Angiomyolipoma of the Colon

Report of a Case and Review of the Literature

Jinn-Shiun Chen, M.D.,* Li-Jen Kuo, M.D.,* Paul-Yann Lin, M.D.,† Chung-Rong Changchien, M.D.*

From the *Division of Colon and Rectal Surgery and †Department of Pathology, Chang Gung Memorial Hospital, Taipei, Taiwan

Angiomyolipoma of the colon is very rare. Only three cases have been reported in the literature. Here we report the case of a 54-year-old male, who presented with a progressive abdominal cramping pain. We performed left hemicolectomy under the impression of a cancerous mass over the splenic flexure of the colon. Histology revealed an angiomyolipoma of the colon. In addition, we review the literature. [Key words: Angiomyolipoma; Colon; Immunohistochemistry]


Angiomyolipomas, a form of mesenchymal hamartoma, are composed of blood vessels, smooth muscle cells, and mature fat cells. The great majority of these tumors occur in the kidney.1 Extrarenal angiomyolipomas are extremely rare and have been reported in the liver,2 nasal cavity,3 vagina,4 spermatic cord,5 skin,6 and mediastinum.7 Preoperative diagnosis is usually difficult and surgical excision is required. Inadequate resection may result in rapid local recurrence.8,9

Report of a Case

A 54-year-old male patient came to our clinic complaining of a two-month history of abdominal pain. He had been well before with no underlying disease such as hypertension, renal, hepatic, or cardiac disease. The pain was located over the left side of the abdomen with cramping characteristics. He had no bowel habit change, tenesmus, or body weight loss. Physical examination showed a palpable mass over the left upper quadrant with tenderness. Digital examination and rigid sigmoidoscopy was performed in the outpatient department, and the results were unremarkable. Colonofiberscope examination and a barium enema revealed a large polypoid mass over the splenic flexure (Figs. 1A and B). Abdominal computed tomography showed an abnormal soft tissue mass lesion with increased thickness of the wall and narrowed lumen over the splenic flexure of the colon (Fig. 1C). The carcinoembryonic antigen level was 2.02 ng/ml. Extended left hemicolectomy was performed. Histology revealed an angiomyolipoma (Fig. 2). Immunohistochemistry staining showed the proliferating smooth muscle cells were positive for HMB-45, desmin, vimentin, and smooth muscle actin and negative for CD-34 and cytokeratin. The postoperative course was uncomplicated, and he was discharged seven days later. He was followed up in the outpatient department and had no tumor recurrence during an 18-month follow-up.

DISCUSSION

Angiomyolipomas are histologically benign tumors derived from mesenchymal tissue. Grossly, they are grayish-yellow with a lobulated appearance and vary in size. The great majority of angiomyolipomas arise in the kidney, and in 45 to 80 percent of patients, they are associated with tuberous sclerosis, a multisystemic disease with autosomal dominant inheritance, with occasional association with the triad of epilepsy, mental retardation, and adenoma sebaceum.1 Extrarenal presentations are very rare and are most commonly found in the liver.2

Preoperative diagnosis is difficult; with a combination of abdominal computed tomography, ultrasound, and magnetic resonance imaging, the diagnostic rate increases to 60 percent.9 Surgical excision is the treatment of choice. The recurrence rate is high in cases of inadequate resection, but when the tumor can be removed completely, the prognosis is excellent, with a nearly 100 percent cure rate.9,10 Microscopically, angiomyolipoma is composed of three components: smooth muscles, adipose tissues and blood vessels. The contents of these components varied in proportion, especially in adipose tissue, and range from less than 10 percent to more than 50 percent of the tumor.
Because of these characteristics, variable imaging features and histologic pictures were noted. Cellular pleomorphism or hyperchromatic appearance is common in smooth muscle cells, and mitosis and multinucleated giant cells may also be present. These features are conducive to an erroneous diagnosis of malignant tumor. The vascular component was composed of tortuous, thick-walled blood vessels with perivascular cell proliferation. The presence of cells intermediate or transitional between lipocytes and smooth muscle cells in the tumor support the hypothesis that the distinctive epithelioid cells are mesenchymal precursor cells that have the ability to differentiate into both myoid and fat cells.\(^2\)\(^1\)\(^1\)\(^1\) Immunohistochemical study of the smooth muscle component of this lesion was positive for HMB-45, desmin, vimentin, and smooth muscle actin, and negative for CD-34 and cytokeratin. Chan and colleagues\(^1\)\(^2\) analyzed 20 cases of renal angiomyolipoma and reported positive results for muscle specific actin, desmin, and HMB-45 in 20, 17, and 18 cases respectively.

Only three cases of angiomyolipoma of the colon have been previously reported.\(^1\)\(^3\)\(^-\)\(^5\) All of the reported cases of colonic angiomyolipoma are summarized in Table 1. We noted several interesting findings in these colonic angiomyolipoma cases. First, all of