Sonographic and Computed Tomography Findings of Intra-abdominal Desmoid Tumor

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Intra-abdominal desmoid tumor is rare and seldom reported in the literature. It can grow large before causing symptoms, such as obstructing bowel loops or urinary system. Here, we present a 29-year-old male who suffered from intra-abdominal desmoid tumor, and describe its imaging findings on ultrasound and abdominal computed tomography. This tumor usually presents as a large homogeneous hypodense solid mass on computed tomography, and demonstrates mild enhancement after contrast medium administration. Although rare, it should be included in the differential diagnosis when a patient presents with a large abdominal mass. [*J Chin Med Assoc* 2010;73(7):393–395]

Key Words: computed tomography, desmoid tumor, familial adenomatous polyposis, sonography

Introduction

Desmoid tumor is rare and can develop in the extremities, chest wall and abdominal wall.[1] However, the abdominal cavity is another common location of this tumor. We report a case of intra-abdominal desmoid tumor, describing its imaging findings on ultrasound (US) and computed tomography (CT).

Case Report

A 29-year-old male, without any previous systemic disease, suffered from gastric ulcer and hemorrhoid for about 2 years. Intermittent hematochezia off and on was noted for 1 year. However, he did not seek any medical help. A large, firm, and growing palpable mass lesion was noted in the upper abdomen. There were no other symptoms such as pain, tenderness, nausea, vomiting, diarrhea or constipation.

He visited our hospital for help and abdominal sonography (Figure 1) revealed a 15-cm homogeneous hypoechoic mass lesion in the upper abdomen. Color Doppler study did not depict any prominent vessels within the tumor. Abdominal CT (Figure 2) was arranged and revealed a 19-cm homogeneous hypodense mass lesion arising from the mesentery in the midline of the upper abdomen, with mass effect deviating the bowel loops laterally, and with mild enhancement after contrast media administration. No cystic change or calcification could be identified. Surgery was performed and a well-encapsulated, firm, yellowish mass

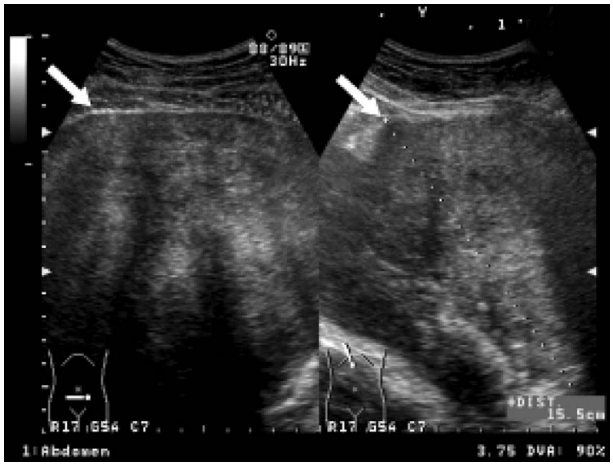

Figure 1. Abdominal sonography revealed a large mass lesion (arrows) in the midline abdomen and deviation of the bowel loops.

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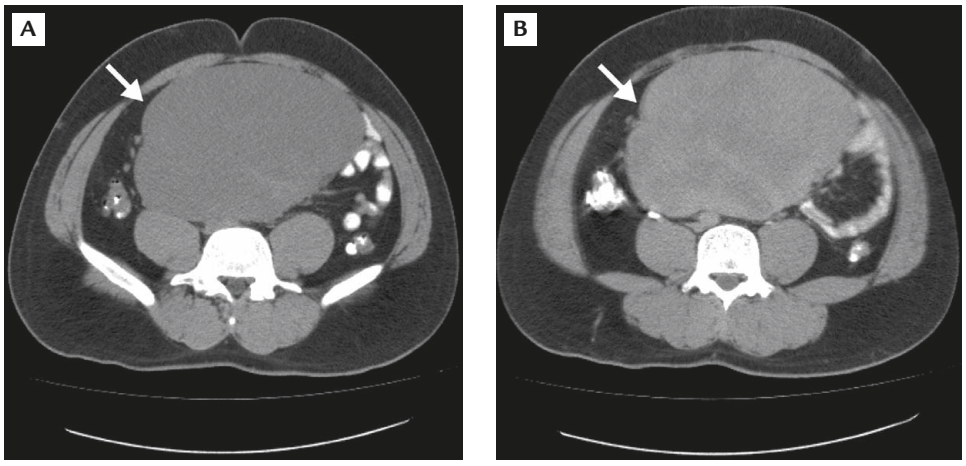

Figure 2. (A) Non-contrast abdominal computed tomography revealed a huge homogeneous mass lesion (arrow) with a well-defined margin in the midline abdomen and deviation of the bowel loops laterally. No inner calcification could be identified. (B) After contrast media administration, the mass (arrow) showed mild enhancement, mainly in the periphery. No vascular structure in the mass could be identified.

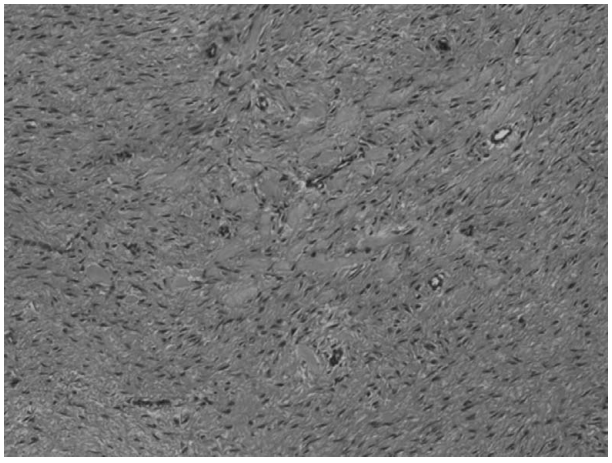

Figure 3. Microscopic pathology revealed nodular or plump aggregates of fibroblasts dispersed throughout dense collagen bundles, with extensive keloid-like collagen deposition.

lesion attached to the right mesocolon was found; right colectomy was performed. Pathology revealed desmoid tumor (Figure 3) and depicted infiltration to adjacent adipose tissue. Postoperatively, chemotherapy was administered and the patient showed good response over 2 years of follow-up.

Discussion

Desmoid tumors were first described in 1832 by MacFarlane. The tumor is rare, estimated to occur in about 2–4 per million population per year, and accounts for less than 3% of all soft-tissue tumors.[1] The tumor can arise from the abdomen, including the abdominal wall, mesentery, and the retroperitoneum. Most are sporadic, but they may occur as a consequence of abdominal surgery. Desmoid tumors may occur at any age, but are more common in people in their 30s and 40s. Abdominal desmoid tumors are rare in patients younger than 20 years. Females are more commonly affected than males in sporadic cases, with a ratio of about 2:1 to 5:1.[2] Multiple desmoid tumors are also more common in women.[3]

Intra-abdominal desmoid tumors are able to grow large before causing symptoms, such as obstruction of the bowel loops or urinary system. In a study of 25 patients, the abdominal desmoid tumors were up to 30 cm in diameter.[4] They also tend to recur after surgical resection or medical therapy.

The extremities, such as the shoulder, pelvic girdle, and chest wall or abdominal wall, are also common locations of these tumors. Abdominal wall desmoid tumors are reported to occur more often in younger women, especially during pregnancy, and recurrence is less frequent.[5] In contrast, desmoid tumors in the extremities are usually smaller, < 5 cm in diameter, and grow slowly.[6]

Desmoid tumors are known to be associated with familial adenomatous polyposis (FAP) in about 5% of cases.[3] In patients with FAP, the incidence of desmoid tumors is about 10–15%, which is 850 times higher than that in the general population. In these patients, the tumors usually arise in the abdominal wall or in the abdominal cavity, infiltrating mesentery, and may form a large and palpable mass.[7] The intra-abdominal desmoid tumors are an important and among the most common causes of death in patients with FAP.[8] A specific tumor-suppressing gene, APC, located on chromosome 5q22, is believed to be related to the development of desmoid tumor in patients with FAP.[7,8] In addition to the APC gene, the risk factors for FAP patients to

develop desmoid tumors include abdominal surgery, female sex, and family history of desmoid tumors. Desmoid tumor is also a component of Gardner syndrome. The incidence of abdominal desmoid tumors in patients with Gardner syndrome is about 4–29%.[4]

The principal aim of using imaging studies for desmoid tumors is for preoperative diagnosis and planning, to detect recurrence after surgical resection, and to follow tumor progression after medical treatment. Abdominal radiography may reveal a soft-tissue mass with mass effect and displacement of bowel loops. On US, abdominal desmoid tumors can present with variable echogenicity, with a smooth, well-defined margin. The imaging features of abdominal desmoid tumors on CT and magnetic resonance imaging are related to their histologic character and vascularity.[9] On CT, desmoid tumors with myxoid stroma will present with a hypodense mass, and those with collagenous stroma will present with a homogeneous, soft-tissue density mass. In this patient, the tumor was composed of both myxoid and collagenous stroma, and thus it presented as a hypodense lesion. The margin of a desmoid tumor can be well-defined, infiltrative, or with mixed pattern.[10] They usually show variable enhancement after contrast agent administration, from mild homogeneous to heterogeneous enhancement.

Magnetic resonance imaging is another imaging modality for evaluating tumor condition, because of its great soft-tissue contrast and high resolution compared with other modalities. Desmoid tumors have low signal intensity on T1-weighted imaging and variable signal intensity on T2-weighted imaging. The higher the signal intensity on T2-weighted imaging, the more rapidly the tumor is growing.[11]

It is important to differentiate abdominal desmoid tumor from other intra-abdominal tumors. Gastrointestinal stromal tumor of the small bowels usually shows areas of hemorrhage and central necrosis when they grow large, and calcification may sometimes be present. Sclerosing mesenteritis usually develops in elderly people around the age of 60 years, occurs more commonly in males, and presents with a well-defined or ill-defined abdominal mass lesion enveloping the mesenteric vessels with a spared fatty collar around the vessels, termed the "fat ring sign".[12] The imaging findings of inflammatory pseudotumor are nonspecific, with heterogeneous attenuation, well-defined margin, variable enhancement after contrast agent administration, and central necrosis in larger lesions. Prominent vascularity may be depicted with Doppler US in inflammatory pseudotumor.[13]

The treatment of desmoid tumors includes surgery, pharmacological therapies, and radiation therapy. However, the tumor tends to recur after surgical resection or medical therapy. Also, in patients with abdominal desmoid tumors, colonoscopy and upper gastrointestinal endoscopy are indicated to determine the presence of FAP.[1] The prognosis of desmoid tumors varies. About 10% resolve spontaneously, 30% progress and resolve intermittently, 50% remain stable, and 10% progress rapidly.[14]

In conclusion, we have presented a case of abdominal desmoid tumor and demonstrated the typical imaging findings, which are homogeneous and hypoechoic on US, and well-defined, hypodense to soft-tissue density, with mild enhancement after contrast media administration on CT. It is important to include desmoid tumor in the differential diagnosis when a huge homogeneous abdominal lesion occurs in a patient.

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