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Case report

Ileal angiomyolipoma as an unusual cause of small-intestinal intussusception

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Angiomyolipomas are benign mesenchymal tumors, but those that arise from the small intestine are exceedingly rare. We report on a 48-year-old woman who had an ileal angiomyolipoma, who presented clinically with vague abdominal pain and bloody stool. Small-bowel intussusception was shown on an abdominal computed tomography (CT) scan. We discuss the clinical manifestations and clinicopathological and immunohistochemical findings of this benign tumor which appeared in this rare location.

Key words: ileum, angiomyolipoma, intussusception

Introduction

Angiomyolipomas (AMLs), benign mesenchymal tumors, were first described by Morgan et al.,[1] in 1951, as renal tumor lesions, characterized by thick-walled vessels, smooth muscle extending outward from the vessels in a peritheliomatous fashion, and mature adipose tissue. Extrarenal AMLs are rarely reported, but locations have included the colon, urinary bladder, vagina, spermatic cord, nasal cavity, skin, and duodenum.[2–11] To the best of our knowledge, no AML arising in the ileum has previously been reported. Herein, we present the first case of an ileal AML. This occurred in a 48-year-old woman, and was confirmed by microscopic examination and immunohistochemical staining, after segmental resection.

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Case report

A 48-year-old woman was admitted to our hospital due to progressive aggravation of intermittent bloody stool for 1 week, without nausea or vomiting. On admission, physical examination showed a flat, soft abdomen, without tenderness or a palpable mass over the abdomen. Normal active bowel sounds were noted during auscultation. Blood test revealed hemoglobin, 10.6 g/dl and hematocrit, 33.4%. Results of a colonoscopic examination and a small-bowel series were negative. An enhanced computed tomography (CT) scan of the abdomen revealed a soft-tissue mass in the terminal ileum, measuring 3 cm in its longest dimension, with an ilio-ileum intussusception (Fig. 1A). There was no evidence of a soft-tissue mass in the liver or in either kidney. Under the impression of a small-bowel tumor combined with intussusception, the patient underwent surgery. During an exploratory laparotomy, a soft-tissue mass was located approximately 20 cm from the ileocecal valve, and an ilio-ileum intussusception, about 15 cm in length, was found. Total resection of the affected small-bowel loop, with end-to-end anastomosis, was performed uneventfully. The patient was discharged in a stable condition, and no recurrence has been noted during 1.5 years of follow up.

Gross examination of the resected small intestine showed a pedunculated polypoid lesion, measuring 4 × 2 × 2-cm, with a grayish-to-yellowish cut surface and focal ulceration (Fig. 1B). 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 (Fig. 1C). The smooth muscle cells were relatively large in number, surrounded the thick-walled vessels, and were intermingled with the adipose cells (Fig. 1D), either in bundles or individually; the smooth muscle cells were confirmed by Masson's trichrome stain. The vascular components were of varied sizes and shapes,

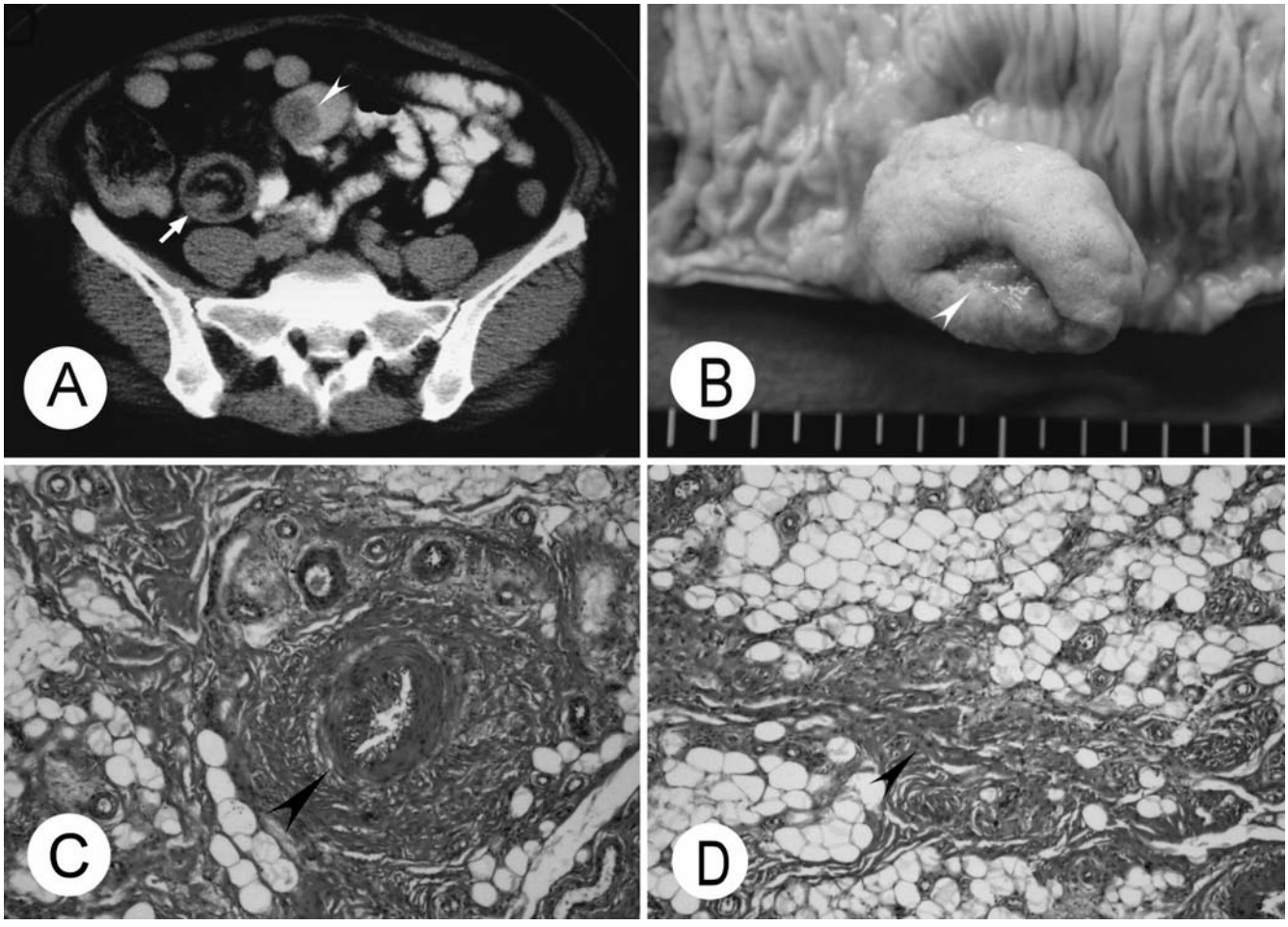

Fig. 1. **A** Enhanced computed tomography (CT) scan of the abdomen showed a soft-tissue mass (*arrowhead*) in the terminal ileum, with intussception (*arrow*). **B** Grossly, the pedunculated polypoid tumor measured 4 × 2 × 2 cm, and had a central ulcerative lesion (*arrowhead*). **C** Photomicrography of the tumor revealed an admixture of three components; thick-walled vessels, areas of interspersed proliferative smooth muscle cells (*arrowhead*), and mature adipose tissues. **D** The adipose tissue cells were intermingled with smooth muscle cells (*arrowhead*). **C** and **D** H&E, ×40

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, the tumor cells showed positive staining for desmin (monoclonal, 1:200; Dako, Copenhagen, Denmark; Fig. 2A), α-smooth muscle actin (SMA; monoclonal, 1:200; Dako; Fig. 2B), and vimentin (monoclonal, 1:200; Biogenex, San Ramon, CA, USA). The vascular components were immunoreactive for CD34 (monoclonal, 1:100; Dako; Fig. 2C), and only scattered tumor cells were stained with HMB-45 (monoclonal, 1:200, Dako; Fig. 2D) and c-*kit* (CD117; monoclonal, 1:200, Dako). Overall, the findings characterized an ileal AML lesion.

Discussion

AMLs are hamartomas which mostly arise from the kidneys and appear in solitary or multicentric mass forms.[12] On occasion, a pedunculated growth or satellite nodule outside the renal capsule pattern can be identified. Approximately one-third of patients with renal AML are associated with tuberous sclerosis complex.[12]

Extrarenal AMLs are a rare event. Clinical symptoms vary and are related to the organ that is involved. An 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 bloody diarrhea, which was unique because it caused intestinal intussusception.

In a review of the previous literature, we found only five cases of intestinal AMLs, four in the colon and one

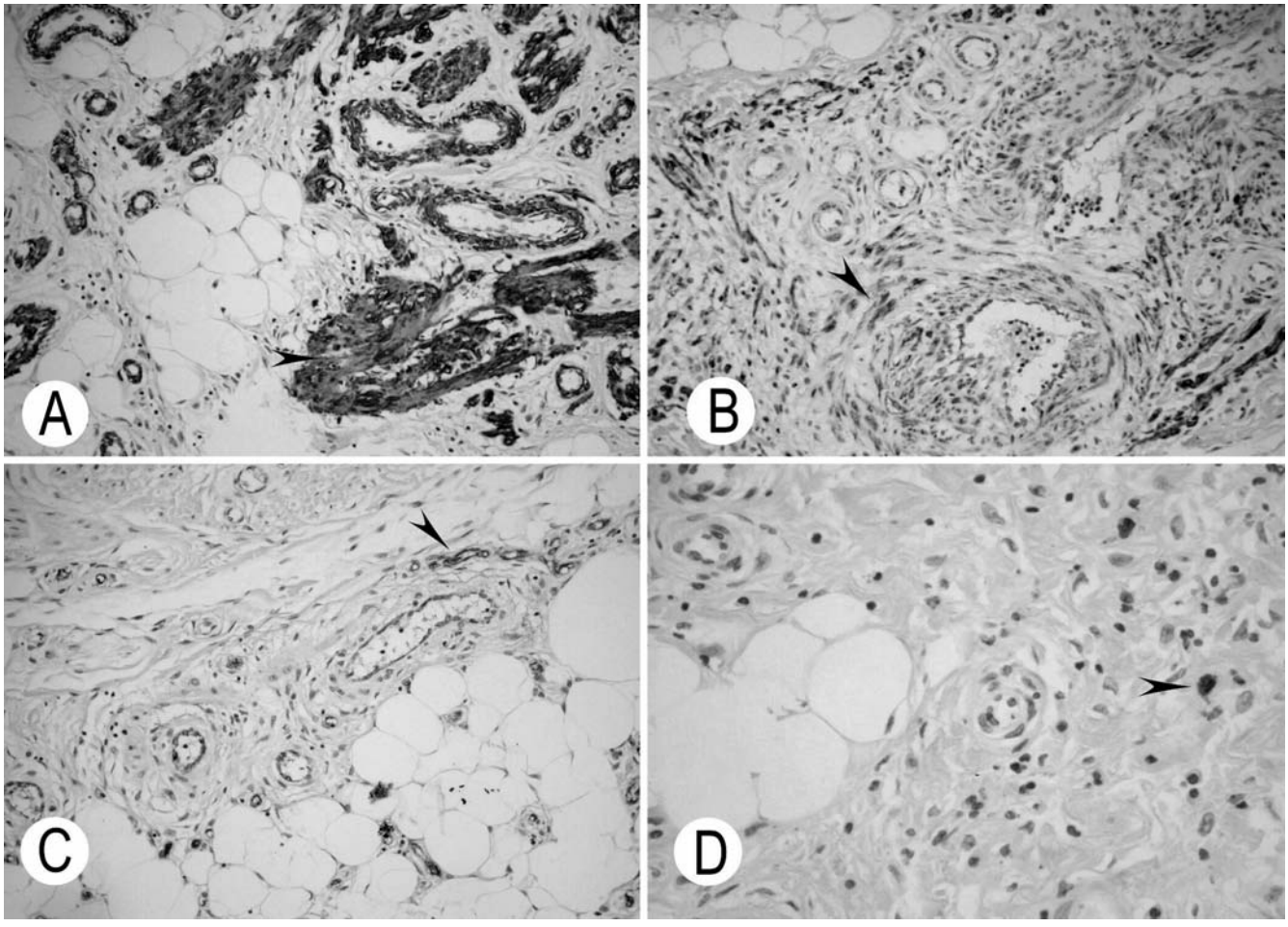

Fig. 2. **A** Thick-walled vessels surrounding smooth muscle cells show strong immunoreactivity for desmin (*arrowhead*). **B** A few interspersed spindle-shaped tumor cells (*arrowhead*) display immunoreactivity for smooth muscle actin (SMA). **C** Immunoreactivity for CD34 is found only in vascular components (*arrowhead*). **D** Scattered tumor cells (*arrowhead*) show immunoreactivity for HMB-45. **A,B,C,D** Immunostaining; **A,B,C** ×100; **D** ×200

in the duodenum. Table 1 summarizes the clinicopathologic data and immunohistochemical staining results of these lesions. All four colonic AML patients were male, while the patient with the duodenal lesion and the present patient were female. The mean age at diagnosis was 55.6 years. The tumor sizes ranged from 1 to 4 cm.[2–5,11] Grossly, all of the lesions appeared in single polypoid, pedunculated,[3–5,11] or sessile[2] patterns. Focal ulceration was seen in three patients. There was no evidence of multiple lesions in any patient. Moreover, none of these patients, including the present one, were associated with tuberous sclerosis complex.[2–5,11]

Immunohistochemically, the smooth-muscle cell components consistently revealed immunoreactivity for desmin, SMA, and vimentin.[2–4,11] However, HMB-45 was positive in only two of the previously reported patients,[2,4] while it was negative in the other two.[3,11] The present patient showed only scattered HMB-45-immunoreactive tumor cells. These results are in contrast to the renal and liver lesions, which usually show immunoreactivity for HMB-45. CD34, cytokeratin, and S-100 were found to be negative in all stained lesions in the previously reported patients.[2–4,11] Antibodies to CD117 have not been well-documented for AMLs until recently; Makhlouf et al.[13] reported that all 15 of their patients with hepatic AML and 6 of their patients with renal AML showed positive results. In the present patient, we found only scattered spindle-shaped tumor cells showing immunoreactivity for CD117.

Microscopically, the characteristic features of AML include an admixture of adipose tissue, smooth muscle cells, and thick-walled vessels, in varying proportions. On occasion, destructive growth into the muscular layer and foci of spindle-shaped tumor cells with an epithelioid appearance, showing nuclear atypia, bizarre nuclei, and mitoses may be present, and this should not be

Table 1. Clinicopathologic data and immunohistochemical staining results in intestinal AML

Case no.	1	2	3	4	5	6
Author	Maluf and Dieckgraefe[2]	Maesawa et al.[3]	Chen et al.[4]	Hikasa et al.[5]	Toye and Czarnecki[11]	Present patient
Sex	Male	Male	Male	Male	Female	Female
Age (years)	55	50	54	67	60	48
Symptoms	Bloody stool	Diarrhea	Abdominal pain	Asymptomatic	Satiety	Intussusception
Location	Ascending colon	Descending colon	Splenic flexure	Sigmoid colon	Duodenum	Ileum
Size (cm)	1	3 × 2.5 × 2.5	3.6 × 3 × 3	1 × 0.8 × 0.7	3.6 × 3.6	4 × 2 × 2
Gross	Sessile	Polyp, ulcerated	Polyp	Polyp	Polyp, ulcerated	Polyp, ulcerated
Treatment	Subtotal colectomy	Partial colectomy	Extended left hemicolectomy	Polypectomy	Segmental resection	Segmental resection
IHC						
VIM	+	+	+	NA	NA	+
SMA	+	+	+	NA	+	+
DES	+	+	+	NA	NA	+
HMB-45	+	−	+	NA	−	+*
CD34	−	NA	−	NA	NA	+**
S-100	NA	−	NA	NA	−	−
CD117	NA	NA	NA	NA	NA	+*

IHC, immunohistochemical staining; VIM, vimentin; SMA, α-smooth muscle actin; DES, desmin; +, positive; −, negative; NA, data not available; +*, positive in few cells; +**, positive in vascular component

misdiagnosed as a malignancy.[2,3] Theoretically, intraoperative diagnosis of the lesion can be made by frozen sections of the polypoid lesions, to identify the occurrence of these three tissue components.

AMLs usually grow slowly. Malignant changes of AMLs are rare, and only sporadically reported cases have been documented in lesions arising in the kidney.[12] Most AMLs that show malignant change are pleomorphic AMLs that reveal the presence of marked cellular pleomorphism and intravascular growth, which eventually progresses, with leiomyosarcomatous transformation and subsequent metastasis. However, to date, no occurrence of malignant change in an intestinal AML has been reported. Surgical resection remains the treatment of choice, and produces an excellent prognosis.[2–5,11] The present patient has shown no evidence of recurrence for 1.5 years after surgery.

In conclusion, intestinal AML is rare; when a patient presents with bloody diarrhea associated with ileal intussusception, AML should be included in the differential diagnosis.

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