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Case Report

Pancreatic cystic desmoid tumor following metastatic colon cancer surgery: A case report☆,☆☆

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ABSTRACT

Desmoid tumors are rare, benign, and locally aggressive neoplasms that stem from connective tissue that have high rates of recurrence after surgery. Intra-abdominal desmoid-type fibromatosis can arise in 2 forms: sporadic or hereditary (associated with familial adenomatous polyposis and Gardner syndrome). The diagnosis of desmoid-type tumors is based on imaging modalities and histopathological examination. The primary treatment is resection surgery. We report a 64-year-old male with a distal pancreatic desmoid tumor. We focus on tumor management by the application of radiological modalities and pathological analysis.

Introduction

Desmoid tumors are noncancerous growths that arise from connective tissues. They can emerge almost anywhere in the body, such as the abdomen, arms, and legs. A retroperitoneal neoplasm such as a pancreatic desmoid tumor is more common in patients with familial polyposis coli and Gardner syndrome and can occur after abdominal surgery. It may present with vague abdominal pain, cramping, and nausea [1]. Such tumors are usually detected on an abdominal computed tomography [CT] scan or magnetic resonance imaging and confirmed with a pathology exam. Slow-growing tumors can be managed conservatively with routine follow-up but those that grow quickly into nearby structures and organs are treated with surgery, radiation therapy, and chemotherapy [2].

Case report

A 66-year-old male with a history of colon cancer metastatic to the caudate lobe with status postsigmoidectomy and hepatic wedge resection presented for a follow-up CT scan 3 years

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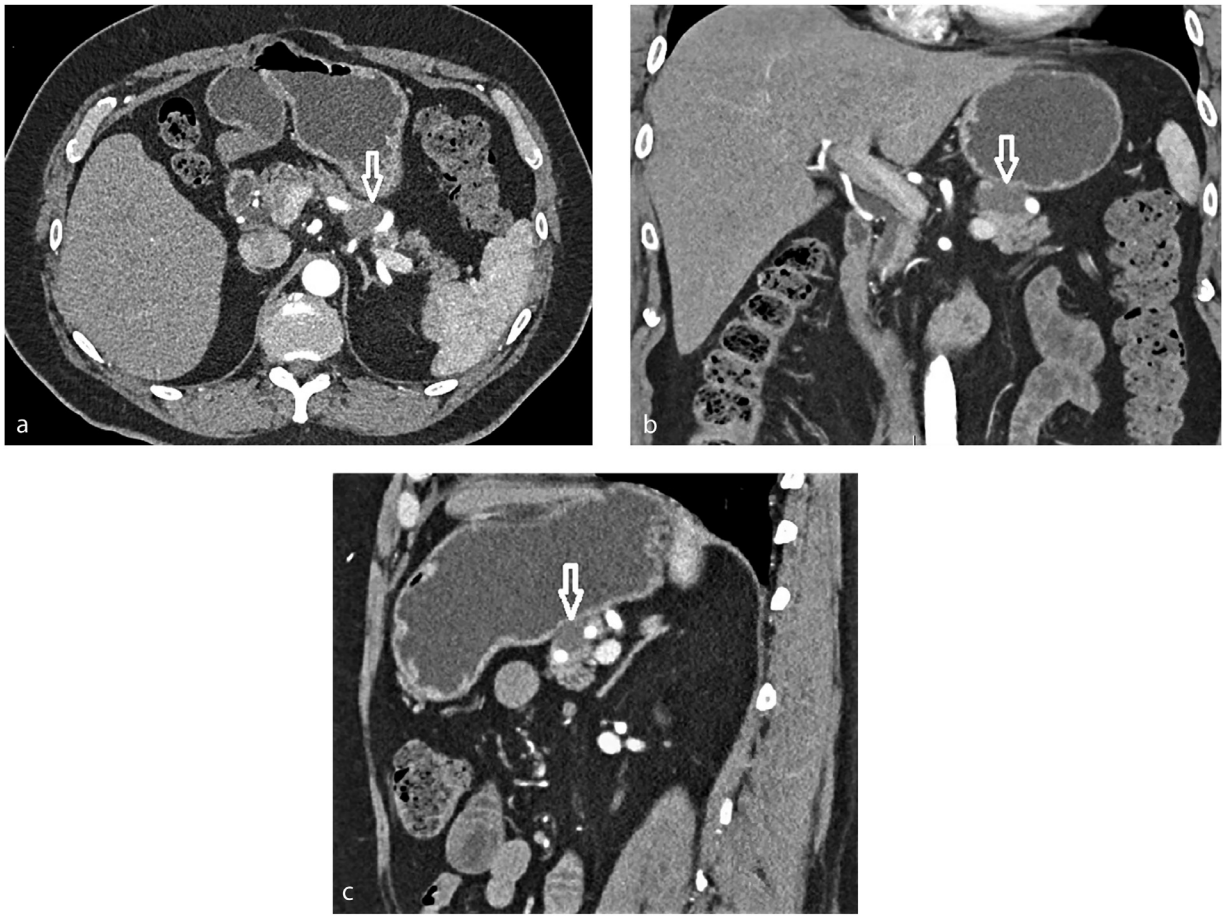

Fig. 1 – Contrast computed tomography scan (arterial phase) showing a hypodense mass (2.9 x 2.0 cm) located in the body of the pancreas abutting the splenic artery in (A) Axial (B) Coronal (C) Sagittal reformations.

after his surgery. The exam showed a new ill-defined hypodense lesion in the body of the pancreas measuring up to a maximum of 2.3 × 1.8 cm, with the abutment of the splenic artery and vein without upstream pancreatic atrophy, ductal dilatation, peripancreatic fat stranding, edema, or significant lymphadenopathy. A CT scan after 1 month showed a slightly larger lesion compared to the prior study, measuring up to 2.9 × 2.0 cm (Fig. 1)

Given the history of colon cancer and clinical suspicion for colon cancer metastases, endoscopic ultrasound-guided fine-needle aspiration was performed, with results revealing a spindle cell lesion without epithelial malignancy It was unlikely to represent a primary pancreatic malignancy, but it was uncertain if this represented a benign lesion or other neoplasms. Concerning the sudden appearance of the lesion and increased size in a repeat CT scan as well as colon cancer history, the patient underwent a distal pancreatectomy and splenectomy for definitive diagnosis. The surgical pathology showed a 3-cm intrapancreatic fibromatosis. The tumor extended to 0.1 cm from the edge of the pancreas. Twelve lymph nodes and associated fibroadipose tissue were negative for tumor cells (Fig. 2). The patient had no evidence of desmoid tumor recurrence on follow-up CT scans after 8 months.

Discussion

Desmoid tumors are rare, noncancerous neoplasms that cytologically result from abnormal proliferation of myofibroblasts throughout the body, and often appear inside the abdomen. These tumors represent approximately 0.03% of all tumors with the peak incidence in females who are in their third decade of life [3]. Firm, well-differentiated outgrowths, desmoid tumors are locally aggressive and found most often with FAP or Gardner syndrome. They are usually asymptomatic but can confer nonspecific symptoms related to the location and degree of invasion of adjacent structures [4]. In cases of pancreatic lesions, patients may present with epigastric pain with radiation to the back or weight loss. Isolated pancreatic desmoid tumors, as described in our patient, are extremely uncommon and mostly located in the body or tail of the pancreas [5]. Imaging modalities like CT scanning and magnetic resonance imaging are used for the diagnosis and follow-up of tumors and determining the extent of the lesion before surgical removal. Its imaging appearance can mimic hypodense solid neoplasms such as pancreatic ductal adenocarcinoma or cystic pancreatic neoplasms. A biopsy can confirm the diagnosis by showing the low proliferative activity of

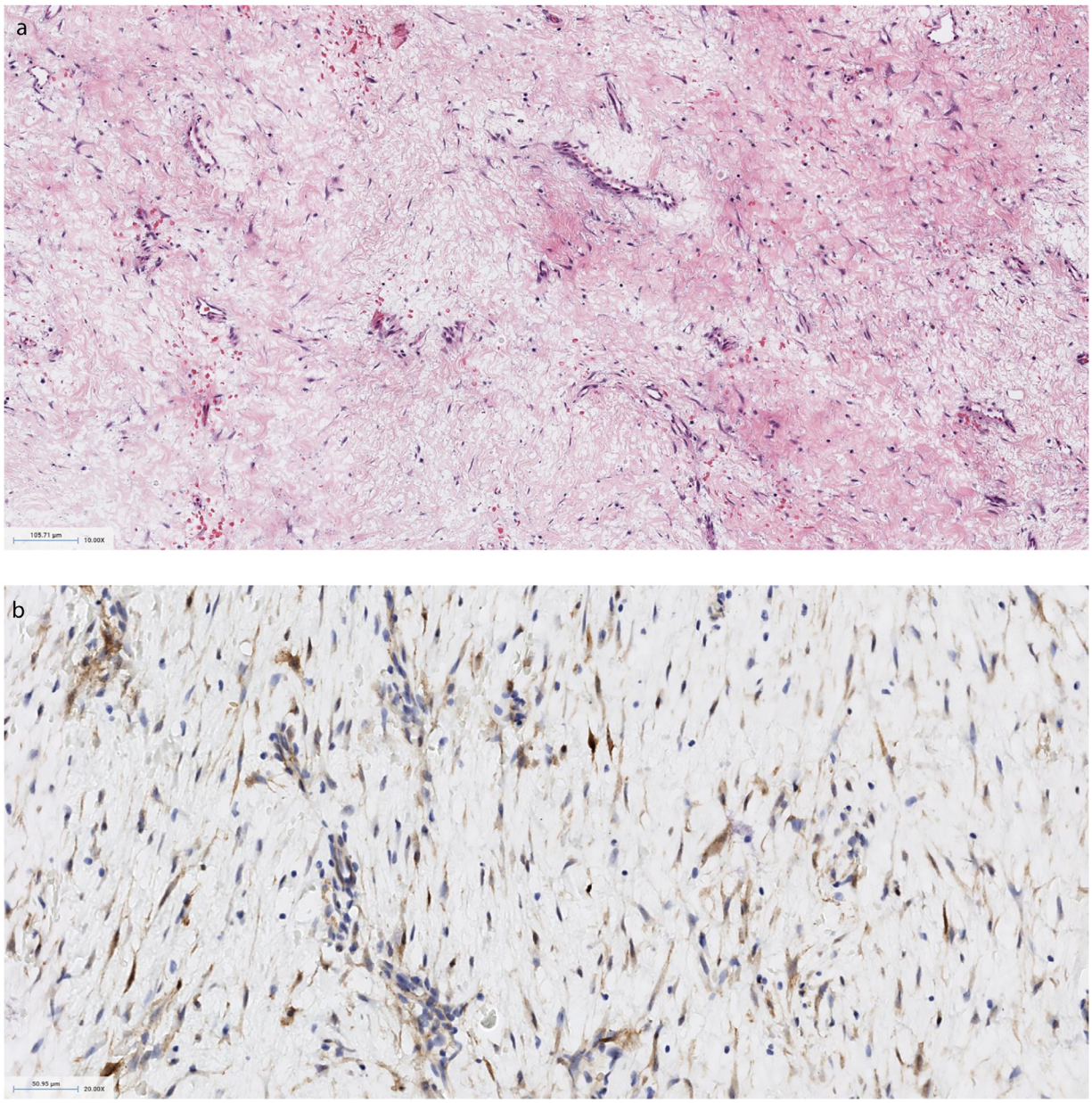

Fig. 2 – Histopathologic evaluation of distal pancreatectomy resection specimen. (A) Hematoxylin and eosin (H&E) stained section from the tumor showing elongated, thin spindled cells present in a collagenous stroma. Note the characteristic thin-walled vessels. (B) Immunohistochemistry for beta-catenin demonstrates abnormal nuclear labeling, confirming the diagnosis.

spindle cells with beta-catenin gene mutation and little cytologic atypia [6]. Surgery with negative surgical margins is the treatment of choice for desmoid tumors. Some studies have revealed that an extended course of nonsteroidal anti-inflammatory medication, hormonal therapy, and cytotoxic chemotherapy can be effective complementary treatments [7].

Polistina et al [8] reported one of the first cases of pancreatic desmoid tumors in 2010. The patient was an asymptomatic 68-year-old man whose incidental desmoid tumor in the tail of the pancreas was found on an abdominal CT scan. The diagnosis was confirmed by pathology and the patient underwent surgery without further complications. Słowik-Moczydłowska et al [1] described a 13-year-old Caucasian boy with recurrent pain for 2 months in the left hypochondrium of his abdomen. The cystic lesion in the hilum of the spleen and tail of the pancreas was discovered in ultrasonography and an abdominal-enhanced CT scan. After confirmation by histological analysis, distal splenopancreatectomy was undertaken, which showed desmoid-type fibromatosis in the postsurgery pathology report. Recently, Torres et al [2] published a report of a 15-year-old boy admitted to the hospital complaining of mild generalized abdominal pain for 1 week, nausea, vomiting, and a low-grade fever. A large cystic lesion in the body of the pancreas was found in an enhanced CT scan of his abdomen. In the pathology report after resection, the diagnosis of desmoid-type fibromatosis of the pancreas was confirmed.

To the best of our knowledge, there have been 27 cases described in the literature of pancreatic desmoid tumors in the last 3 decades, of which only 6 were cystic masses [3]. Compared to the available case reports in the literature, our patient was unique in terms of incidental discovery of the cystic lesion on an abdominal CT scan following metastatic colon cancer surgery, which can mimic the appearance of metastatic disease. It was diagnosed as a desmoid tumor by showing spindle cells and positive beta-catenin immunohistochemical staining. Therefore, a combination of radiologic images and pathology evaluation played an essential role in identifying the tumor [9]. Generally, the best treatment is decided based on location, the expansion of the tumor to neighboring organs, and risk to surrounding structures. In the case presented here, surgical intervention with wide, free margins for the prevention of tumor recurrence was performed. Follow-up was also necessary, despite the low rate of recurrence. The follow-up CT scan revealed no sign of recurrence. Although a pancreatic cystic desmoid tumor is extremely rare, it should be considered in the differential diagnosis of pancreatic lesions.

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