VASCULAR GASTROINTESTINAL MALFORMATION IN CHILDREN - A CASE REPORT

Smaranda Diaconescu[1], P. Plamadeala[2], Doina Mihaila[2], S.G Aprodu[3], Nicoleta Gimiga[1*], Ancuta Ignat[1], Gabriela Paduraru[1], M. Burlea[1], V.V Lupu[1], Claudia Olaru[1]

[1]"Grigore T. Popa" University of Medicine and Pharmacy - Iaşi, Romania, Faculty of Medicine, Department of Pediatrics

[2]„St. Mary" Children's Emergency Hospital, Pathology Department, Jassy, Romania

[3]„St. Mary" Children's Emergency Hospital, Surgery Department, Jassy, Romania

*Corresponding author: Nicoleta Gimiga, MD, PhD
"Grigore T. Popa" University of Medicine and Pharmacy - Iaşi, Romania
e-mail : chiti_nico@yahoo.com

INTRODUCTION

Children suffering from intestinal vascular malformations may present with symptoms such as abdominal pain, emesis, gastrointestinal bleeding, obstruction or intussusception, diarrhoea, ascites, protein-losing enteropathy [1]; some cases are asymptomatic and the diagnosis may be delayed which lead to severe complications.

CASE REPORT

A boy aged 5 presented to the ER unit of our hospital with impaired general condition, high fever, vomits, intense abdominal pain and constipation for three days. The patient had no significant clinical history and didn't underwent previous abdominal ultrasonography. Clinical examination revealed abdominal guarding and contracture and a firm tumoral mass in the right abdominal flank and fossa of about 15 cm in diameter. Laboratory data showed severe pancitopenia (WBC=2670/mmc, NEU=89%, Hb=6,2 g/l, Ht=19,1%, PLT=16000/mmc), metabolic acidosis (HCO3=16,3 mmol/L), impaired renal function (urea=44 mg/dl, creatinine=1,13 mg/dl), hypoproteinemia (TSP=31 g/l) , hepatic cytolysis (AST= 257 U/l, ALT= 88 U/l).

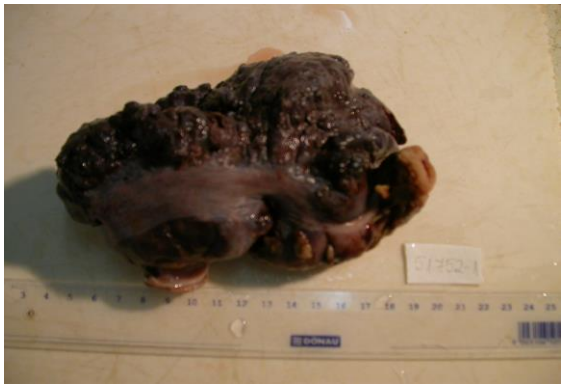

Figure 1. Operative piece distal ileum, appendix and colon with extended hemorrhagic area

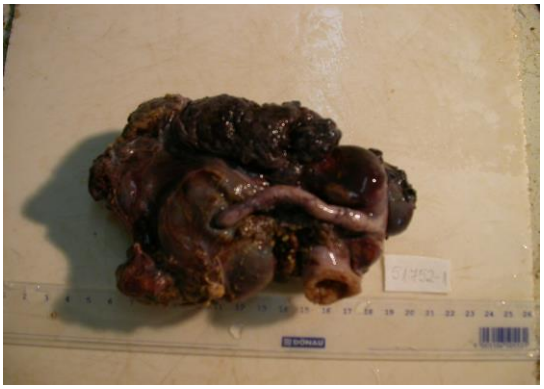

Figure 2. Operative piece: the lumen of the hemorrhagic mass that infiltrates the cecum wall

Ultrasonography and plain X-Ray showed massive overflow fluid in the peritoneum; emergency surgery was decided with the suspicion of acute peritonitis. Intraoperative findings were hemoperitoneum, intestinal occlusion caused by a „tumor" including the cecum and ascending colon and perforation of the transvers colon; a right extended hemicolectomy was performed.

Postoperative course was progressively worsening, with disseminated intravascular coagulation, hemorrhagic shock, sepsis and exitus within 24 hours with multiple organ failure. Histopathological report identified the „tumoral" mass as being a giant cavernous vascular malformation involving the cecum, the ascending and transverse colon.

Figure 3. Hemorrhagic colic mucosa with dilated vessels, HEx40

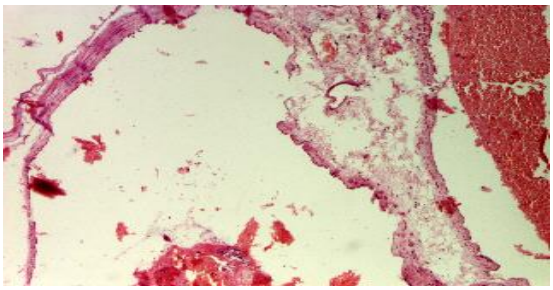

Figure 4. Dilated vessel with thrombus surrounded by hemorrhagic area, HEx40

DISCUSSIONS

To our knowledge, there are worldwide only very few reports on vascular malformations of the small intestine in small children, mostly not larger than 3 cm [2,3]. They may occur anywhere along the intestinal system; the small bowel is the most frequent site with hemangiomas and vascular malformations accounting for 10% of all small bowel tumours [4]. Colonic and anorectal hemangiomas and vascular malformations are even rarer yet, with 200 cases documented from 1931 to 1974 [5,6]. Misdiagnosis is the theme with these lesions. Eighty percent of patients undergo one prior inappropriate surgical procedure [7,8,9]. In a series with five patients, four had undergone a hemorrhoidectomy [10]. A series evaluating rectosigmoid cavernous hemangiomas had misdiagnosed the GI bleeding as hemorrhoids and ulcerative colitis [7,8]. Another series of 47 patients had estimated a delay in diagnosis of 16 years [11]. Hemangiomas and malformations provide few exam findings. Distal lesions can be detected on digital examination, though these masses are not usually overt. These tumours are soft and compressible, with a nodular sensation. When large, a mass on abdominal examination can sometimes be palpated. Evidence of chronic or acute blood loss will be present. In large consumptive masses, a decrease in clotting factors such as fibrinogen, platelets, and factors V and VIII is seen. Obstructive or polypoid lesions are the primary sign of hemangiomas and malformations identified by contrast studies [12]. Anterior displacement of the rectum and widening of the presacral space can be the result of the mass effect and soft tissue component of large cavernous rectal malformations. These masses may collapse with air insufflation.

On CT, pathognomonic findings consist of transmural enhancing bowel-wall thickening with or without phleboliths [12]. The extent of extramural extension and surrounding invasion can also be accurately evaluated on a CT scan. MRI can also add to diagnosis, especially in situations of rectal malformations [13]. On a T-2 weighted MRI, a high signal intensity can be seen, thought to be due to the slow flow. Increased signal

intensity is also noted in the perirectal fat with serpiginous structures correlating to the small vessels supplying the hemangioma. The findings mentioned are very specific findings, and not seen in any other clinical entities. That is why MRI can be more useful, perhaps, than CT. Colonoscopy is crucial in the evaluation and workup of hemangiomas and malformations [14,15]. As noted with air-contrast barium enemas, the polypoid lesions can collapse with insufflation. The upper GI tract should be evaluated to aid in the identification of synchronous lesions, and a complete colonoscopy done to assess the proximal extension. Biopsy is not recommended [16,17,18,19 ,20,21] due to the obvious potential for bleeding, although some have suggested biopsy with caution to promote an accurate diagnosis [22].

As with all GI bleeding, critical care and resuscitative efforts take first priority to ensure that hemodynamic stability is achieved. In true histologic hemangiomas, treatments with corticosteroids have been successful [23,24,25]. However, most GI-associated hemangiomas are actually vascular malformations; therefore, pharmacological options are not effective.

Ideal lesions of polypoid tumours with a narrow base have been successfully treated with snare polypectomy and cauterization [26,27,28]. The argon plasma coagulation has also been reported to have success, even in instances of severe hematochezia [29]. This technique should only be used if surgery is not a feasible option.

Despite more-conservative options, the treatment of choice is surgical resection. The recommended treatment for rectosigmoid malformations prior to 1971 was abdominoperineal resection [16,17,30, 31, 32, 33]. Because sphincter preservation is the goal, low anterior resection with mucosal resection is the current standard [34].

CONCLUSIONS

The benign vascular lesions of the gastrointestinal tract, extremely rare in children include hemangiomas, lymphangiomas and vascular malformations. Early imaging is essential in the diagnosis of these conditions and may prevent life-threatening events; if the abdominal ultrasound is unclear, barium enema, colonoscopy, scintigraphy, CT, MRI and also mesenteric arteriography are useful. In our case the disease had a prolonged silent evolution with a huge increase in the volume of malformation that led to intraperitoneal rupture with severe postoperative complications and exitus.

REFERENCES

1 Yoo, S: GI-Associated Hemangiomas and Vascular Malformations. Clin Colon Rectal Surg. Sep 2011; 24(3): 193–200.
2 Munn J, Hussain AN, Castelli MJ, Diamond SM, Black PR: Ileal perforation due to arteriovenous malformation in a premature infant. J Pediatr Surg 1990, 25:701-703.
3 Frémond B, Yazbeck S, Dubois J, Brochu P, Garel L, Ouimet A: Intestinal vascular anomalies in children. J Pediatr Surg 1997, 32:873-877
4 Varma J D, Hill M C, Harvey L AC. Hemangioma of the small intestine manifesting as gastrointestinal bleeding. Radiographics. 1998;18(4):1029–1033.
5 Lyon D T, Mantia A G. Large-bowel hemangiomas. Dis Colon Rectum. 1984;27(6):404–414.
6 Allred H W, Jr, Spencer R J. Hemangiomas of the colon, rectum, and anus. Mayo Clin Proc.1974;49(10):739–741
7 Amarapurkar D, Jadliwala M, Punamiya S, Jhawer P, Chitale A, Amarapurkar A. Cavernous hemangiomas of the rectum: report of three cases. Am J Gastroenterol. 1998;93(8):1357–1359.
8 Levy A D, Abbott R M, Rohrmann C A, Jr, Frazier A A, Kende A. Gastrointestinal hemangiomas: imaging findings with pathologic correlation in pediatric and adult patients. AJR Am J

Roentgenol.2001;177(5):1073–1081.
9 Oner Z, Altaca G. Diffuse cavernous rectal hemangioma—clinical appearance, diagnostic modalities and sphincter saving approach to therapy: report of 2 and a collective review of 79 cases. Acta Chir Belg.1993;93(4):173–176.
10 Wang H T, Tu Y, Fu C G, et al. Diffuse cavernous hemangioma of the rectosigmoid colon. Tech Coloproctol. 2005;9(2):145–148.
11 Nader P R, Margolin F. Hemangioma causing gastrointestinal bleeding. Case report and review of the literature. Am J Dis Child. 1966;111(2):215–222.
12 Hsu R M, Horton K M, Fishman E K. Diffuse cavernous hemangiomatosis of the colon: findings on three-dimensional CT colonography. AJR Am J Roentgenol. 2002;179(4):1042–1044
13 Djouhri H, Arrivé L, Bouras T, Martin B, Monnier-Cholley L, Tubiana J M. MR imaging of diffuse cavernous hemangioma of the rectosigmoid colon. AJR Am J Roentgenol. 1998;171(2):413–417
14 Fenoglio-Preiser C M, Pascal R R, Perzin K H. Atlas of Tumor Pathology, Second Series, Fascicle 27: Tumors of the Intestines. Washington, DC: Armed Forces Institute of Pathology; 1990. pp. 473–483.
15 Yorozuya K, Watanabe M, Hasegawa H, et al. Diffuse cavernous hemangioma of the rectum: report of a case. Surg Today. 2003;33(4):309–311
16 Coppa G F, Eng K, Localio S A. Surgical management of diffuse cavernous hemangioma of the colon, rectum and anus. Surg Gynecol Obstet. 1984;159(1):17–22.
17 Chaimoff C. On the treatment of hemangioma of the rectum. Dis Colon Rectum. 1985;28(8):632.
18 Borum M L. Cavernous colorectal hemangioma: a rare cause of lower gastrointestinal bleeding and a review of the literature. Dig Dis Sci. 1997;42(12):2468–2470.
19 Aylward C A, Orangio G R, Lucas G W, Fazio V W. Diffuse cavernous hemangioma of the rectosigmoid—CT scan, a new diagnostic modality, and surgical management using sphincter-saving procedures. Report of three cases. Dis Colon Rectum. 1988;31(10):797–802
20 Stening S G, Heptinstall D P. Diffuse cavernous haemangioma of the rectum and sigmoid colon. Br J Surg. 1970;57(3):186–189
21 Cunningham J A, Garcia V F, Quispe G. Diffuse cavernous rectal hemangioma—sphincter-sparing approach to therapy. Report of a case. Dis Colon Rectum. 1989;32(4):344–347.
22 Wang C H. Sphincter-saving procedure for treatment of diffuse cavernous hemangioma of the rectum and sigmoid colon. Dis Colon Rectum. 1985;28(8):604–607.
23 Bartoshesky L E, Bull M, Feingold M. Corticosteroid treatment of cutaneous hemangiomas: how effective? A report on 24 children. Clin Pediatr (Phila) 1978;17(8):625–638, 629–638.
24 Akyüz C, Yariş N, Kutluk M T, Büyükpamukçu M. Management of cutaneous hemangiomas: a retrospective analysis of 1109 cases and comparison of conventional dose prednisolone with high-dose methylprednisolone therapy. Pediatr Hematol Oncol. 2001;18(1):47–55.
25 Ezekowitz R AB, Mulliken J B, Folkman J. Interferon alfa-2a therapy for life-threatening hemangiomas of infancy. N Engl J Med. 1992;326(22):1456–1463.
26 Fraiberg E N, Ahmed S. Colonoscopic excision of a polypoidal cavernous hemangioma of the cecum.Gastrointest Endosc. 1985;31(2):109.
27 Liang L C, Forbes N, David J, Ozick L. Endoscopic polypectomy of an unusually long polypoid colorectal cavernous hemangioma. Gastrointest Endosc. 1998;47(3):307–308.
28 Levitt R E. Colonic cavernous hemangioma. Gastrointest Endosc. 1998;48(3):337.
29 Benson J M, Orlay G. Colorectal haemangioma and its relationship to haemorrhoids in childhood. Aust N Z J Surg. 1991;61(7):537–540.
30 Bland K I, Abney H T, MacGregor A MC, Hawkins I F. Hemangiomatosis of the colon and anorectum: case report and a review of the literature. Am Surg. 1974;40(11):626–635
31 Jaques A A. Cavernous hemangioma of the rectum and rectosigmoid colon. Am J Surg. 1952;84(5):507–509.
32 Hellstrom J, Hultborn K A, Engstedt L. Diffuse cavernous hemangioma of the rectum. Acta Chir Scand.1955;109(3-4):277–283.
33 Westerholm P. A case of diffuse haemagiomatosis of the colon and rectum. Acta Chir Scand.1967;133:173–176.
34 Demircan O, Sönmez H, Zeren S, Coşar E, Bicakci K, Ozkan S. Diffuse cavernous hemangioma of the rectum and sigmoid colon. Dig Surg. 1998;15(6):713–715