Cavernous Hemangioma of Colon and Pregnancy: A Case Report

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Abstract- Cavernous hemangioma of the colon is a rare vascular malformation but is clinically important because it can sometimes cause massive bleeding. We report a case of thrombocytopenia in a post-partum young woman with large cavernous hemangioma of the ileum and colon, culminating in Kasabach Merritt syndrome (KMS), massive uncontrollable hemorrhage, and death. CT scan shows multiple submucosal bulgings in all segments of the colon. Macroscopic examination showed multiple well-defined masses, with gray-brown color in ileum, cecum and ascending colon. The masses were found to be a cavernous hemangioma with a thick fibrotic wall and extensive intra-cystic hemorrhage. KMS is an uncommon complication of colonic large hemangiomas that, as in our patient, can lead to uncontrollable bleeding and death and should be kept in mind by visceral surgeons as one differential diagnosis of large intra-abdominal tumorous masses, especially in young adults.

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Keywords: Cavernous hemangioma; Colon; Kasabach-meritt syndrome

Introduction

Hemangiomas are vascular tumors that rarely involve colon. They can be very small lesions, up to very large ones (1). These vascular tumors are often seen in young patients (2). Generally, hemangiomas are found when patients present with abdominal pain, hematochezia, and melena, or incidentally identified by ultrasonography (US), computed tomography (CT), angiography, or magnetic resonance imaging (MRI) and diagnosed by histopathological examination after resection of suspected masses (1). Hemangiomas of the large intestine can occur anywhere in this organ (3). They can present as solitary, multiple, or part of a more complex syndrome with cutaneous manifestations (4). Although a variety of histological and clinical types of hemangioma exist, the cavernous and capillary types are most commonly encountered (5-9). Steroids, estrogen therapy, and pregnancy can increase the size of an already existing hemangioma (10). Kasabach Merritt syndrome (KMS) is an uncommon complication of large hemangiomas in which there is thrombocytopenia and coagulopathy. Hemorrhoidal bleeding is common during pregnancy. Other preexisting anorectal conditions can also be exacerbated by the increased vascular volume and pelvic congestion (2). Recent advances in colonoscopic techniques have led to successful endoscopic resection in selected cases (5-9). Surgical resection is the mainstay of most large lesions' treatments, with an emphasis on sphincter preservation. Nonsurgical endoscopic treatment with banding and sclerotherapy has been reported with success, especially in instances where an extensive resection is not feasible (2-10). We report a 21-year-old woman with a large cavernous hemangioma involving ileum, the whole colon and pericolic fat with subsequent development of KMS.

Case Report

A 21-year-old woman presented with 17 days history of generalized abdominal pain radiating to back after normal vaginal delivery. Laparotomy in another town was done with suspicion of uterine perforation. The surgeon saw multiple masses in the colon and referred the patient to our center. On admission, she had generalized abdominal pain and distension. There were no signs of other disorders, including fever, chills, jaundice, nausea, vomiting, melena, or hematuria. She had anorexia and mild dyspnea. Her past medical history included chronic hepatitis B. On physical examination she was pale with abdominal distension, and a 5cm mass

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was palpated in the right lower quadrant. Laboratory studies, including tests for serum amylase, creatinine, aspartate aminotransferases, bilirubin, and urea nitrogen gave normal results. CBC showed anemia (HB:6.3 mg/dl) and thrombocytopenia (PLT: 89000). Other laboratory tests showed PT:16.3, PTT:35 and INR:1.8. HBsAg was positive. A fibroscan of the liver was performed. The median liver fibrosis was F2 on metavir histological index. The cap score for liver steatosis was 340 which is equal to 82%. US (ultrasonography) showed a giant retroperitoneal microcystic mass (180×102 mm) on psoas muscle, but no additional abnormalities of the uterus, ovaries, liver, gallbladder, pancreas or urinary tract were found. A spiral CT scan of the abdomen was done (Figure 1).

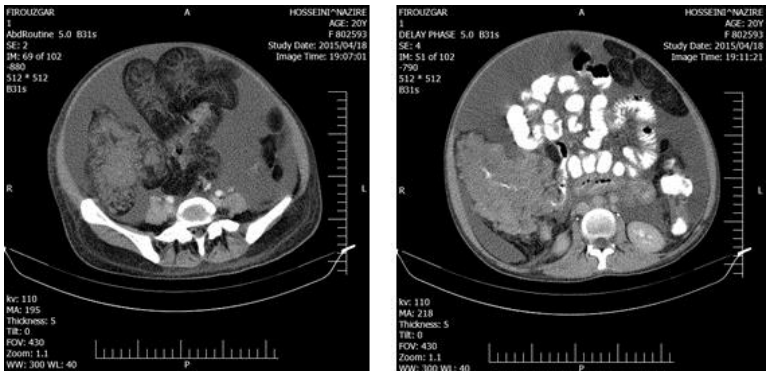

Figure 1. A giant colon microcystic mass (180×102 mm) on psoas muscle using spiral CT scan

Multiple submucosal bulgings were seen in all segments of the colon, along with some fresh blood in colonoscopy. Also ,a large polypoid mass in the rectum and multiple small polyps in rectosigmoid were reported. EUS study revealed rectal sessile non-circumferential polyp extending proximally and pelvic ascites. Rectal wall thickening was seen up to 12 cm in non-circumferential pattern limited to mucosa; submucosal layers and muscularis propria layer were intact (Figure 1).

At surgery, 5 liters of intra-abdominal blood was aspirated. A large intact mass involving the cecum and ascending colon without bleeding and similar lesion in rectum were evident. A superficial laceration with oozing was seen on the surface of the uterus. Right hemicolectomy and cholecystectomy were done. Macroscopic examination showed multiple well-defined masses, with gray-brown color in ileum, cecum and ascending colon. The largest one was 230×50×50 mm in the ileum. Tumors had thick walls and spongy appearance, with large dilated spaces filled with clots and blood, and surrounded by multiple lacunas containing a light-yellow gelatinous liquid mixed with blood (Figure 2).

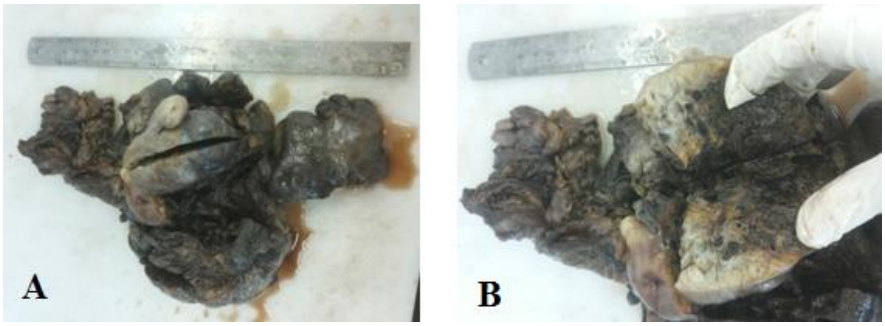

Figure 2. (A and B) Multiple thick-walled masses, with gray-brown color in the ileum, cecum and ascending colon

Microscopically there were multiple vascular spaces of various sizes, lined by a single layer of flattened cells in all parts of the bowel wall, but most prominently in submucosa and subserosa, greatly involving pericolic fat. Fibrosis and infiltration of inflammatory cells were seen in the tumor wall (Figure 3 A-D).

After surgery platelet count was 44000 and Hb 9, so she was transfused with pack cell blood and platelets and FFP and was transferred to ICU for assisted ventilation. She had GI bleeding from NG tube, but endoscopy just showed oozing of the gastric mucosa. She had fever, leukocytosis, anemia, and persistent thrombocytopenia despite platelet transfusions. Her INR fell below 5.5, and she expired.

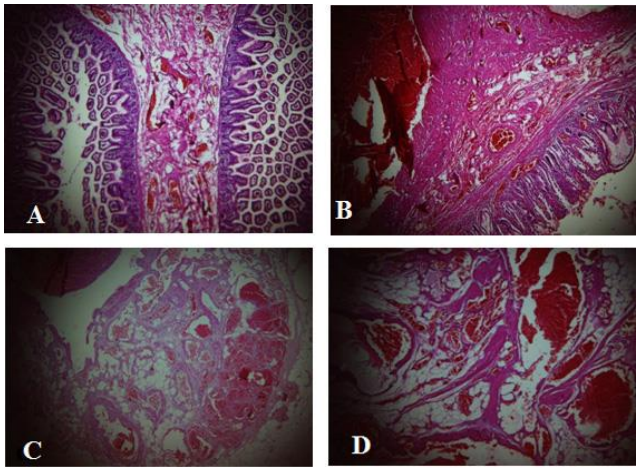

Figure 3. Microscopical findings: a cavernous hemangioma (CH); A., B., C., D)

Discussion

Hemangiomas are rare lesions of the colon, but vascular malformations of the gastrointestinal tract have been reported since 1839 (11). According to the literature, these lesions originate from embryonic sequestrations of mesodermal tissue (12). Most colonic hemangiomas are capillary or cavernous type. Capillary hemangiomas consist of a proliferation of small capillaries composed of thin-walled spaces lined by endothelial cells, while cavernous hemangiomas consist of large spaces lined by single or multiple layers of endothelial cells (13). The capillary subtype is usually solitary and causes no symptoms, while the cavernous subtype presents with bleeding (60%-90%), anemia (43%), obstruction (17%), and, rarely, with platelet sequestration. Approximately 10% of patients remain asymptomatic (14). The endoscopic findings of colon hemangiomas vary. Grossly, hemangiomas appear as soft, compressible bluish or deep red submucosal lesions, with dilated, engorged veins in the rectal wall (9). The patient's presenting symptoms were unremarkable, with the main symptom being dull abdominal pain, which would have been due to the slow-growing CH. However, large CHs can cause compression of surrounding organs and corresponding complications, such as hydronephrosis and upper hydroureter in the patient (15,16).

KMS is an uncommon complication of large hemangiomas in which there is thrombocytopenia and coagulopathy (17,18). Sylla *et al*., (2008) believed that large hemangioma should not be biopsied. Abdominal CT can provide useful information about the size of the lesion and its extension to adjacent organs. They suggested that complete surgical resection is the best treatment for large or diffuse lesions. Polypoid hemangioma sometimes involves the entire wall of colon, extending through the mesocolon and mesentery, as in our case (9). Most patients with hemangiomas of the colon are young men. Misdiagnosis often occurs because of lack of awareness of classic clinical features. Endoscopy, plain abdomen x-ray, barium enema, CT scan, MRI and selective angiography are useful means of investigation for accurate diagnosis (19-20). In our patient, abdominal CT scan revealed useful information about the size and location of the lesion and the extension to adjacent organs. Expansive cavernous hemangiomas may infiltrate organs, such as the bladder or uterus that lie in anatomical proximity to the colorectum. Misdiagnosis often occurs due to the lack of specific clinical features. Hemangiomas have been frequently mistaken for internal hemorrhoids, carcinoma, inflammatory bowel disease, adenomatous polyps, and other conditions (19,21-24).

The first impression of the case was a cystic tumor originating from the colon. For such cases, the differential diagnosis included malignancies such as sarcomas, or benign lesions such as paraganglioma,

neurofibroma, lipoma, teratoma, and neurilemmoma. Observation showed that the tumor was localized and there was no evidence of invasion or metastasis to peripheral organs, so resection of the tumor to relieve the pressure on neighboring organs was performed. Pathologically, the gross findings of the tumor included dilated spaces filled with blood, consistent with the typical appearance of CH. However, the thick walls seen in our patient's tumor is rare in CHs, and this was distinct from lesions in the kidney, pancreas, adrenal gland, or even previously reported primary retroperitoneal cavernous hemangioma (PRCH) (25). Microscopically, the inner components of the tumor consisted of variously sized vascular spaces lined by a single layer of flattened cells, which stained positive with CD34 and vimentin. CD31 and factor VIII-related antigen are also informative markers for the diagnosis of CH (25,26). There was inflammation and fibrotic thickening of the tumor wall in this case, which probably prevented the contrast medium from filling the tumor on CT scan. Treatment in hemangioma with thrombocytopenia is different as De Pree (1963) showed in his work (27). Steroids, estrogen therapy and pregnancy can increase the size of an already existing hemangioma (8). Ryu *et al*., (2008) and Gottlieb *et al*., (2008) reported hemangiomas in pregnant women (28,29). Gottlieb *et al*., (2008) presented the case of a young woman who developed life-threatening rectal bleeding requiring early delivery. Through the use of endorectal endoscopic ultrasonography (EUS), the condition was diagnosed as a diffuse cavernous rectal hemangioma (29). Ryu *et al*., (2008) reported a case of hepatic hemangioma rupture in a 36-year-old woman with a 34-week twin pregnancy. Because of non-specific symptoms and the presence of the large gravid uterus during pregnancy, it was difficult to reach this diagnosis. Under spinal anesthesia, a cesarean section for twin delivery and removal of the hematoma in the abdominal cavity were performed. However, the source of active bleeding was not found during the operation. The spontaneous rupture of hepatic hemangioma was diagnosed after postoperative hepatic angiography and treated successfully by embolization of the left hepatic artery (28). Chatzoulis et al (2008) and Xiao *et al*., (2004) found that steroid therapy, estrogen therapy and pregnancy can increase the size of an already existing hemangioma (30,31). Experimental studies have revealed that estrogens augment endothelial cell proliferation, migration and organization into capillary-like structures (30). Xiao *et al*., (2004) described that hemangiomas have estrogen receptors, an indication that these tumors may be a target tissue for estrogens (31).

Complete surgical resection is the definitive therapy for colonic hemangioma, although: 1. a pedunculated polypoid hemangioma, 2. less or equal to 2.5 cm, with 3. endoscopic ultrasonography demonstrating that the depth is limited to the submucosal layer are the three major criteria for endoscopic resection (32).

There are currently no consensus guidelines for the treatment of KMS. Some studies have reported good therapeutic effects with comprehensive sequential therapy, including steroid therapy, interferon, arterial embolization, vincristine, radiotherapy, and surgery (33-35). Nonsurgical endoscopic treatment with banding and sclerotherapy has been reported with success, especially in instances where an extensive resection is not feasible (34). Azad *et al*., (2012) reported a case of a newborn male baby presented with a large hemangioma on the right upper extremity and his initial laboratory tests were consistent with consumptive coagulopathy. He was diagnosed as a case of KMS and treated with oral prednisolone (36). Children with a large vascular tumor and associated Kasabach-Merritt coagulopathy respond inconsistently to therapy and have a high mortality rate. De Pree (1963) reviewed 49 cases of giant hemangiomas with thrombocytopenia and found great variation in the localization and histological, clinical course, and type of treatment in them (27). Arunachalam *et al*., (2012) reported a case series of four patients who presented with large surface vascular tumors and low platelet count and their management. They did medical management with steroids, propranolol and vincristine in different combinations. Their results showed excellent final response without surgery (37). It is concluded that KMS is a complicated disease with high mortality and difficult treatment modalities. Surgical treatment is still controversial but the only curative modality.

In this study, we presented a case of colon angiomatosis with thrombocytopenia, a rare type of colon hemangioma, in an adult patient. The tumor was separate from surrounding organs. Thus it was diagnosed as a colon CH. The colon CH, in this case, had thick fibrous walls and extensive intracystic old hemorrhage, leading to the initial cyst-like appearance. The clinical features of colon CH may be more subtle than other types of CH and clinicians need to be alert to the possibility of colon CH as such tumors can grow to a very large size and cause serious complications. Surgical resection is curative treatment for colon CH, which reduces the risk of hemorrhage and relieves the pressure on neighboring organs. Presence of thrombocytopenia in angiomatous tumors is an ominous sign and highly

increases morbidity and mortality. KMS is an uncommon complication of colonic large hemangiomas that, as in our patient, can lead to uncontrollable bleeding and death.

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