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Angiomyolipoma of the Jejunum Mimicking Metastatic Disease in a Patient with Colonic Adenocarcinoma

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Abstract

Angiomyolipoma is a benign tumor most commonly arising in the kidney. Very few cases have been reported to be located in the small intestine. Here we report the first case located in the jejunum in a patient who was diagnosed with a colonic adenocarcinoma. In the preoperative evaluation this benign lesion was thought it might represent a metastatic nodule.

Keywords: Angiomyolipoma, Colon Adenocarcinoma, Jejunum

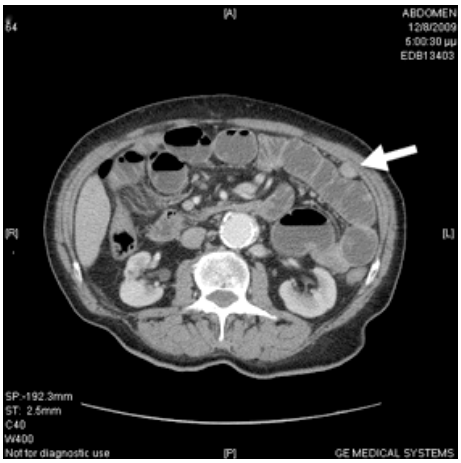
1. Introduction

Angiomyolipoma (AML) is a benign neoplasm composed of blood vessels, smooth muscle cells, and mature fat cells [1]. AML most commonly arises in the kidney. Other locations include the liver (most frequent extrarenal site) [2], the uterus[3], the lungs [4], the skin [5], the subgaleal space [6], the anterior mediastinum [7], the urinary bladder [8], and mucosal membranes such as the lips [9] and the nasal cavity [10]. Very few cases have been reported in the gastrointestinal tract, including nine cases arising from the colon [11-19], and four cases arising from the small bowel [20-23]. Two of the latter cases were located in the duodenum [20,21], and the other two in the ileum [22,23]. In this paper we report a fifth case of AML of the small intestine, this time located at the jejunum. The benign small intestinal tumor was masquerading a metastatic nodule in the preoperative imaging evaluation, since our patient also presented a tumor in the ileocecal region.

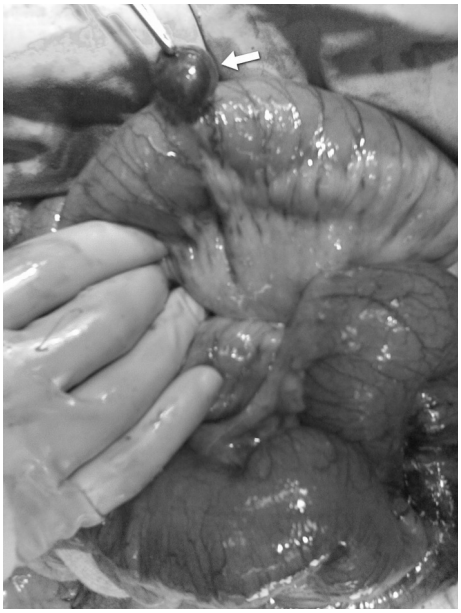
2. Case Report

An 80 years old male was urgently admitted to our surgical department with abdominal pain, vomiting and abdominal distension. On clinical examination the patient was afebrile and the abdomen was moderately distended, with mild tenderness on palpation of the right iliac fossa, but no signs of peritonism. Bowel sounds were reduced and digital examination of the rectum was normal. Plain abdominal films confirmed small bowel obstruction and full blood count and biochemistry were unremarkable. Conservative treatment was initiated, including intravenous fluids, nasogastric suction and monitoring of the vital organs. After three days of conservative treatment, although there was no clinical deterioration, the obstructive ileus was not resolved and a colonoscopy was performed, to further investigate the etiology of obstruction, which revealed an obstructive tumor of the ileocecal valve. A preoperative CT scan of the abdomen performed the next day for staging purposes showed regional lymphadenopathy of the mesentery of the ileocecal region, but no hepatic metastases. However, a soft tissue mass was found in the proximal jejunum with most probable diagnosis to be metastatic implant (**Figure 1(a)**).

In order to resolve the persisting ileus, an exploratory laparotomy was decided and performed through a high suprasubumbilical incision. A large obstructive tumor of the ileocecal valve with regional lymphadenopathy were detected with no signs of hepatic or other metastatic disease. However, a round, well circumscribed tumor of the antimesenteric border of the proximal jejunum was detected (**Figure 1(b)**), which was excised in healthy tissues macroscopically through a limited small bowel enterectomy. A typical right hemicolectomy was then performed with ileocolic anastomosis to the transverse colon. Following an uneventful postoperative period, and after gradual mobilization and feeding, the patient was discharged home on the seventh postoperative with instructions for regular follow-up and referral to the medical



(a)



(b)

Figure 1. (a) CT scan of the abdominal area showing a nodule attached to the jejunum (arrow); (b) The surgical specimen of the nodule of the jejunum as found at the operation theater (arrow).

oncology department for the colon cancer. The patient refused any further treatment, but still today, 12 months after operation, is alive and well.

2.1. Pathological Findings

The hemicolectomy specimen consisted of a segment of the terminal segment of ileum 22 cm in length, the ileocecal valve, the cecum along with the appendix, and a segment of the ascending colon 16 cm in length. A large exophytic and invasive tumor, 4 cm in diameter was found in the ascending colon, close to the ileocecal valve. 20 lymph nodes, 0.3-1.2 cm in diameter were removed from the surrounding pericolic fat. On microscopy this tumor proved to be a well-differentiated adenocarcinoma of the large intestine, invading the whole thickness of the intestinal wall, and extending to the pericolic fat. One of the removed lymph nodes presented metastasis from the aforementioned adenocarinoma. The tumor of the jejunum was reddish, and soft, 2 cm in diameter. It was composed by spindle cells arranged in fascicles, vacuolated (fat) cells and many blood vessels (**Figure 2(a)**). No significant nuclear atypia, mitotic activity, necroses or increased cellularity were detected. In order to sustain a definitive diagnosis, immunohistochemical evaluation followed.

Immunohistochemistry was performed on formalin-fixed, paraffin-embedded tissue sections, 3-μm in thickness. High temperature antigen unmasking in electric pressure cooker, and TrilogyTM solution (Cell Marque, Rocklin, CA, USA) pretreatment preceded the main procedure. A standardised automated (Nexes, Ventana, Tuscon, AZ, USA), streptavidin-biotin method (I-VIEW Paraffin DAB, Ventana) followed the application of the monoclonal antibodies. The latter were directed against HMB-45 (dilution 1:50, Dako, Glostrup, Denmark), Melan-A (dilution 1:50, Dako), a-smooth muscle actin (SMA, dilution 1:40, Novocastra, Newcastle, UK), Desmin (dilution 1:100, Novocastra), S-100 (dilution 1:300, Dako), CD117 (dilution 1:50, Dako), and CD34 (dilution 1:50, Dako). The tumor cells were positive to HMB-45 (**Figure 2(b)**), Melan-A, SMA (**Figure 2(c)**), and desmin, while they were negative to CD117, and CD34 (except of the endothelial cells of the vessels, **Figure 2(d)**). Our findings in total were consistent with a diagnosis of angiomyolipoma.

3. Discussion

Traditionally, AML was considered as hamartoma. However, AML today is regarded as a true neoplasm, since it has been found to present clonal nature [24]. Moreover, AML is thought to arise from the perivascular epithelioid cell (PEC), which has certain morphologic and immunohistochemical characteristics, even though no known normal cell counterpart has been found until now [1]. Immunohistochemically, PEC expresses myogenic and melanocytic markers, such as HMB-45, HMSA-1, MelanA/Mart1, microophtalmia transcription factor (Mitf), actin and, less commonly, desmin. Other (rare) tumors which arise from PEC include clear-cell "sugar" tumor of the lung and extrapulmonary sites, lymphangioleiomyomatosis, clear-cell myomelanocytic tumor of the falciform ligament/ligamentum teres, and rare clear-cell tumors of other anatomical sites. In addition, some PEComas are related to the tuberous sclerosis

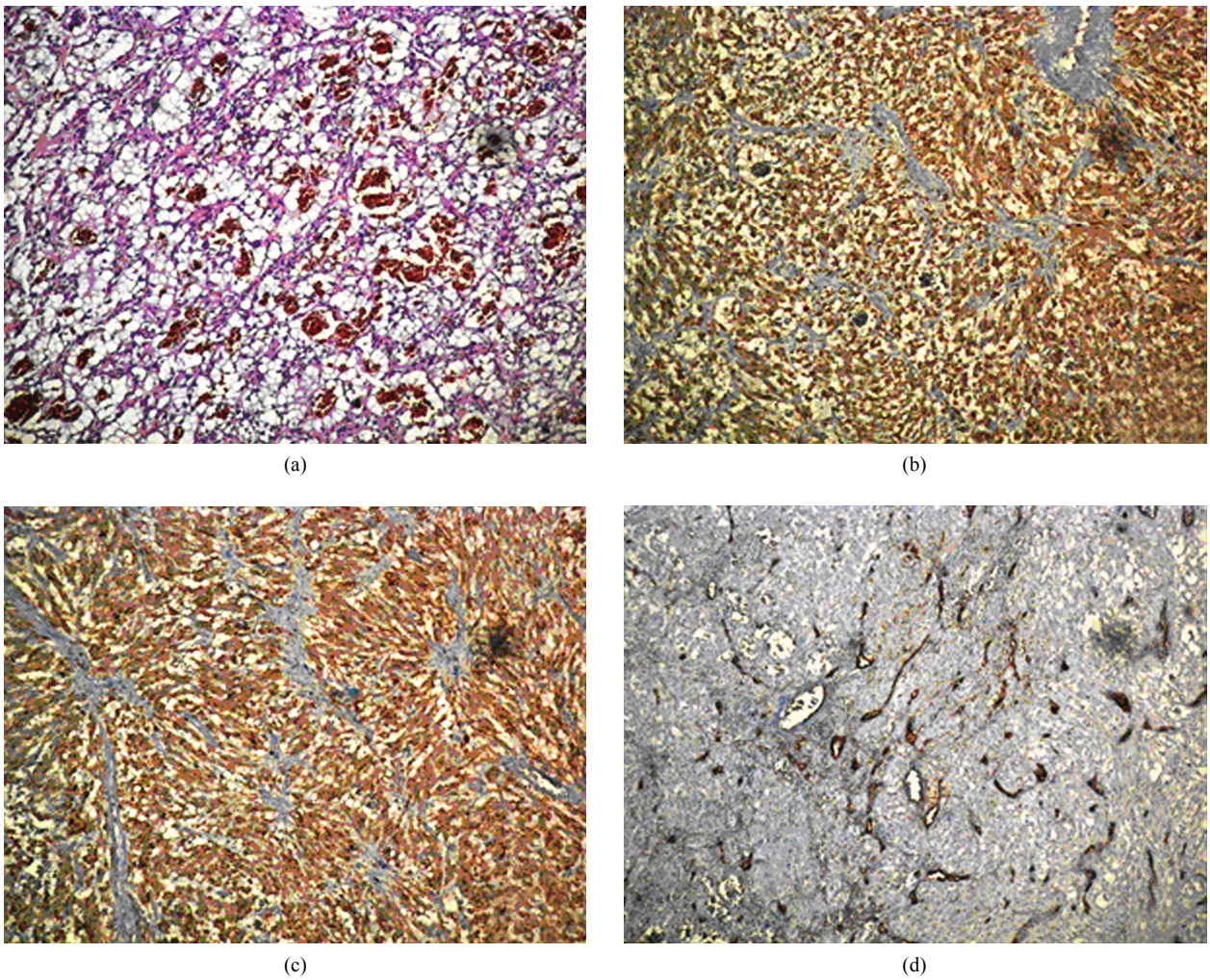


Figure 2. (a) The tumor is composed of fascicles of spindle cells, interspersed with groups of vacuolated fat cells and numerous blood vessels (H&E, X100); (b) The spindle cells of the lesion are strongly positive to HMB-45 (DAB, Hematoxylin, X100); (c) The spindle cells of the lesion are strongly positive to smooth muscle actin (DAB, Hematoxylin, X100); (d) Only blood vessels show positive reaction to CD34 (DAB, Hematoxylin, X100).

complex (TSC), an autosomal dominant genetic disease due to losses of TSC1 (9q34) or TSC2 (16p13.3) genes [25,26]. This syndrome is characterized by mental retardation, seizures and cellular proliferations (AMLs, subependymal giant cell tumors, cutaneous angiofibromas, cardiac rabdomyomas, lymphangioleiomyomatosis, pulmonary multifocal micronodular hyperplasia). Similar alterations of the TSC genes have been demonstrated in a significant number of PEComas, both occurring within the TSC and in sporadic cases.

Sporadic AMLs occur in older patients, in the fourth to sixth decades of life, with a female predominance; they are single, unilateral and larger than those associated with TSC [27]. Classic AML contains more than one cell type; if a particular cell type predominates, AML is consequently named (lipoma-like AML or leiomyoma-like AML). AMLs arising in the GI tract are extremely uncommon and usually present with melena, anemia, diarrhea, abdominal pain, and may even be clinically asymptomatic [11-23]. Radiological diagnosis of extrarenal AMLs is difficult because of the rarity of the condition. Even though four other angiomyolipoma cases have been reported to affect small intestine, our case is unique, since to our knowledge is the first to involve jejunum. In addition, in our case the intestinal nodule was thought to represent metastatic disease in the pre-operative evaluation, because our patient was having a colonic tumor as well. So, it represented a critical differential diagnostic problem in terms of severity, and staging of his primary disease. Moreover, we consider ours to be a genuine AML case, since it is both HMB-45, and Melan-A positive. At least two cases of small intestinal AML reported in the literature [8,9] were HMB-45 negative (**Table 1**), when HMB-45 positive immureaction

Table 1. Angiomyolipomas of the small intestine reported in the literature: Immunohistochemical reaction to various antibodies.

Author	SMA	Desm	KIT	CD34	Melan-A	HMB-45	S-100	Vim
De Padua *et al.*	+	NA	NA	NA	NA	+	NA	NA
Toye and Czarnecki	+	NA	NA	NA	NA	-	-	NA
Lee *et al.*	+	+	-	+	NA	-	NA	NA
Lin *et al.*	+	+	+	+	+	+	NA	+
Miliaras and Miliaras	+	-	-	-	+	+	-	NA

SMA = smooth muscle actin, Desm = desmin, Vim = vimentin, NA = not available.

is currently considered as a prerequisite for such a diagnosis [1,14]. In conclusion, intestinal AMLs are very rare, and may cause various symptoms or mimick other conditions as metastatic disease in our case. For this reason it is quite important to differentiate such a case from other mesenchymal small intestinal tumors, and especially from gastrointestinal stromal tumor, which is the most frequent tumor in this location and merits specialized targeted immunotherapy.

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