American Journal of Case Reports

ISSN 1941-5923
© Am J Case Rep, 2014; 15: 374-377
DOI: 10.12659/AJCR.891044

Received: 2014.05.16
Accepted: 2014.05.28
Published: 2014.09.02

# A Giant Mesenteric Desmoid Tumor Revealed by Acute Pulmonary Embolism due to Compression of the Inferior Vena Cava

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

BCDE Elisa Palladino
BCDF Joseph Nsenda
ABCD Renaud Siboni
CFG Christian Lechner

Department of General and Digestive Surgery, Chalons en Champagne Hospital, Chalons en Champagne, France

**Corresponding Author:** Elisa Palladino, e-mail: palladinoelisa@hotmail.com
**Conflict of interest:** None declared

**Patient:** Male, 69
**Final Diagnosis:** Mesenteric desmoid tumor
**Symptoms:** —
**Medication:** —
**Clinical Procedure:** —
**Specialty:** Surgery

**Objective:** Rare disease

**Background:** Intra-abdominal fibromatosis is a benign rare tumor of fibrous origin with a significant potential for local invasion and no ability to metastasize, but it can recur. The etiology of desmoid tumors is unknown. It is often associated with conditions such as familial adenomatous polyposis and Gardner syndrome.

**Case Report:** We report the case of a 69-year-old man who presented to our hospital with an acute pulmonary embolism. The patient had a past history of colic surgery for a polyp with a high-grade dysplasia. Pulmonary angiography showed partial occlusion of the right superior lobe artery and partial occlusion of the middle lobe artery. The patient was given thrombolytic therapy. Abdominal computerized tomography revealed a mesenterial giant mass with compression of the inferior vena cava (IVC). A biopsy of the mass, confirming aggressive fibromatosis. A laparotomy was performed, which revealed a massive growth occupying the abdomen and attached to the previous ileocolic anastomosis. One day after surgery, his condition deteriorated.

**Conclusions:** This report underlines the potential of imaging investigations of abdomen and vena cava if pulmonary embolism is suspected, especially when there is no evidence of peripheral venous thrombosis or other predisposing factors. Unfortunately, data on the surgical management of desmoid tumor is scarce. Therefore, the standard of treatment is a surgical resection for resectable tumors.

**MeSH Keywords:** Fibromatosis, Abdominal • Fibromatosis, Aggressive • Pulmonary Embolism

**Full-text PDF:** http://www.amjcaserep.com/abstract/index/idArt/891044

750 — 4 20

## Background

A desmoid tumor, also referred to as aggressive fibromatosis, is a rare benign tumor but has highly locoregional aggressive potential arising from a proliferation of fibrous tissue, with an incidence of 2–4 per million per year and a slight female preponderance. It has no known etiologic factor; however, it is associated with certain familiar syndromes such as familial adenomatous polyposis (FAP) [1]. Other risk factor include: previous trauma, prolonged estrogen intake, and previous surgery. Desmoid tumors can localized in a variety of anatomic sites: abdominal wall, retroperitoneum, and extremities with a high aggressive potential of adjacent structures.

## Case Report

A 69-year-old-man was admitted into our unit through the emergency room for a suspected acute pulmonary embolism. The diagnosis was confirmed by pulmonary angiography, which showed partial occlusion of the right superior lobe artery and partial occlusion of the middle lobe artery. The patient had a medical history of tuberous sclerosis, a right colectomy for a polyp with a high-grade dysplasia, prostate cancer, and chronic constipation. He reported no history of abdominal trauma or prior venous thromboembolic event.

On admission the patient was stable and abdominal examination revealed a mobile abdominal mass involving the entire right abdomen. An abdominal computed tomography (CT) scan showed an intraperitoneal heterogeneous, tissular mass (20×11×16), arising from the mesentery, involving the duodenojejunal angle, and the inferior vena cava was severely compressed (Figures 1 and 2).

The inferior vena cava was dilated and showed no laminar thrombi distal to the obstruction. The patient was given thrombolytic therapy followed by heparin administration for 3 weeks. A biopsy of the mass confirmed the aggressive fibromatosis. The patient was prepared for surgery. A laparotomy was performed and confirmed the large mesenteric mass involving the proximal jejunal region and the ileocolic anastomosis

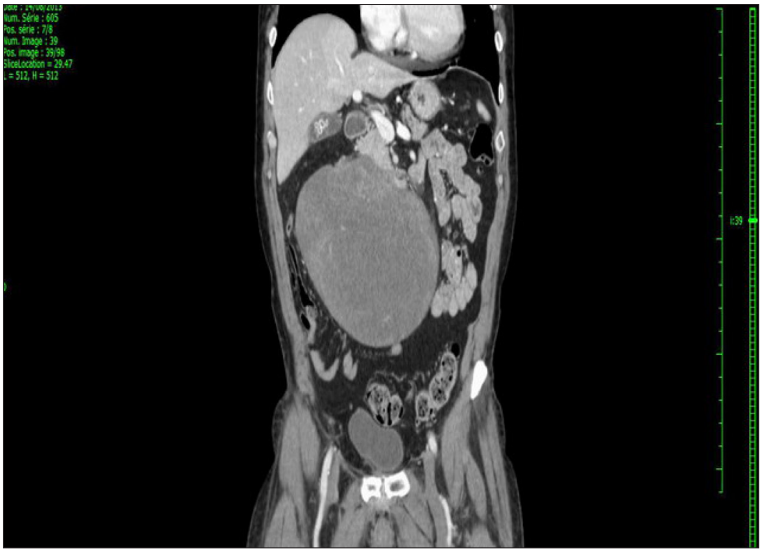

**Figure 1.** Contrast-enhanced computed tomography abdominal scan (sagittal section) showed a mesenterial mass occupying almost the entire right abdomen, with compression of inferior vena cava.

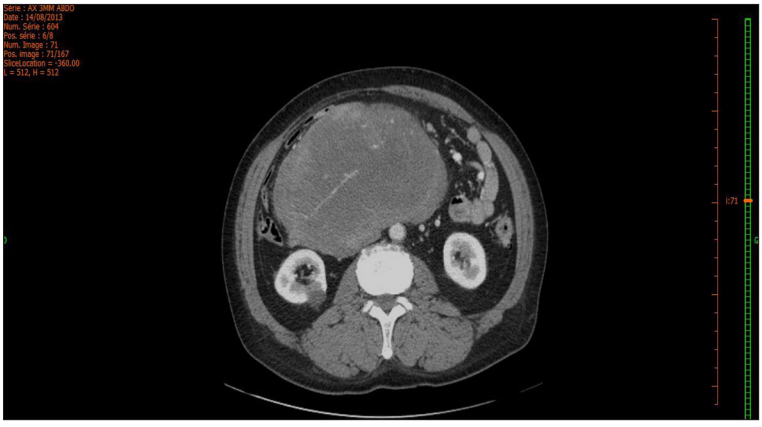

**Figure 2.** Contrast-enhanced computed tomography abdominal scan showing a heterogenous mesenterial mass (axial section).

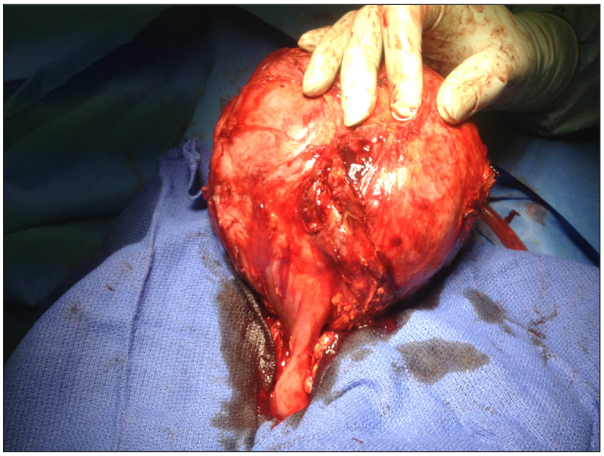

**Figure 3.** Intraoperative image showing attached jejunal segment.

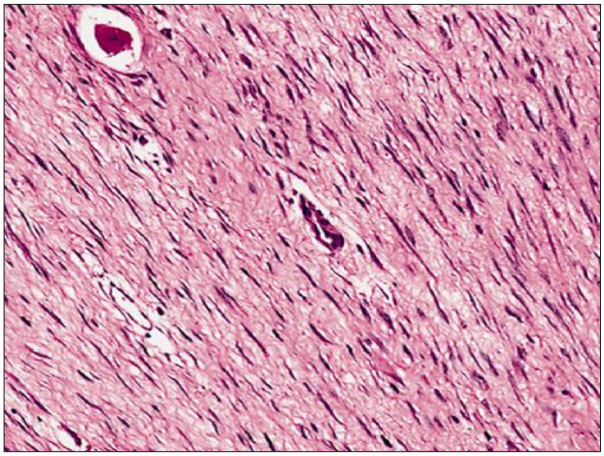

**Figure 4.** Histopathological picture of mesenteric desmoid tumor showing a tumor composed of stellate and spindle cells.

(Figure 3). No lymph nodes or ascites were seen. No focal lesion was noted in the liver. The mass was resected with a part of the adherent jejunal and cholic segment, using linear GIA stapler anastomosis. The postoperative course was complicated by a fatal acute myocardial infarction and the patient died on post-operative day 1. The microscopic and immunohistochemical findings were compatible with the diagnosis of an aggressive mesenteric fibromatosis (Figures 4) (the tumor was positive for vimentin and to beta-catenin).

## Discussion

Fibromatosis (or desmoid tumor) is an uncommon but highly aggressive group of benign tumors arising from fascial or musculoaponeurotic tissues, including mesenteric fibromatosis [2,3]. Fibromatosis comprises 0.03% of all tumors, with a slight young adult preponderance and no identifiable etiologic factor. It is, however, associated with familiar syndromes such as familial adenomatous polyposis (FAP) [1].

A possible genetic predisposition may be implicated, especially in patients with Gardner's syndrome having a high potential for developing mesenteric fibromatosis compared with the normal population. Most of the patients are clinically asymptomatic and the symptoms are associated with progressive invasion of contiguous structures and organ compression [4,5].

In 2007 Koh et al. published their experience and reported 5 mesenteric fibromatosis patterns: 1) spontaneous regression pattern, 2) stable pattern, 3) variable growth pattern, 4) progressive growth pattern, and 5) aggressive growth pattern [1].

They found that 75% of all patients had a progressive growth pattern necessitating early surgical treatment [6–13]. Because of the rarity of desmoid tumors and the unavailability of data on their surgical management, there are neither specific guidelines nor a standardized therapeutic approach to its management.

Therefore, curative surgical resection remains the standard of treatment for resectable tumors [14–18]. Preoperative biopsy is recommended for differential diagnosis with intestinal carcinoma, carcinoid tumor, mesenteric fibromatosis, lymphoma, and retroperitoneal fibrosis [17].

Radiotherapy is indicated for unresectable or recurrent tumors [8]. Acute pulmonary embolism due to compression of the inferior vena cava related to mesenteric fibromatosis is very rare. Few cases of pulmonary embolism caused by gastrointestinal disease appear in the literature [19].

In this report the patient died and the origin of the thrombosis was the tumor itself. This report underlines the potential interest of imaging investigations of the abdomen and vena cava if pulmonary embolism is suspected, especially when there is no evidence of peripheral venous thrombosis or other predisposing factors [7,8].

## Conclusions

Mesenteric fibromatosis is usually asymptomatic and presents with a variety of clinical features [20]. The physicians should be suspicious when a pulmonary embolism is present without evidence of deep vein thrombosis or other predisposing factors. Consequently, therapeutic strategies should be specifically tailored for each patient and their unique presentation according to anatomic location [14,15].

### Conflicts of interest

The authors declare no conflicts of interest.

## References:

1. Koh PK, Loi C, Cao X et al: Mesenteric desmoid tumours in Singapore familial adenomatous polyposis patients: clinical course and genetic profile in a predominantly Chinese population. Dis Colon Rectum, 2007; 50: 75–82
2. Shields CJ, Winter DC, Kirwan WO, Redmond HP: Desmoid tumours. Eur J Surg Oncol, 2001; 27: 701–6
3. Faria SC, Iyer RB, Rashid A et al: Desmoid tumor of the small bowel and the mesentery. Am J Roentgenol, 2004; 183(1): 118
4. Lath C, Khanna PC, Gadewar SB, Agrawal D: Inoperable aggressive mesenteric fibromatosis with ureteric fistula.Case report and literature review. Eur J Radiol, 2006; 59: 117–21
5. Rakha EA, Kandil MA, El-Santawe MG: Gigantic recurrent abdominal desmoid tumor: a case report. Hernia, 2007; 11: 193–97
6. Smith AJ, Lewis JJ, Merchant NB et al: Surgical management of intra-abdominal desmoid tumor. Br J Surg, 2000; 87: 608–13
7. Yang CH, Sheen-Hen SM, Lu CC et al: Computed tomographic presentation of mesenteric fibromatosis. Dig Dis Sci, 2005; 50: 348–50
8. Teo HE, Peh WC, Shek TW: Case 84: desmoid tumor of the abdominal wall. Radiology, 2005; 236: 81–84
9. Middleton SB, Phillips RK: Surgery for large intra-abdominal desmoids tumors: report of four cases. Dis Colon Rectum, 2000; 43: 1759–62; discussion 1759–62
10. Kabra V, Chaturvedi P, Pathak KA, deSouza LJ: Mesenteric fibromatosis: a report of three cases and literature review. Indian J Cancer, 2001; 38: 133–36
11. Lev D, Kotilingam D, Wei C et al: Optimizing treatment of desmoids tumors. J Clin Oncol, 2007; 25: 1785–91
12. Spiridakis K, Panagiotakis G, Grigoraki M et al: Isolated giant mesenteric fibromatosis (intra-abdominal desmoids tumors). Case Report. G Chir, 2008; 29(10): 413–16
13. Gari MK, Guraya SY, Hussein AM, Hego MM: Giant mesenteric fibromatosis: report of a case and review of the literature. World J Gastrointest Surg, 2012; 4(3): 79–82
14. De BreeE, Keus R, Melissas J et al: Desmoid tumors: need for an individualized approach. Expert Rev Anticancer Ther, 2009; 9: 525–35
15. Fiore M, Rimareix F, Mariani L et al: Desmoid-type fibromatosis: a front-line conservative approach to select patients for surgical treatment. Ann Surg Oncol, 2009; 16: 2587–93
16. Stockle E, Coindre JM, Longy M et al: A critical analysis of treatment strategies in desmoids tumours: a review of a seies of 106 cases. Eur J Surg Oncol, 2009; 35: 129–34
17. Stoidis CN, Spyropoulos BG, Misiakos EP et al: Surgical treatment of giant mesenteric fibromatosis presenting as a gastrointestinal stromal tumor: a case report. J Med Case Report, 2010; 4: 314
18. Peng PD, Hyder O, Mavros MN et al: Management and recurrence patterns od desmoids tumors: a multi-institutional analysis of 211 patients. Ann Surg Oncol, 2012; 19(13): 4036–42
19. Paolillo V, Sicuro M, Nejrotti A et al: Pulmonary Embolism due to compression of the Inferior vena cava by a hepatic emangioma. Tex Heart Inst J, 1993; 20: 66–68
20. Disher AC, Biswas M, Miller TQ, Kuvhenguhwa A: Atypical desmoids tumor of the abdomen: a case report. J Natl Med Assoc, 1993; 85: 309–11