AMERICAN JOURNAL OF DIAGNOSTIC IMAGING, 2018
VOL 4, NO. 1, PAGE 1–4
10.5455/ajdi.20180219040806

eJM eJManager

CASE REPORT

Open Access

Pelvic fibromatosis reaching cavo-atrial junction enroute inferior vena cava—a rare presentation

Nilu Malpani Dhoot[1], Arpita Bhaumik[1], Anasua Chattopadhyay[1], Usha Goenka[1], Sanjib Majumdar[1], Surabhi Goenka[1], Enam Murshed Khan[2], Rashmi Chand[1]

[1]Imaging and Intervention, Department of Radiology, Apollo Gleneagles Hospital Kolkata, West Bengal, India

[2]Histopathology Division, Department of Pathology, Apollo Gleneagles Hospital Kolkata, West Bengal, India

ABSTRACT

Fibromatosis is a group of soft tissue tumors with unpredictable clinical presentation distinguished as locally belligerent and non metastasizing. Deep fibromatosis is rarer than superficial fibromatosis. We report an unique case of intravascular extension of fibromatosis where the tumor epicenter in the pelvis reached the cavo-atrial junction enroute Inferior vena cava.

ARTICLE HISTORY

Received February 19, 2018
Accepted March 04, 2018
Published July 16, 2018

KEYWORDS

Pelvic fibromatosis
intravascular invasion

Introduction

Deep fibromatosis is characterized by a variable and unpredictable clinical presentation. They are a rare fibroblastic cell proliferation distinguished as locally belligerent and non-metastasizing [1]. Pelvic fibromatosis is rare, with only 31 cases having been described previously [2]. We recount an unique case where fibromatosis epicenter in pelvis reached the cavo-atrial junction travelling via Inferior vena cava (IVC).

Case Report

A 50-year female presented to the gynecology department with complaints of pain in lower abdomen and difficulty in micturition since last 7 months. She had undergone trans-abdominal hysterectomy and bilateral salpingo-oophorectomy for abnormal uterine bleeding. She had two prior caesarian sections. No other co-morbidities were known at the time of presentation. She was not on any regular medications.

On examination, a lobulated lump was felt in the pelvis. On per vaginal examination, a mass could be felt in the right fornix. An ultrasound of the whole abdomen showed multiple heterogeneous masses extending throughout abdomen and pelvis. No definite organ of origin could be identified. Hence, a contrast enhanced computed tomography (CECT) was done.

The CECT revealed, large mass in the pelvis measuring 10.2 × 9.6 × 8.3, filling the pelvis more on the right side, displacing the urinary bladder and bowel loops anteriorly and towards the left with maintained fat planes (Fig. 1). The mass infiltrated the internal and common iliac veins on the right with tumor thrombus extending to IVC and reaching the cavo-atrial junction mildly projecting into the right atrium (Figs. 2–4). Numerous nodular masses were also seen in the mesentry, contiguous with each other and the IVC tumor (Fig. 5). All of these solid masses showed similar morphological and enhancement characteristics having moderate and fairly homogenous enhancement with very few necrotic areas. The right ureter was compressed with proximal hydroureteronephrosis and delayed contrast excretion. Mild ascites was noted.

Based on the above CT findings a diagnosis of leiomyoma or leiomyosarcoma was suggested. Ultrasonography (USG) guided core biopsy was done from one of the mesenteric nodules which suggested benign spindle cell neoplasm (Fig. 6).

Contact Nilu Malpani Dhoot ✉ drniludhoot@gmail.com Department of Radiodiagnosis and Intervention, Apollo Gleneagles Hospital, Kolkata, India.

© EJManager. This is an open access article licensed under the terms of the Creative Commons Attribution Non-Commercial License (http://creativecommons.org/licenses/by-nc/3.0/) which permits unrestricted, noncommercial use, distribution and reproduction in any medium, provided the work is properly cited.



Figure 1. Contrast enhanced axial section of the lower abdomen shows nodular masses in the pelvis and mesentery (solid arrow). The masses invades the right iliac veins (*). The right iliac artery is pushed anteriorly (arrow). The mass is more on the right side. The bowel loops are pushed anteriorly and to the left.



Figure 2. Coronal CT reconstruction shows large pelvic mass travelling along the iliac vein to the IVC (solid arrow). There is right hydronephrosis and mild ascites.

Beta catenin was positive on immunohistochemistry (Fig. 7) and negative for CD34, CD45, CD117, desmin, DOG1, Ki67, S100, and smooth muscle actin. The pathology findings favored fibromatosis desmoid type. Extensive local spread of the tumor rendered it inoperable. The patient was put on tamoxifen and is under regular follow up. At 3 months follow up the disease showed no interval change.

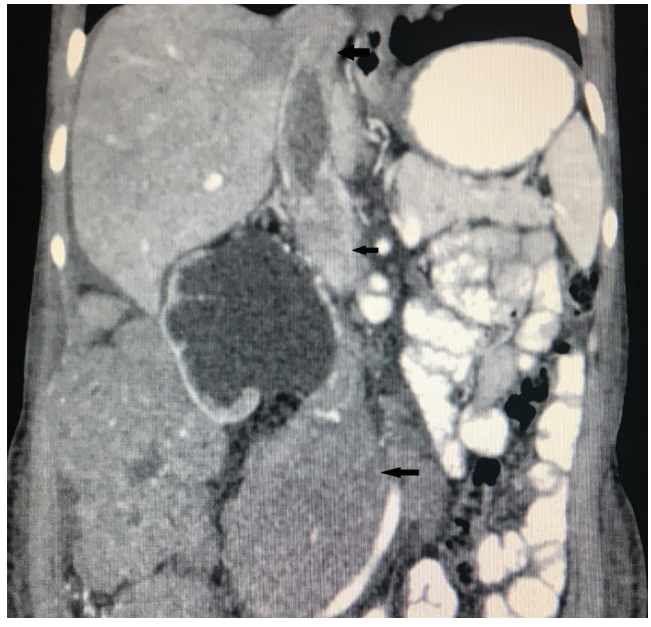


Figure 3. Coronal CT reconstruction shows large pelvic mass travelling along the iliac vein to the IVC (solid arrow). There is right hydronephrosis and mild ascites.

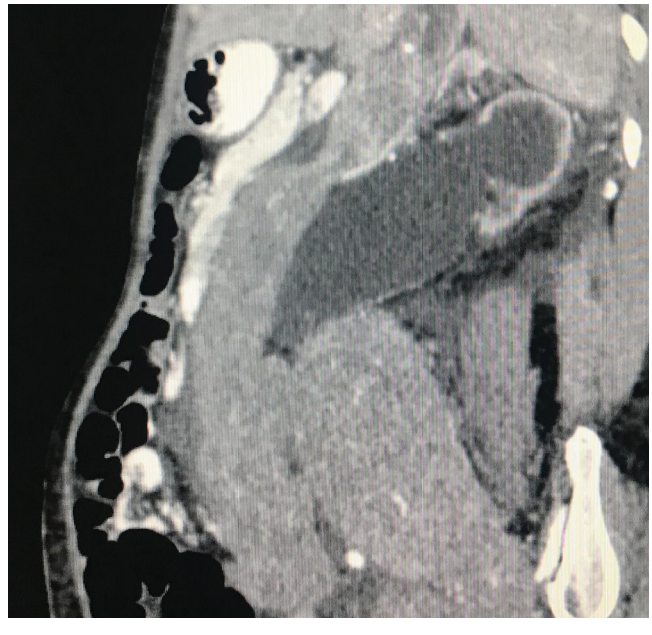


Figure 4. Oblique sagittal reconstruction image shows mass reaching the cavo-atrial junction (solid arrow). There is a mesenteric mass and the pelvic mass appears contiguous with each other and the IVC tumor thrombus.

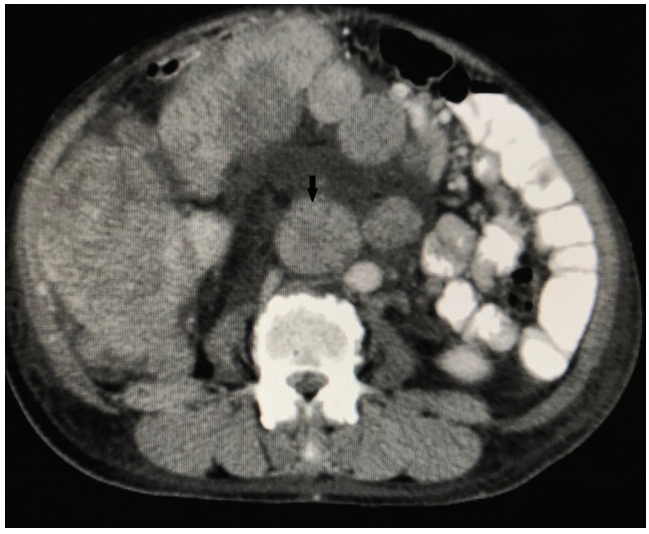


Figure 5. Axial CT shows tumor thrombus in the IVC (solid arrow), nodular mesenteric masses and ascites.

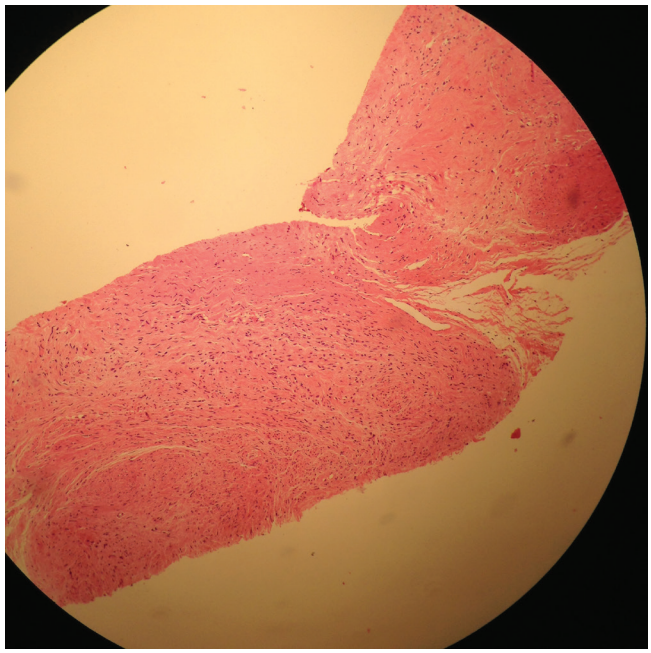


Figure 6. Histopathology shows interlacing fascicles of benign spindle shaped cells with spindly nuclei, inconspicuous nucleoli and moderate cytoplasm favoring spindle cell tumor.

Discussion

Fibromatosis or desmoid tumor defines a fibromatous proliferative tumor that is classified between benign fibrous tissue proliferation and fibrosarcoma in its biological behavior [3]. It is classed by the World Health Organization as an intermediate soft tissue tumor related to sarcoma family having locally aggressive nature. They are predominantly idiopathic in origin and can also be caused due to traumatic, hormonal and genetic factors [4].

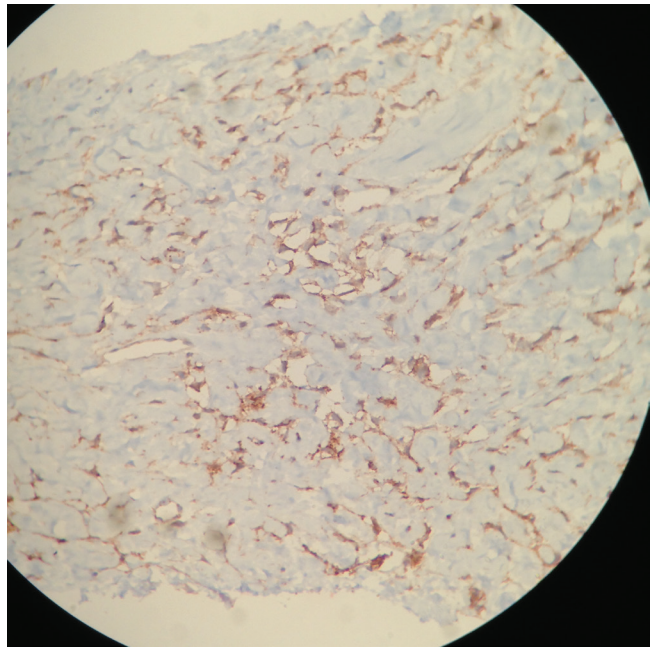


Figure 7. Immunohistochemistry shows beta catenin positive.

Based on location fibromatosis may be classified as superficial or deep. Although superficial and deep fibromatosis possess identical histologic morphology, superficial-type fibromatosis are beta catenin negative, more frequent and are seen at plantar, palmar, or penile locations as compared to deep, visceral and extra-abdominal locations (i.e., abdominal or chest wall) of the deep desmoid-type which are rare and beta catenin positive [5].

Clinical presentation is variable depending on the local extension of the tumor. Our case was a deep fibromatosis with intravascular extension growing along the IVC to the right atrium. There was extension to the mesentery as well.

A hallmark of aggressive fibromatosis is its unpredictable clinical behavior. Although some tumors have an aggressive course with a high tendency for local recurrence even after apparently adequate resection, other cases present with an initial growth phase followed by disease stabilization, in the absence of treatment [6]. Significant proportion of desmoid tumors remain stable or regress while others exhibit progression. Many patients with desmoids benefit from a primary nonaggressive treatment, because growth arrest is a common feature of this disease [6]. Conversely, an aggressive strategy (surgery and/or radiotherapy) as a primary policy for all patients might over treat large amount of them and may be associated with significant sequelae. Surgical intervention can be disfiguring and often requires reconstruction

and associated with post operative complications. Undoubtedly, the majority of patients exhibiting indolent disease will respond well to aggressive treatments; however, it is also likely that to have had an equally good outcome without any intervention.2In our institute, treatment algorithm commences with more conservative approaches. In our case due to extensive invasion, high dose Tamoxifen (120 mg/day) was advised along with anti inflammatory drugs. On 3 month follow up there was no significant change in the tumor volume. The tamoxifen treatment provides a receptor-mediated mechanism for the action of antiestrogenic compounds in the treatment of fibromatosis [7].

The cause of fibromatosis remains unclear. It is thought that the condition may be related to trauma, surgery, hormonal factors, or have a genetic association. Desmoid tumors may occurs sporadically having somatic beta catenin-activating mutations and within the context of familial adenomatous polyposis exhibiting germline inactivating mutations of Adenomatous polyposis coli (APC) gene [6]. Our patient had history of previous hysterectomy and positive beta catenin.

There are few case reports where pelvic fibromatosis showing local aggressive behavior and have been successfully treated [8,9]. There are multiple cases of deep fibromatosis in the mediastinum and head and neck [10–12], causing superior vena cava syndrome [11] by venous compression.

However, intravascular invasion and continuation of fibromatosis has not been reported till date. To our knowledge, this is the first reported case of pelvic fibromatosis travelling intravascularly via IVC upto the right atrium.

References

[1] Mariani A, Nascimento AG, Webb MJ, Sim FH, Podratz KC. Surgical management of desmoid tumors of the female pelvis. J Am Coll Surg 2000; 191:175–83.

[2] Huang GS, Lee HS, Lee CH, Chen CY. Pelvic fibromatosis with massive ossification. Am J Roentgenol 2005; 184:1029–30.

[3] Kasper B, Ströbel P, Hohenbergera P. Desmoid tumors: clinical features and treatment options for advanced disease. Oncologist 2011; 16(5):682–93.

[4] Weiss SW, Goldblum JR. Soft tissue tumours: fibromatoses. 5th edition. Mosby Elsevier, Philadelphia, PA, 2008.

[5] Ross JA, Zhang X. Desmoid-type fibromatosis. Atlas Genet Cytogenet Oncol Haematol 2013; 17(8):571–8.

[6] Bonvalot S, Desai A, Coppola S, Le Péchoux C, Terrier P, Dômont J, et al. The treatment of desmoid tumors: a stepwise clinical approach. Ann Oncol 2012; 23:158–66.

[7] Lev D, Kotilingam D, Wei C, Ballo MT, Zagars GK, Pisters PW, et al. Optimizing treatment of desmoid tumors. J Clin Oncol 2007; 25:1785–91.

[8] Ariën F1, Aleman JM, Op de Beeck B, Tjalma WA. Treatment of aggressive pelvic fibromatosis with interferon. Obstet Gynecol 2015; 126(6):1219–21.

[9] Fishman A, Girtanner RE, Kaplan AL. Aggressive fibromatosis of the female pelvis. A case report and review of the literature. Eur J Gynaecol Oncol 1996; 17(3):208–11.

[10] Xie Y, Xie K, Gou Q, He J, Zhong L, Wang Y. Recurrent desmoid tumor of the mediastinum: a case report. Oncol Lett 2014; 8(5):2276–8.

[11] Inase N, Ichioka M, Akamatsu H, Usui Y, Miyake S, Yoshizawa Y. Mediastinal fibromatosis presenting with superior vena cava syndrome. Respiration 1999; 66(5):464–6.

[12] Abdelkader M, Riad M, Williams A. Aggressive fibromatosis of the head and neck (desmoid tumours). J Laryngol Otol 2001; 115(10):772–6.