Multiple Gastrointestinal Haemangiomatosis: A Report of Two Cases

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Abstract

Gastrointestinal (GI) haemangiomatosis is a very rare cause of GI bleeding in children. Due to this fact, making the diagnosis sometimes is delayed. In this case report, two different patients with multiple GI haemangiomatosis are presented. Of note is an 8 year old male with a previous history of a vascular malformation of the face presented with features of bowel obstruction (intussusception) and a history of recurrent blood in stool. At laparotomy, multiple haemangiomatosis were seen on the small bowel, right colon, liver, diaphragm and on the body of the gallbladder. Surgical management with excision of the enteric lesions and stapled anastomosis was done.

Keywords: Haemangiomas, intussusception, sirolimus

Introduction

Gastrointestinal (GI) haemangiomatosis is an uncommon vascular tumour.[1] Diagnosis and management of this condition can be quite challenging, especially when patients present with unexplainable recurrent GI bleeding which can sometimes be difficult to diagnose.

The exact aetiology and pathogenesis of infantile haemangioma (IH) remains to be elucidated. There is emerging evidence for an endothelial stem/progenitor cell as the cellular origin of haemangioma. The source of these endothelial progenitors remains elusive.[1]

We report two children with multiple haemangiomas with lesions on the liver, gall bladder, small bowel and large bowel.

Case Reports

Case 1

A 7-year-old boy, known with a haemangioma on the arm, presented with chronic iron deficiency anaemia for several months and occult blood in stool. He required several admissions and transfusions for very low haemoglobins. He was referred to the paediatric surgical unit for gastroscopy and colonoscopy, but these could not reveal the possible cause. On laparotomy, the small bowel was studded with haemangiomas [Figure 1]. A decision was made to resect as much as possible of the lesion to limit haemorrhages. His recovery was uneventful and has maintained normal haemoglobin levels without need for further transfusion and haematinics over the past 2 years.

Histology revealed ulcerating haemangiomas.

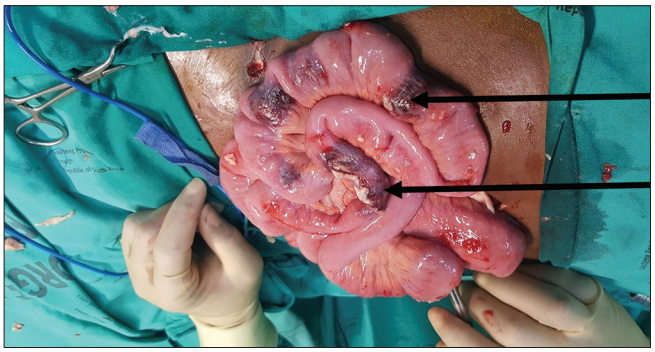


Figure 1: Multiple haemangioma lesions on small bowel

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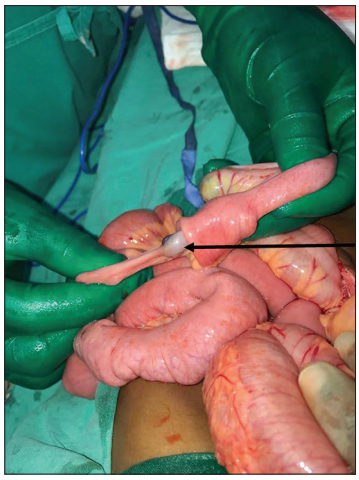


Figure 2: Haemangiomas with intussusception

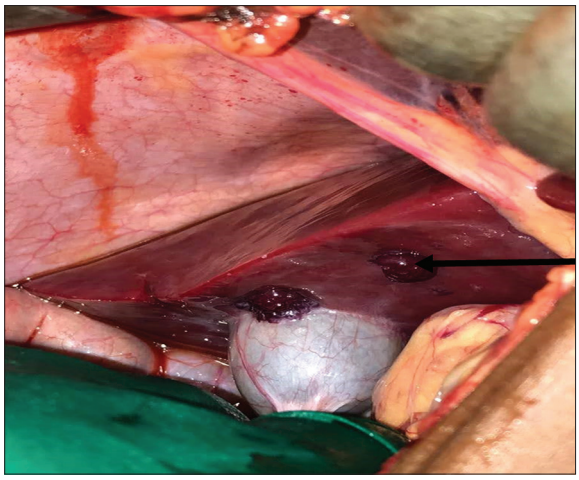


Figure 3: Haemangiomas on the liver



Figure 4: Stapled anastomosis after excision of haemangiomas

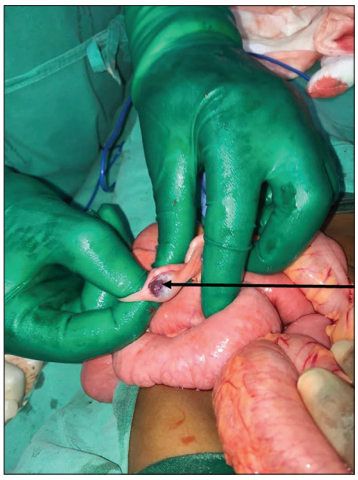


Figure 5: Haemangiomas on small bowel

Case 2

An 8-year-old male, known with facial haemangioma, presented with signs and symptoms of bowel obstruction. Prior to presentation, a previous history of recurrent rectal bleeding was present. On presentation, his serum haemoglobin level was 2.5 g/dL and he was transfused to correct the anaemia.

Abdominal ultrasound and computed tomography (CT) scan of the abdomen confirmed bowel obstruction due to intussusception.

At laparotomy, a jejunojejunal intussusception [Figure 2] about 10 cm from the ligament of treitz was found and was manually reduced. Multiple haemangiomatosis [Figures 3 and 4] were seen on the small bowel with intussusception. Other lesions were also present on the right colon, liver, diaphragm, pancreas and on the body of the gallbladder. On gastroscopy, there were multiple prepyloric gastric haemangiomas.

He was managed with haematinics and followed up in the clinic with a plan to start the patient on sirolimus for the treatment of the haemangiomas. He, however, represented with the same symptoms of anaemia and colicky abdominal pains a few weeks later.

A decision was made for excision of lesions as we were unable to get hold of the sirolimus medication and the patient had represented with abdominal pains and recurrent bleeding a second time. At surgery, a total of 20 lesions were found with multiple areas of small bowel intussusception. The lesions were excised and stapled anastomosis done (18 lesions on the small bowel and 2 lesions on the large bowel were excised in total) [Figure 4]. On the small bowel, 3 lesions on the jejunum and 2 lesions on the ileum as well as 2 lesions on the ascending colon were excised and stapled anastomosis done [Figure 5]. The child's recovery was uneventful and is currently asymptomatic with no further episodes of GI bleeding over the past 2 years. His last serum haemoglobin level on clinic review was 12.7 g/dL.

Discussion

GI haemangiomas are uncommon cause of GI bleeding. The diagnosis can be quite difficult to make as it is not the first differential diagnosis that comes to mind in a patient with upper or lower GI bleeding or a child presenting with chronic anaemia.

The exact aetiology and pathogenesis of IH remains to be elucidated. There is emerging evidence for an endothelial stem/progenitor cell as the cellular origin of haemangiomas. The source of these endothelial progenitors remains elusive.[1]

Some studies suggest a population of resident angioblasts, arrested in an early stage of vascular development, as a source. Haemangioma endothelial cells may be of placental origin because they coexpress several markers: glucose transporter-1, type III iodothyronine deiodinase, Fcg-RIIb, merosin and Lewis Y antigen.[1]

It is important to recognise several discrete diseases, anomalies and syndromes associated with haemangiomas. Multiple cutaneous haemangiomas may be associated with haemangiomas in deep organs, particularly the liver and when affecting the GI tract involves the small bowel most frequently.[2]

GI haemangioma can be histologically classified as capillary, cavernous and mixed types. Of these types, cavernous haemangioma is the most common.[3]

The diagnosis of GI haemangiomas usually includes the use of abdominal ultrasound, CT scan, magnetic resonance imaging, angiography and laparoscopy. Oesophagogastroscopy and colonoscopy can be used depending on the clinical presentation.[4]

Capsule endoscopy has been reported to be useful in the diagnosis of small bowel haemangioma in young infants.[5]

Management of GI haemangiomas is difficult because of sheer number of lesions as presented in the cases above. Asymptomatic patients can be conservatively managed and observed closely; however, the use of steroids, antiangiogenic agents and interferon alpha has been described.

More recently, the mTOR inhibitor, sirolimus, has been shown to be highly effective. Low-dose sirolimus as an antiangiogenic agent has been described for the treatment of these lesions; the vascular masses were reduced rapidly and there was no GI bleeding and muscular haematoma after sirolimus therapy.[6]

In our experience with managing this case, we found that non-surgical management is not always possible in all patients.

Although conservative non-surgical management of these lesions has been described in extensively in literature, persistent bleeding from residual untreated small bowel disease and recurrent disease has lead the reliability and consistency of results using this approach to be questioned.[7-10]

Surgical resection of the lesions is used for patients with multiple lesions or patients unresponsive to conventional medical management depending on the extent and site of the lesion.

Conclusion

Management of symptomatic multiple GI haemangiomas is challenging. There is a place for surgical resections of bowel lesions and may save the patients from long term medical use. Surgery is necessary when medical treatment fails or in emergency situation like intestinal obstruction or massive haemorrhage where immediate intervention is required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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