anterior abdominal wall and the peritoneum were found bluish. There were large clots filling the Morison's pouch and right paracolic gutter, with a lot of altered blood [Fig 3a and b].



Fig. (3a) Huge Clot in Morrison's Pouch.



Fig. (3 b) Anterior Abdominal Wall muscles show some color change

The entire bowel was found distended but intact with no evidence of strangulation. The clots were evacuated. Formal laparotomy revealed multiple hepatic haemangiomas, on the inferior surface of the liver, one with active bleeding [Fig 4 a&b].



Fig. (4-a) Multiple Liver haemangiomas.



Fig. (4-b) Multiple Liver haemangiomas.

Packing of the site of the active bleeding was done successfully stopped the bleeding. Biopsy of one peripheral lesion was taken after haemostatic suture over the biopsied site, which arrested the bleeding. Again the peritoneal cavity was washed with warm saline, sucked and mobbed dry. The laparotomy and right inguinal incisions were closed in layers after ligation of the patent process vaginalis with vicryl 3/0.

The recovery was uneventful. He was admitted to the nursery, where he was transfused with blood again. His pulse, oxygen saturation, respiratory rate and urine output remained unchanged. Three hour later he suddenly collapsed and arrested. Immediate CPR was performed and the patient had uneventful recovery. Unfortunately, two hour later he had another arrest from which he could not be revived.

#### Discussion

Hepatic haemangiomas are vascular malformations that rarely affect infants. Out of 25 cases of haemangiomas in infancy during 10 years period Enjolras O, et al found only three patients to have hepatic hemangiomas<sup>1</sup>. They have protean presentation and may be associated with high morbidity and mortality in affected infants despite their histologically benign nature. Clinical manifestations range from asymptomatic self-limiting lesions, symptomatic hepatomegaly, congestive heart failure associated with high-volume vascular shunting and increased pulmonary vascular tone, abdominal compartment syndrome, anaemia, consumptive coagulopathy to sudden death<sup>2-4</sup>. Because of the complex nature of these lesions and their variable angioarchitecture and a spectrum of angiographic findings<sup>5</sup>, multiple imaging techniques were required for precise diagnosis<sup>6</sup>. Although there is variation in imaging features of infantile haemangiomas, MRI remains the technique of choice in most patients<sup>7</sup>. Ultrasonography is no longer used for discovering haemangiomas, and

liver scintigraphy does not always show the shape of these tumours<sup>8</sup>.

The natural history of the disease as well as the treatment options is confusing. The treatment options include conservative management, medication (steroids and interferons), radiological options (irradiation and selective embolization), and surgical intervention (ligations of feeding vessels, tumor excision)<sup>4, 9-11</sup>. Surgical intervention should be considered only on symptomatic lesions, progressively growing haemangiomas or those tumors which show high risk of bleeding<sup>8</sup>. However, there is no supportive evidence to favor any of the above mentioned modalities of treatment<sup>2</sup>. Of the radiological treatment, intra-arterial embolization was found to be a valuable method for treating symptomatic cavernous liver haemangiomas in newborns, however, its role in multifocal ones is questionable<sup>12-14</sup>.

Ruptured haemangiomas ranged in size from 3.0 to 25.0 cm, and many were located on the inferior surface of the liver. It is a rare complication and when it occurs the outcome is gloomy with over 70% reported mortality. Our patient was not an exception<sup>4, 15, 16</sup>. There fore, surgical resection of ruptured haemangiomas is recommended in low risk patients, while those with high risk may benefit from transcatheter hepatic arterial embolization<sup>14</sup>.

## References

1. Enjolras O, Riche MC, Merland JJ et al. Management of alarming hemangiomas in infancy: a review of 25 cases. *Pediatrics* 1990; 85(4):491-8.
2. Christison Lagay ER, Burrows PE, Alomari A et al. Hepatic hemangiomas: subtype classification and development of a clinical practice algorithm and registry. *J Pediatr Surg* 2007; 42(1):62-7; discussion 67-8.
3. Sidwell RU, Daubeney PE, Porter W et al. Neonatal hemangiomatosis and atrial septal defect: a rare cause of

- right heart failure in infancy. *Pediatr Dermatol* 2004; 21(1):66-9.
4. Samuel M, Spitz L. Infantile hepatic hemangioendothelioma: the role of surgery. *J Pediatr Surg* 1995; 30(10):1425-9.
5. Kassarian A, Dubois J, Burrows PE. Angiographic classification of hepatic hemangiomas in infants. *Radiology* 2002; 222(3):693-8.
6. Burrows PE, Duois J, Kassarian A. Pediatric hepatic vascular anomalies. *Pediatr Radiol* 2001; 31(8):533-45.
7. Kassarian A, Zurakowski D, Dubois J et al. Infantile hepatic hemangiomas: clinical and imaging findings and their correlation with therapy. *AJR Am J Roentgenol* 2004; 182(3):785-95.
8. Jesic R, Radojkovic S, Tomic D et al. Personal experience in embolization of liver hemangiomas. *Srp Arh Celok Lek* 1998; 126(9-10):349-54.
9. Iyer CP, Stanley P, Mahour GH. Hepatic hemangiomas in infants and children: a review of 30 cases. *Am Surg*. 1996; 62(5):356-60.
10. Srivastava DN, Gandhi D, Seith A et al. Transcatheter arterial embolization in the treatment of symptomatic cavernous hemangiomas of the liver: a prospective study. *Abdom Imaging* 2001; 26(5):510-4.
11. Feldman M, ed., et al. Sleisenger & Fordtran's Gastrointestinal and Liver Disease: Pathophysiology/ Diagnosis/ Management, 6th ed. Philadelphia: WB Saunders, 1998; 1379-80.
12. Kullendorff CM, Cwikiel W, Sandstrom S. Embolization of hepatic hemangiomas in infants. *Eur J Pediatr Surg* 2002; 12(5):348-52.
13. Fok TF, Chan MS, Metreweli C et al. Hepatic hemangioendothelioma presenting with early heart failure in a newborn: treatment with hepatic embolization and interferon. *Acta Paediatr* 1996; 85(11):1373-5.
14. Warmann S, Bertram H, Kardorff R et al. Interventional treatment of infantile hepatic hemangioendothelioma. *J Pediatr Surg*. 2003; 38(8):1177-81.
15. Yamamoto T, Kawarada Y, Yano T et al. Spontaneous rupture of hemangioma of the liver: treatment with transcatheter hepatic arterial embolization. *Am J Gastroenterol* 1991; 86(11):1645-9.
16. Davenport M, Hansen L, Heaton ND, Howard ER: Hemangioendothelioma of the Liver in Infants. *J Pediatr Surg* 1995; 30(1):44-7.