Colon Tumor Malignancy vs. Retroperitoneal Fibrosis?

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ABSTRACT
51 y/o man with a clinical presentation of severe abdominal pain for several weeks localized to lower abdomen progressively worsening with defecation. Associated symptoms include: hematochezia, a 30 lb weight loss, and change in caliber of stool described as “pencil like”. On CT abdomen, the patient had a heterogeneously enhancing mass 7.8 x 5.4 x 6.6 cm along the sigmoid colon. Colonoscopy evaluation showed a completely obstructing mass in the sigmoid colon that was unable to traversed by the colonoscope. Pathology showed adenomatous polyp, negative for high grade dysplasia or malignancy. Due to his severe pain and large bowel obstruction on clinical presentation, as well as the clinical suspicion of malignancy, the patient underwent a Laparoscopic laparotomy where the surgeon identified retroperitoneal fibrosis which involved the left ureter and pelvic side wall along with the sigmoid colon. The dissected and resected sigmoid mass showed pathology consistent with a fibrotic tissue mass with no evidence of malignancy. This case is an unusual presentation of Retroperitoneal Fibrosis presenting as a colon mass and suspicious for malignancy. Due to its rarity we decided to share this experience as a case report and learning experience.

CASE PRESENTATION
51 y/o Hispanic male presented with severe 10/10 abdominal pain progressively worsening and associated with a change in bowel movements for several weeks. He also reported a 30 lbs. weight loss and a change in the caliber of his stool described by the patient as “pencil like” form. He had associated symptoms of hematochezia, loss of appetite and fatigue. He denied fever, chills, melena, or hematemesis. He denied a family history of colon malignancy. He has never had a previous colonoscopy before. Upon physical examination of the patient he had stable vital signs with a temperature of 36.8, respiratory rate of 18, blood pressure of 112/77, pulse of 87 and oxygen saturation of 96% at room air. He was in no acute distress and had a remarkable abdomen examination which included tenderness to left lower quadrant and suprapubic area, no masses palpated, and no rebound. HEENT, Cardiovascular, pulmonary and neurological exams were unremarkable. Laboratory data findings were: WBC 11,000, Hemoglobin 14 mg/dL, Hematocrit 41, Platelet count 279,000. The comprehensive metabolic panel was unremarkable except for an albumin of 3.3.

Imaging studies included a CT abdomen and CT thorax. The CT abdomen showed a 7.8 x 5.4 x 6.6 heterogeneously enhancing mass arising along the wall of the sigmoid colon with diffuse mural thickening noted along the descending and sigmoid colon. Mesenteric fat stranding was noted along the distal descending and sigmoid colon. The liver showed multiple well-circumscribed hypodense lesions within the hepatic segments 2, 5, 6 and 8 with the largest diameter of 1.2 cm on the lesion within hepatic segment 2. The CT thorax showed multiple non-calcified pulmonary nodules noted within the right middle and left lower lobes, measuring 0.6 cm and 0.4 cm respectively.

The Gastroenterology service was consulted for further evaluation. After examination and assessment of the patient’s case it was decided to proceed with colonoscopy to evaluate the colon mass and obtain tissue for pathological diagnosis there was a high suspicion for malignancy. Colonoscopy evaluation showed a completely obstructing sigmoid mass at 40 cm from anal verge described as friable, irregular and fungating. (Image 1) The endoscopist was unable to traverse the lesion with the colonoscope. Biopsies were taken of the lesion. Pathology showed a adenomatous polyp without findings of high-grade dysplasia or malignancy. Prior to obtaining pathology results the patient had been discharged on the basis of presumed malignancy and need to see oncology and his Primary care physician to determine further treatment plans. However, the patient returned 6 days later to the hospital due to worsening pain. Surgery was immediately involved in the case due to the presume diagnosis of malignancy, despite pathology results. Due to the clinical presentation being consistent with a large bowel obstruction the decision was made by the surgeon to proceed to the operating room for sigmoid mass resection. Operative examination revealed a large amount of adhesive tissue hard to dissect as it involved the left side pelvic wall and the left ureter along with the sigmoid colon. After careful distention of the sigmoid colon from the other anatomical structures a fresh frozen pathology sample was examined and preliminary results showed fibrotic tissue without malignancy. (Image 2) The sigmoid mass within a 22 cm long segment of sigmoid colon was removed. Sigmoid-rectal anastomosis was performed afterwards and was examined endoscopically to ensure no anastomotic leaks. The Official report of pathology of sigmoid mass confirmed a fibrotic lesion with acute

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Image 1: Irregular, friable and fungating sigmoid mass at 40 cm from anal verge seen on colonoscopy evaluation.

Image 2: Fibrotic lesion including acute/chronic inflammation with a diverticular picture to the left. Negative for neoplasia. High power magnification of the fibrotic lesion showing fibroadipose tissue array. (picture to the right)

Image 3: Fibroadipose tissue showing acute and chronic inflammation in high power magnification (picture to the left) and in low power magnification. (picture the right)

on chronic inflammation findings and crypt abscesses and crypt architectural distortion with acute inflammation, findings consistent with inflammatory colitis. (Image 3)

DISCUSSION
Retroperitoneal fibrosis is a spectrum of rare diseases with the hallmark of aberrant fibro-inflammatory tissue that most commonly develops around the infra-renal portion of the abdominal aorta and iliac arteries and frequently entraps neighboring structures such as ureters and the inferior vena cava. Rarely, other anatomical structures such as the colon may also be involved. Retroperitoneal fibrosis is subdivided into two categories either Idiopathic or Secondary. Idiopathic accounts for two thirds of the cases while the remaining third are secondary to other causes such as malignancy, infection, inflammatory disease, trauma, radiotherapy, surgery and intake of certain medications (e.g. infliximab, etanercept, hydralazine, pergolide among others). The diagnosis is mainly obtained by means of radiographic imaging (e.g. Abdomen computed tomography or magnetic resonance imaging) and biopsies. The diagnosis of this disease still remains challenging as it can clinically present as an obstruction of an organ such as the colon and often requires surgery and evaluation of full tissue to achieve the diagnosis while definitely excluding malignancy.

Secondary retroperitoneal fibrosis can be multifactorial. It has been described to involve other anatomical structures, but rarely presents as a bowel obstruction. Because of this reason we have decided to present our case as an unusual presentation of a man with a large bowel obstruction and a sigmoid mass concerning for malignancy but discovered to be fibroadipose tissue on pathological examination. In addition our patient was not taking any of the medications that have been incriminated in this entity.

Only one case similar to this, but localized to the rectum, has been described before in 1998 making this a very unusual presentation. In that Case presentation a male with a chief complaint of constipation and found on abdominal computed tomography to have a rectal mass suspicious for rectal malignancy had an exploratory laparotomy with findings consistent with retroperitoneal fibrosis. Unlike our case this patient had a needle biopsies confirming retroperitoneal fibrosis but as malignancy could not be excluded a surgical approach was performed.

Our case is also unusual, unlike the case described above, due to an underlying finding of ulcerative colitis on pathology. Could this be purely coincidence or is this a true association. It has been proposed that retroperitoneal fibrosis could be associated with ulcerative colitis as a secondary cause. Unlike our case, the three cases with ulcerative colitis and retroperitoneal fibrosis in the literature had the main presentation of ureteral involvement with urologic obstructive symptoms. In addition our patient had an otherwise normal rectum and sigmoid colon plus no past history or family history of ulcerative colitis.

One of the main problems of this entity it’s the challenge of diagnosis as it can appear radiographically and clinically concerning for malignancy. Despite technological advances there are still no specific approaches or criterion for diagnosis. Therefore many cases could end up in with surgical resection for final diagnosis. Other treatment approaches in the literature are corticosteroids medications. The aims of treatment in this disease are to induce regression of fibro-inflammatory reaction and to relieve obstruction of the ureters and others retroperitoneal structures, and to avoid recurrence. Induction therapy with the use of high doses steroids (1mg/kg/day for the first month) is the best option to curb disease activity. Reevaluation of disease after a month is needed for further treatment management with a medical management approach of prednisone tapering to 5-10 mg daily with 3 to 4 months if steroid-responsive and maintenance for an additional 6 to 9 months.

CONCLUSION
In conclusion Retroperitoneal fibrosis is a rare entity that warrants an early diagnosis in order to promptly start systemic therapy and to treat obstructive complications. Retroperitoneal

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fibrosis is a challenging diagnosis that may require exploratory or laparoscopic laparotomy in order to exclude malignancy as in our patient whose disease mimicked colon cancer. Further investigation to create a list of criteria and imaging clues that can aid in the diagnosis of this disease is needed.

REFERENCES


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