Ileal angiomyolipoma manifested by small intestinal intussusception

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Abstract

Angiomyolipomas (AMLs), benign mesenchymal hamartomas, arise primarily in the kidneys of patients with or without tuberous sclerosis. Extra-renal AMLs are very rare and are most commonly found in the liver. AMLs of the small intestine are exceedingly rare. Here, a case of a 28-year-old man, who presented with ileal intussusception caused by ileal AML is reported. The clinicopathological and immunohistochemical findings of ileal AMLs are discussed and the literature on small intestinal AMLs is reviewed.

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Key words: Angiomyolipoma; Intussusception; Hamartoma; Ileum; Colectomy

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CASE REPORT

A 28-year-old man presented to the emergency room complaining of progressive abdominal cramping pain, accompanied by nausea and vomiting for 6 h. He had no significant medical history except for having been admitted with pneumothorax 13 years previously. The pain was located over the right lower quadrant of the abdomen. He had no change in bowel habits, bloody stools, or tenesmus. Physical examination showed a palpable mass over the right lower quadrant with direct tenderness. Increased bowel sounds were noted during auscultation. Laboratory findings were normal; red blood cell count 5.02 × 10¹²/mm³, hemoglobin 15.7 g/dL and hematocrit 44.1%, platelets 237 × 10⁹/mm³, and white blood cell count 7700/mm³. Serum analysis was as follows: total protein 8.6 g/dL, aspartate aminotransferase 19 U/L, alanine aminotransferase 13 U/L, blood urea nitrogen 13.5 mg/dL, and creatinine 0.7 mg/dL. An enhanced computed tomography (CT) scan of the abdomen showed ileocolic intussusception caused by an abnormal 3.0-cm soft tissue mass with increased thickness of the wall (Figure 1A). There was no evidence of any other soft tissue mass either in the liver or in the kidney. Colonoscopic examination could not be performed because of severe abdominal pain. The patient underwent surgery with the diagnosis of intussusception. During an exploratory laparotomy, the main polypoid mass was located approximately 60 cm from the ileocecal valve. An ileo-ileal intussusception, about 15 cm in length, was found. This mass protruded into the right colon, including the cecum. A right hemicolectomy was uneventfully performed.

Grossly, an approximately 3 cm × 3 cm × 2.5 cm sized polypoid mass was located in the ileum, 60 cm from the ileocecal valve. Other than this mass, no other soft tissue mass was found. The ileum was resected en bloc with the cecum, appendix, and ascending colon (Figure 1B). There were no soft tissue masses found around the liver or kidney. A right hemicolectomy was uneventfully performed.

INTRODUCTION

Angiomyolipomas (AMLs), benign mesenchymal tumors, are composed of blood vessels, smooth muscle cells, and mature fat cells. These mesenchymal hamartomas arise primarily in the kidney[1] and extrarenal AMLs are very rare. AMLs of the small intestine are exceedingly rare and, to the best of our knowledge, only three cases have been reported in the literature[2,3]. Herein, a case of ileal AML, manifested by small intestinal intussusception, is presented. This occurred in a 28-year-old man, and was confirmed by microscopic examination and immunohistochemical staining after right hemicolectomy.
ileocecal valve, and many small polyps were located in the ileum about 25 cm from the ileocecal valve (Figure 1B). The cut surface of the main mass was grayish-yellow in color, with a lobulated appearance and exhibiting no hemorrhage or necrosis. Microscopically, it revealed a mixture of three components: mature adipose tissue, thick-walled vessels, and interspersed areas of spindle-shaped smooth muscle cells (Figure 2A). Immunohistochemically, the smooth muscle cell components of this lesion were consistently positive for α-smooth muscle actin (SMA: Clone 1A4; dilution 1:100; Dako; Figure 2B), desmin (Clone D33; dilution 1:200; Dako; Figure 2C), and vimentin (Clone V9; dilution 1:100; Novocastra). The vascular components were immunoreactive for CD34 (Clone QBEnd/10; dilution 1:50; Novocastra; Figure 2D), but tumor cells were negative for HMB-45 (Novocastra, 1:60) and C-kit (Clone 104D2; dilution, 1:200; Dako). The final pathological diagnosis was AML. The numerous small polyps were diagnosed as lymphoid polyps. The postoperative course was uncomplicated, and the patient was discharged 8 d later. He was seen in the outpatient department for follow-up, and had no tumor recurrence over the subsequent 6 mo.

**DISCUSSION**

AMLs are histologically benign tumors derived from mesenchymal tissue. The vast majority of AMLs arise in the kidney. In 45 to 80% of patients, they are associated with tuberous sclerosis, a multi-systemic disease with autosomal dominant inheritance, and the occasional association with the triad of epilepsy, mental retardation, and adenoma sebaceum[1]. Extrarenal AMLs are very rare and have been reported in the liver[4], nasal cavity[5], vagina[6], spermatic cord[7], skin[8], mediastinum[9], and gastrointestinal (GI) tract, including the colon[10-13] and small intestine[12,13]. AMLs arising in the GI tract are extremely uncommon and usually present with melena, anemia, diarrhea, abdominal pain, and may even be clinically asymptomatic[10-13]. Radiological diagnosis of extrarenal AMLs is difficult because of the rarity of the condition. CT is an effective means of imaging and identifying AMLs. Thin-section (3-5 mm) scanning is performed in an attempt to show fatty-tissue attenuation. Sonography can suggest the diagnosis, showing a well-defined hyperechoic lesion, but it is not diagnostic. On magnetic resonance imaging, lesions are bright on T1-
Table 1  Clinicopathological data and immunohistochemical staining results in small-intestinal AMLs

| Case No. | 1 | 2 | 3 | 4 |
|----------|---|---|---|---|
| Author   | Han et al | Toye and Czarnecki | Lin et al | Present |
| Age (yr) | 60 | 60 | 48 | 28 |
| Gender   | Male | Female | Female | Male |
| Tuberous | Absence | Absence | Absence | Absence |
| sclerosis | | | | |
| Symptoms | Polypoid | Polypoid | Pedunculated polypoid | Polypoid |
| Intussusception | Present | Absent | Present | Present |
| Location | Duodenum | Ileum | Ileum | Ileum |
| Size (cm) | 4 × 4 × 3 | 3.6 × 3.6 | 4 × 4 × 2 | 3 × 3 × 2.5 |
| Gross morphology | Polyoid | Polyoid | Pedunculated polypoid | Polyoid |
| IHC       | SMA | + | + | + |
|           | DES | + | NA | + |
|           | VIM | + | NA | + |
|           | CD34 | NA | NA | + |
|           | HMB-45 | - | - | + |
|           | CD117 (c-kit) | - | NA | + |

DES: Desmin; VIM: Vimentin; +: Positive; -: Negative; NA: Data not available; ++: Positive in few cells.

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