

Case Report

Angiomyolipoma of colon: unusual presentation

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ABSTRACT

Angiomyolipomas are benign mesenchymal tumors mostly arising from the kidney. Angiomyolipoma of the colon is extremely rare. Here we report the findings of a 72 years gentleman who presented with recurrent episodes of abdominal pain and fullness of one year duration. Colonoscopy was suggestive of polypoidal lesion in the descending colon. CECT abdomen revealed a colocolic intussusception in the descending colon with lipoma as a leading point. He underwent a standard left hemicolectomy. Histopathological examination showed that the tumor of 5.7 cm in diameter included smooth muscle (spindle cell type), mature adipose tissue, and vessels, and therefore a diagnosis of angiomyolipoma was made. We believe this is the second report of colonic angiomyolipoma presenting with colocolic intussusception.

Keywords: Angiomyolipomas, Intussusception, Colon

INTRODUCTION

Angiomyolipomas (AML) are benign mesenchymal tumors, first reported by Morgan et al, found mostly in the kidney.¹ Histologically they compose, thick-walled vessels, smooth muscle cells, and variable amounts of mature adipose tissue. Extra renal angiomyolipomas are uncommon, and occurrence in the gastrointestinal tract is extremely rare.² AML at various sites including colon, urinary bladder, vagina, spermatic cord, nasal cavity, ileum, skin and duodenum have been reported. Clinical symptoms vary depending upon the organ of origin. There is only one report of colonic AML presenting with intussusception. We report an elderly gentleman who presented with colocolic intussusception with histological surprise of AML on the resected specimen. We also briefly review the literature.

CASE REPORT

A 72 year gentleman reported with repeated episodes of colicky abdominal pain, without nausea or vomiting.

Clinical examination revealed soft abdomen. His laboratory profile was normal. Ultrasound abdomen showed hypo echoic mass in the left lumbar region with dilated small bowel. Colonoscopy showed large polypoidal growth in the descending colon (Figure 1A). An enhanced computed tomography (CT) scan of the abdomen revealed whorled bowel loops involving the left colon suggestive of colocolic intussusception (Figure 1B). A 3.1 × 1.5 cm well defined non enhancing soft tissue lesion within the intussusceptum was seen (Figure 1B). There was no evidence of a soft-tissue mass in the liver or in either kidney. Under the impression of a descending colon lesion combined with intussusception, the patient underwent surgery. During an exploratory laparotomy, a colo-colic intussusception involving the descending colon was found. Left hemicolectomy with hand sewn anastomosis was performed. The patient was discharged in a stable condition and is doing well on follow up.

Gross examination of the resected large intestine showed a polypoid lesion, measuring 5×4×4 cm, with a yellowish cut surface and focal ulceration. Microscopically, the

polypoid lesion revealed an admixture of three components: mature adipose tissue, thick-walled vessels, and interspersed areas of spindle-shaped smooth muscle cells (Figure 2A). The smooth muscle cells were relatively large in number, surrounded the thick-walled vessels, and were intermingled with the adipose cells. The vascular components were of varied sizes and shapes, but were most often round and thick-walled, with narrow lumens. There was no significant nuclear atypia, pleomorphism, or active mitoses in these three cell types. Immunohistochemically (IHC), the tumor cells showed positive staining for desmin. and scattered spindle cells were stained with HMB- 45 (monoclonal, 1:200, Figure 2 B). Overall, the findings characterized a colonic AML lesion.

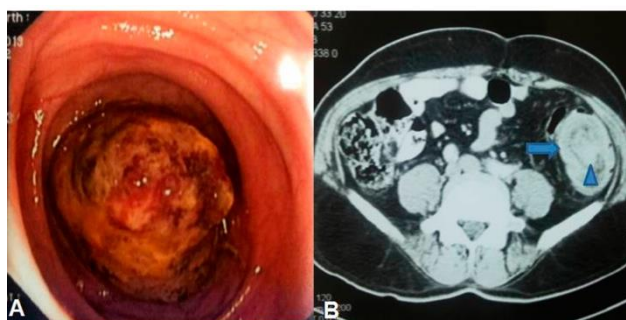


Figure 1: (A) Colonoscopic picture showing polypoid lesion; (B) CECT abdomen showing characteristic whorled appearance (blue arrow) and soft tissue lesion within the intussusceptum (blue arrow head).

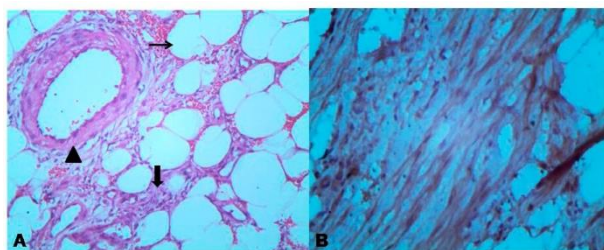


Figure 2: (A) Microscopic picture (HE stain) showing admixture of three components: mature adipose tissue (thin arrow), thick-walled vessels (arrow head), and interspersed areas of spindle-shaped smooth muscle cells (thick arrow); (B) Spindle cells stained with HMB 45.

DISCUSSION

Angiomyolipomas are benign mesenchymal tumors that arise mostly from the kidney, and are recognized as a kind of hamartoma. Renal AML are associated with tuberous sclerosis, which is a multisystemic disease with an autosomal dominant inheritance.³ AML occur in 80% of patients with tuberous sclerosis.⁴ AML consist of varying proportions of three components: smooth muscle, adipose tissue, and vessels. The tumor appears yellowish

on cut section due to fat content. The blood vessels in AML are frequently tortuous and thick walled. These patients are at risk of developing aneurysms and spontaneous bleed due to lack of elastic tissue in the vessel wall. Distinctive and pathognomonic histological findings on microscopy help to make the diagnosis of AML.

AML arising in the colon usually presents with melena, anemia, diarrhea, and abdominal pain, and may even be clinically asymptomatic.²⁻⁵ Our patient presented with abdominal pain, which was unique because it caused colocolic intussusception.

Extrarenal AML are rare, and most extrarenal AML arise from the liver.² AML of the colon are extremely rare. In our review of the literature, there have been only eight reported cases of AML of the colon.⁵⁻¹² Although there is a female predominance noted in renal AML, all of the reported colonic AML patients were middle-aged men, our patient was an elderly gentleman. Although renal AML is often associated with tuberous sclerosis, in none of the reported cases of colonic AML including ours tuberous sclerosis was noted. Seven patients were symptomatic, but one patient was asymptomatic. In all the reported cases including ours the tumors were located in left colon except in one case, where it was found in the ascending colon. The tumor sizes ranged from 1 to 5.7 cm. The tumor morphology in our case was of polypoid as noted most of the other reports. Five patients were treated with surgical resection and three were treated with an endoscopic procedure. In immunohistochemical (IHC) examinations, all of the AML of the colon reacted to SMA. Most renal AML react to HMB- 45, but AML of the colon reacting to HMB- 45 is rare. In our case the tumor cells reacted to desmin and HMB-45.

Renal AML can present with acute abdomen and shock as a result of spontaneous hemorrhage in the tumor. Yamakado et al reported that a tumor of 4 cm or larger was a predictor of spontaneous rupture.¹³ In 4.5% of angiomyolipomas, fatty component can be missed or is difficult to notice on CT scan, increasing the difficulty of pre operative diagnosis.^{14,15} On the contrary in our case the radiologist had raised the suspicion of lipomatous component in the intussusceptum.

In general, an AML is a tumor with a benign clinical course.³ However, an incomplete excision may result in local recurrence. Moreover, there have been several reports of AML with high malignant potential, known as epithelioid AML.^{16,17} These tumors have the potential for local invasion and distant metastases, but the involvement of regional lymph nodes by these tumors is uncommon.³ Kawaguchi et al reported that the IHC findings of SMA were positive in 30% of typical angiomyolipomas but that they were negative in malignant types.¹⁶ Since all tumors of AML of the colon are positive for SMA, it is thought that the tumors are typical AML, not malignant types. Due to the small number of cases of colonic AML, there

is still no established treatment. At present, we believe that AML of the colon should be treated by complete resections of the tumor. Although there is no evidence of local recurrence, these patients should be followed regularly.

In summary, we report a case of AML of the colon that we believe to be the second case presenting with colocolic intussusception. Although pre operative suspicion can be made, it is often difficult to differentiate it from malignancy. Surgical resection should be considered due to the risk of hemorrhage and obstruction.

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