MULTIPLE INFANTILE HEPATIC HAEMANGIOMA WITH KASABACH–MERRITT SYNDROME PRESENTING WITH RAISED INTRABDOMINAL PRESSURE AND TREATED WITH PROPRANOLOL: A CASE REPORT

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ABSTRACT

Background: **Infantile hepatic haemangioma is a common benign vascular malformation in the liver, however, its association with Kasabach–Merritt syndrome, raised intrabdominal pressure and intestinal obstruction is rare.**
We report this case of syndromic infantile hepatic haemangioma to raise awareness for its suspicion in this unusual presentation and the role of propranolol in the management.

Case summary: **We report a case of a 3-month-old female who presented with abdominal swelling, difficulty in breathing, constipation, bilateral inguinal swellings and protrusion of the rectal mucosa. There were multiple cutaneous vascular lesions on the palms, chest and abdomen. Examination under mild sedation revealed multiple kernel-like nodules in the enlarged liver.**
She had thrombocytopenia and a computerised tomographic scan of the abdomen showed multiple rim enhancing hyperdense vascular lesions involving the entire liver. She was placed on oral propranolol with regression of the lesion and resolution of symptoms.

Conclusion: **Multiple Hepatic haemangiomas with Kasabach-Merritt syndrome can present with raised intrabdominal pressure and features of intestinal obstruction. This possibility should be considered particularly in infants also presenting with cutaneous vascular conditions. Conservative care with propranolol is an effective treatment option.**

Keywords: Hepatic haemangiomas, Infantile vascular malformations, Abdominal compartment syndrome, Propranolol

INTRODUCTION

Infantile hepatic haemangioma (IHH) is the most common benign vascular tumour of the liver in infancy, accounting for the majority of hepatic vascular lesions in this age group.[1,2] It typically presents with hepatomegaly, abdominal distension, or incidental findings on imaging. Although most cases follow a relatively benign course, complications such as high-output cardiac failure, consumptive coagulopathy, and syndromic associations can occur.[1-3]

One of the most serious complications of haemangiomas is Kasabach–Merritt syndrome (KMS), characterized by thrombocytopenia, hypofibrinogenemia, and consumptive coagulopathy. KMS is more frequently associated with kaposiform haemangioendotheliomas and tufted angiomas, making its occurrence in IHH unusual and clinically significant.[4,5]

The present case highlights an atypical presentation of IHH with raised intra-abdominal pressure, intestinal obstruction, and rectal prolapse. Such gastrointestinal manifestations are rarely reported and may mimic other abdominal pathologies.[5,6] Recognition of cutaneous vascular lesions in conjunction with hepatic involvement was pivotal in raising suspicion for a syndromic vascular disorder.

Case summary

CCM was a 3-month-old female who presented to our hospital through the Children's Emergency Unit with a history of skin lesions from birth, abdominal swelling 2 months earlier and constipation

Multiple reddish-black, shiny skin lesions were noticed at delivery, located on the palms, abdomen, chest, nostrils and right shoulder. They were rapidly increasing in size, not itchy, and not painful with an associated episode of bleeding when traumatised by bathing sponge. The bleeding was said to have resolved after pressure was applied, with no associated history of fainting spells or weakness.

Abdominal distension was noticed 1 month after delivery and progressively increased in size.

There was a history of intermittent inconsolable episodes of crying, no vomiting but has been having constipation. She opened bowel every 4 to 5 days. There was history of protrusion of fleshy mass through the anus with bilateral groin swellings.

Antenatal history was eventful with threatened miscarriage for which she was managed conservatively. There was no history of maternal febrile illness, diabetes or hypertension.

She was delivered via elective Caesarean section on account of a history of previous myomectomy, still birth, and secondary infertility, in addition to advanced maternal age.

She had presented to a neighbouring tertiary health centre where, after initial investigations with plain abdominal x-rays and ultrasound, she was planned for colostomy on account of suspected Hirschsprung's disease before referral to our centre for expert care.

The mother was a 38-year-old school teacher and the father was a 41-year-old trader who deals in phone accessories. Baby was the only surviving offspring of both parents and no similar history in the family.

Examination findings revealed an irritable infant who was restless, pale, anicteric and afebrile.

She weighed 3.3 kg. There were multiple erythematous and blanching papular lesions on her right palm, left shoulder, left upper thoracic region and the right nostril (Figure 1).

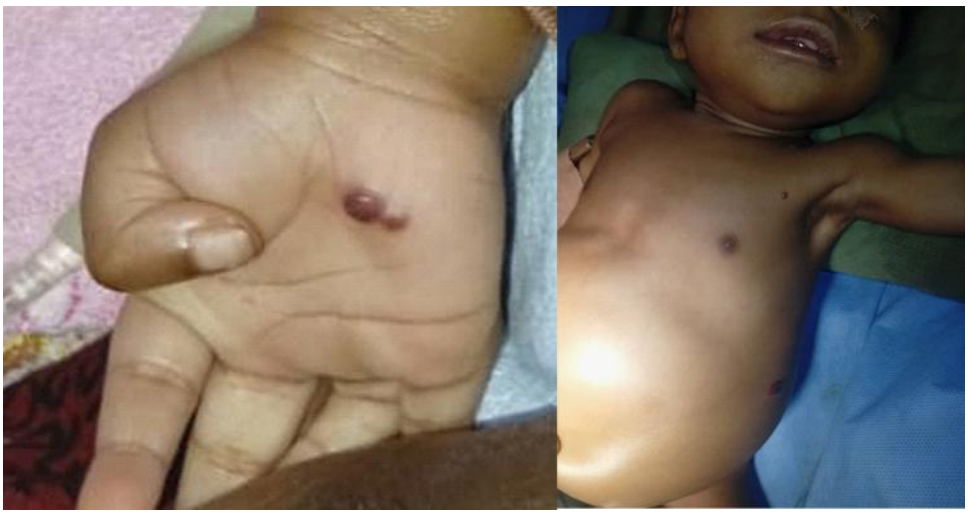

Figure 1: Multiple cutaneous haemangiomas involving the palm, abdomen and chest

Abdominal examination showed a uniformly distended and tensed abdomen with inability to palpate the internal organs however, the bowel sounds were increased. There were bilateral reducible inguinal swellings and protrusion of the rectal wall which appeared to be full thickness. Figure 2

The initial diagnosis of Hirschsprung's disease at the referring hospital was entertained in our centre until investigation results were out.

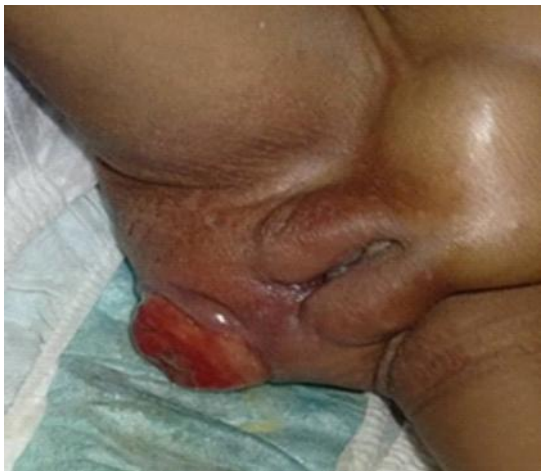

Figure 2: Bilateral inguinal hernias and rectal prolapse

Her full blood count showed white blood cell count of 0.43x10^6/L, haemoglobin of 1.2g/dl, haematocrit of 3.8% and platelet count of 41x10^9/L. Serum electrolytes and blood urea and creatinine were within normal limits as were the liver function test parameters. Random plasma glucose done was 9 mmol/litre. She subsequently had examination under mild sedation revealing hepatomegaly of 8 centimetres below the costal margin with kernel-like firm masses palpated all over the liver surface.

Abdominopelvic ultrasound scan showed hepatomegaly with multiple masses with heterogenous echotexure with compression of the intestines to the contralateral side. The computerised tomographic scan

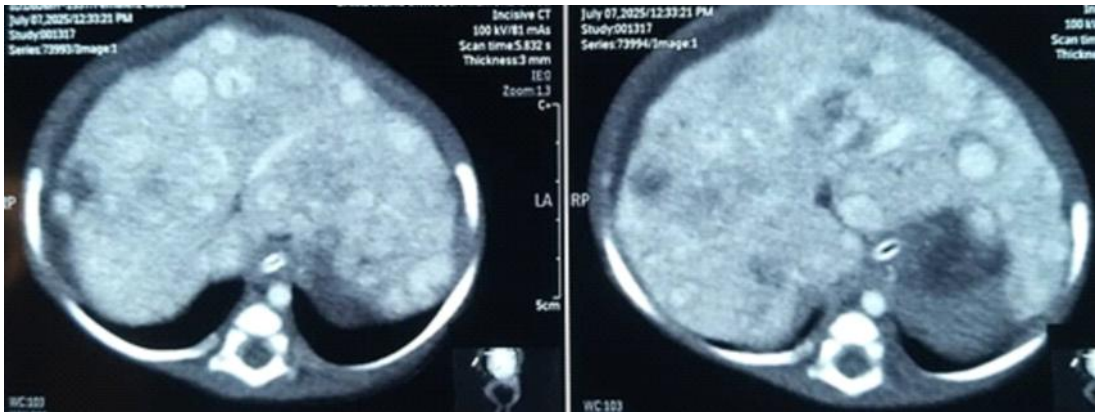

Figure 3: Computerised tomographic scan showing the enlarged liver with multiple enhancing lesions

showed the liver riddled with multiple rim enhancing lesions within the entire lobes of the liver (Figure 3).

She was transfused with fresh whole blood and the blood parameters improved with WBC of 11.69x10^9/L, haemoglobin of 9.1 g/dl, haematocrit of 27.2% and platelet count of 91x10^9/L

A diagnosis of Multiple Hepatic Haemangioma with Kasabach–Merritt syndrome was made.

She was immediately commenced on oral propranolol at 2 mg/kg in two divided doses for a period of 3 months with immediate resolution of symptoms within days to weeks of commencement of treatment.

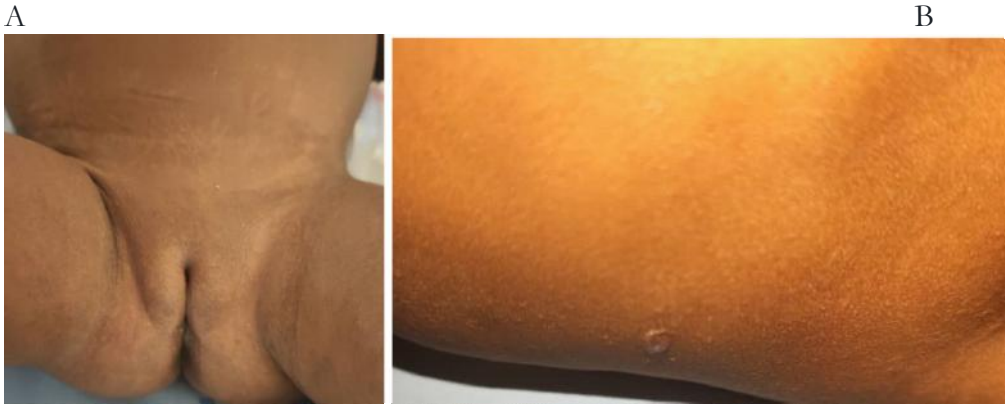

Figure 4: A- Showing the spontaneous reduction of the rectal prolapse and B- resolution of the left anterior chest lesion

She was discharged after two weeks of in-patient care with commencement of oral intake on day five of admission. Complete resolution of both cutaneous lesions and hepatic masses was achieved after 6 months of commencement of therapy without sequalae.

She has been followed up for 10 months with no recurrence and complete resolution of the hepatic and cutaneous symptoms (Figure 4). Follow –up was with clinical examinations, ultrasound scans which showed no residual lesions with return of the liver to normal size and normalized haematological parameters. Computerized tomographic scan was not repeated due to cost and risk of ionizing radiation. She is currently is being planned for elective bilateral herniotomies.

Discussion

Infantile hepatic haemangioma with KMS is a very rare association and its presentation with increased intra-abdominal pressure is an uncommon presentation that could present a diagnostic dilemma.[3,5,6] This index case was evaluated initially as a case of Hirschsprung's disease at the referring hospital and initially in our own hospital.

Findings of multiple cutaneous haemangiomas alongside any other gastrointestinal symptom, such as features of intestinal obstruction, should indeed raise the suspicion IHH. [2] The hepatic involvement could be diffuse, multifocal or localized.[2,3]The present case had multiple lesions in both lobes of the liver as determined by the abdominal examination and confirmed by the abdominal computerised tomographic scan.

The exact mechanism of raised intra-abdominal pressure and causation of intestinal obstruction could not be explained beyond a mass effect of the multiple vascular lesions in the liver.

However, Yang *et al*,[3] had reported two cases of hepatic haemangiomas causing intestinal obstruction as a result of adhesion of the intestines to the lesions, though the cases involved focal lesions rather than multifocal involvement of the liver as in this index case.

The association with Kasabach-Merritt syndrome is diagnosed with findings of thrombocytopenia in the complete blood count and imaging diagnosis with abdominopelvic ultrasound scans and computerised tomographic scans are quite helpful for confirmation of hepatic haemangiomas.[7,8] This index case did not only have a thrombocytopenia but pancytopenia which could be from the systemic vascular lesion, consumption coagulopathy or the chronicity of the illness. The abdominopelvic ultrasound aside including hepatomegaly in the differential diagnosis was not quite helpful in the diagnosis. This could be as a result of the experience of the sonographer and because ultrasound is operator dependent.

Management of IHH has historically included corticosteroids, interferon-á, chemotherapy, embolization, and surgical resection. However, these approaches are often associated with significant morbidities. Since 2008, propranolol, a non selective beta blocker, has been recognized as a safe and effective therapy for infantile hemangiomas, including hepatic lesions.[8] It has also been found to be safe and effective. Its mechanism is thought to involve vasoconstriction, inhibition of angiogenic factors, and induction of apoptosis in capillary endothelial cells. Several reports have documented successful regression of hepatic haemangioendotheliomas with propranolol monotherapy, even in cases complicated by KMS.[2,3]

In our case, propranolol therapy resulted in regression of both the hepatic nodules, cutaneous lesions and resolution of the systemic symptoms, reinforcing its role as a first-line treatment. Compared to invasive approaches, propranolol offers the advantages of minimal toxicity, ease of administration, and suitability for fragile infants.[3,7] This case therefore contributes to the growing body of evidence supporting propranolol in the management of complex haemangiomas.[8,9] Although generally safe, propranolol therapy requires careful monitoring. Reported side effects include bradycardia, hypotension, hypoglycemia, bronchospasm, sleep disturbances, and gastrointestinal upset.[10] None of these side effects was observed in the index case.

Clinically, this report emphasizes the importance of considering IHH with KMS in infants presenting with unexplained abdominal distension, respiratory compromise, and cutaneous vascular anomalies. Early recognition and initiation of conservative therapy with propranolol can avert complications and improve outcomes.

Limitation

As a single case report; generalizability is limited. Further studies with large sample size are needed to establish standardized treatment protocols and to clarify the mechanisms underlying the association between IHH, KMS, and gastrointestinal manifestations.

Conclusion

Infantile hepatic haemangioendothelioma (IHH) is generally considered a benign vascular tumour of infancy, but its association with Kasabach–Merritt syndrome and gastrointestinal complications such as raised intra-abdominal pressure and intestinal obstruction is rare and diagnostically challenging.

Our experience demonstrates that propranolol therapy can be an effective and safe first-line treatment, leading to regression of hepatic lesions and resolution of systemic symptoms.

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