CASE REPORT

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Imaging findings of familial adenomatous polyposis-associated aggressive mesenteric fibromatosis: A case report

Agresivna mezenterijalna fibromatoza udružena sa familijarnom adenomatoznom polipozom – karakteristike dobijene primenom *imaging* tehnika snimanja

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Abstract

Introduction. Aggressive fibromatosis, also known as desmoid type fibromatosis (DF) is a locally aggressive fibroblastic neoplasm that can arise anywhere in the body with no potential for metastasis and a high recurrence rate after surgical resection. Mesenteric fibromatosis are locally aggressive DF of the mesentery with a high propensity for bowel involvement. The real etiology of these tumors remains unknown, occurring sporadically or in association with familial adenomatous polyposis (FAP), as Gardner's syndrome. **Case report.** A 34-year-old female patient presented with a palpable solid tumefactive mass in the left hemiabdomen. Contrast enhanced computed tomography (CT) and magnetic resonance imaging (MRI) revealed multiple massive solid tumefactions in the mesentery and in between the small bowel loops. Colonoscopy confirmed the presence of multiple sessile polyps characteristic of FAP. Tissue samples of the mesenteric mass were acquired via ultrasound guided biopsy with histopathologic confirmation of desmoid fibromatosis with imunohistochemical analysis. The risk of surgery was deemed too high at the time due to the size of the mass and proximity to mesenteric vascular structures, therefore the patient was planned for chemotherapy with a potential for further surgical reevaluation. **Conclusion.** Mesenteric fibromatosis is a rare neoplasm that presents with a wide range of histologic and imaging features. CT and MRI play a crucial role in evaluation and planning an optimal treatment model for patients with mesenteric fibromatosis.

Key words:
biopsy, needle; colonoscopy; diagnosis; fibromatosis, abdominal; fibromatosis, aggressive; immunohistochemistry; magnetic resonance imaging; mesentery; tomography, x-ray computed.

Apstrakt

Uvod. Agresivna fibromatoza, takođe poznata kao dezmoidna fibromatoza (DF), je lokalno agresivna fibroblastična neoplazma koja se može javiti bilo gde u ljudskom telu, bez potencijala metastaziranja, sa visokom stopom recidiviranja nakon hirurškog uklanjanja. Mezenterijalne fibromatoze su lokalno invazivne fibromatoze mezenterijuma sa čestim zahvatanjem crevnih vijuga. Etiologija ovih tumora je nepoznata, a javljaju se sporadično ili udruženi sa familijarnom adenomatoznom polipozom (FAP) kao Gardnerov sindrom. **Prikaz bolesnika.** Bolesnica, stara 34 godine, javila se na pregled sa palpabilnom solidnom tumorskom masom u levom hemiabdomenu. Kompjuterizivanom tomografijom (KT) sa aplikacijom kontrastnog sredstva i magnetno-rezonantnim (MR) snimanjem otkrivene su višestruke solidne tumefakcije u mezenterijumu, kao i između crevnih vijuga tankog creva. Kolonoskopijom su viđeni multipli sesilni polipi karakteristični za FAP. Ultrazvučno navođenom biopsijom dobijeni su uzorci tkiva mezenterične mase sa patohistološkom verifikacijom DF, uz primenu imunohistohemijske analize. Procenjeno je da je rizik od hirurške intervencije prevelik s obzirom na veličinu promene i njen odnos prema mezenterijalnim vaskularnim strukturama, zbog čega je planirana hemioterapija, uz potencijalnu naknadnu hiruršku reevaluaciju. **Zaključak.** Mezenterijalna fibromatoza je retka neoplazma sa širokim spektrom histoloških i slikovnih karakteristika. KT i MR snimanje igraju važnu ulogu u proceni i planiranju optimalnog terapijskog modela kod bolesnika sa mezenterijalnom fibromatozom.

Ključne reči:
biopsija iglom; kolonoskopija; dijagnoza; fibromatoza, abdominalna; fibromatoza, agresivna; imunohistohemija; magnetna rezonanca, snimanje; mezenterijum; tomografija, kompjuterizovana, rendgenska.

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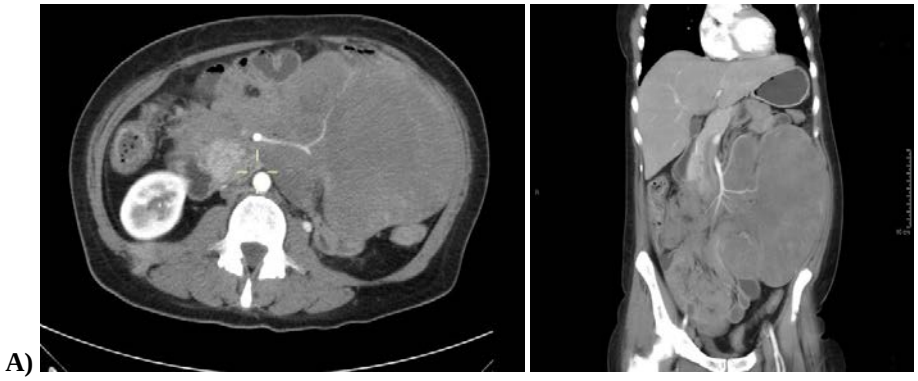
Introduction

Aggressive fibromatosis, also known as desmoid type fibromatosis (DF) is a locally aggressive fibroblastic neoplasm that can arise anywhere in the body with no potential for metastasis and a high recurrence rate after surgical resection. The term desmoid was first used by Muller in 1838 and is derived from the Greek word *desmos* meaning band or tendon [1]. DF is a rare neoplasm with an estimated annual incidence of 2–4 cases per million people, accounting for approximately 0.03% of all neoplasms and less than 3% of all soft tissue tumors [2]. It is commonly seen in the reproductive years of women, often during and after pregnancy [1]. By their location, these tumors can be classified as intraabdominal, extraabdominal, or abdominal wall. Mesenteric fibromatosis is locally aggressive DF of the mesentery with a high propensity for bowel involvement [3]. The real etiology of these tumors remains unknown, occurring sporadically or in association with familial adenomatous polyposis (FAP) as Gardners's syndrome [2, 3]. While they are non-metastasising, they can be locally aggressive, causing symptoms due to pressure effect, which can lead to complications such as small bowel obstruction, ischemia and perforation [4].

We presented a case of FAP-associated aggressive mesenteric fibromatosis, presenting as a palpable abdominal mass, with computed tomography (CT) and magnetic resonance imaging (MRI) findings of this rare neoplasm.

Case report

A 34-year-old female patient presented with a palpable solid tumefactive mass in the left hemiabdomen. On abdominal examination, the mass was firm, approximately 22 x 20 cm in diameter, immobile with no associated tenderness. Her general physical examination was unremarkable. All her baseline blood investigations were normal. Contrast enhanced CT revealed multiple massive solid tumefactions in the mesentery and in between the small bowel loops, inhomogeneous in structure, the largest one being approximately 131 × 101 × 190 mm in diameter (Figure 1). CT angiography of the abdomen revealed the tumor was vascularized via branches of the superior mesenteric artery, with venous drainage via the superior mesenteric vein. There was no significant reduction of lumen of the celiac trunk and superior mesenteric artery. A colonoscopy was performed



A) **B)**

Fig. 1 – Contrast enhanced computed tomography (CT) of the abdomen: A) transversal and B) coronal images show. A large inhomogeneous solid mass in the mesentery with arterial supply via branches of the superior mesenteric artery.

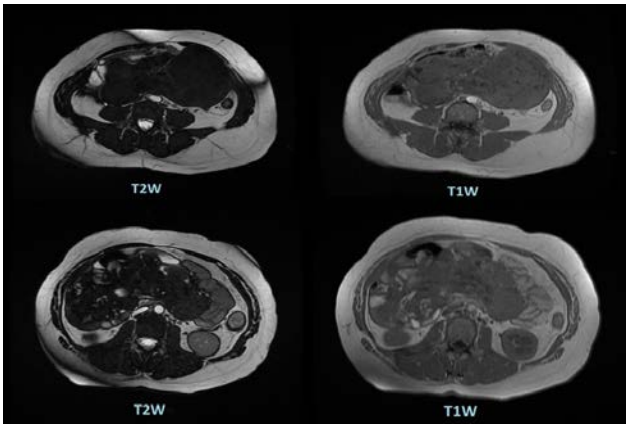


Fig. 2 – Magnetic resonance imaging scan. Transversal T2W and T1W images. Massive tumor formation stretching from the ascending colon to the anterior abdominal wall bearing a T2W hypointense signal with relative T1W isointensity.

which confirmed the presence of multiple sessile polypomatous lesions of different diameter characteristic of FAP with similar lesions visible in the stomach and duodenum on esophagogastroduodenoscopy. An MRI scan of the abdomen was performed a little bit later, showing a massive intraabdominal tumor formation, stretching from the ascending colon through the mesentery to the anterior abdominal wall, with slight compression of the left rectus abdominis muscle and contact with the descending colon. The tumor was hypointense on T2 weighted images, with a relative isointensity on T1W scans and no signs of restricted diffusion (Figure 2). There was mild dominantly peripheral inhomogeneous alteration of signal after a paramagnetic contrast was administered with prominent central hypovascularity (Figure 3). The superior mesenteric artery and celiac trunk were within close proximity of the tumor tissue with no signs of vascular compression or thrombosis.

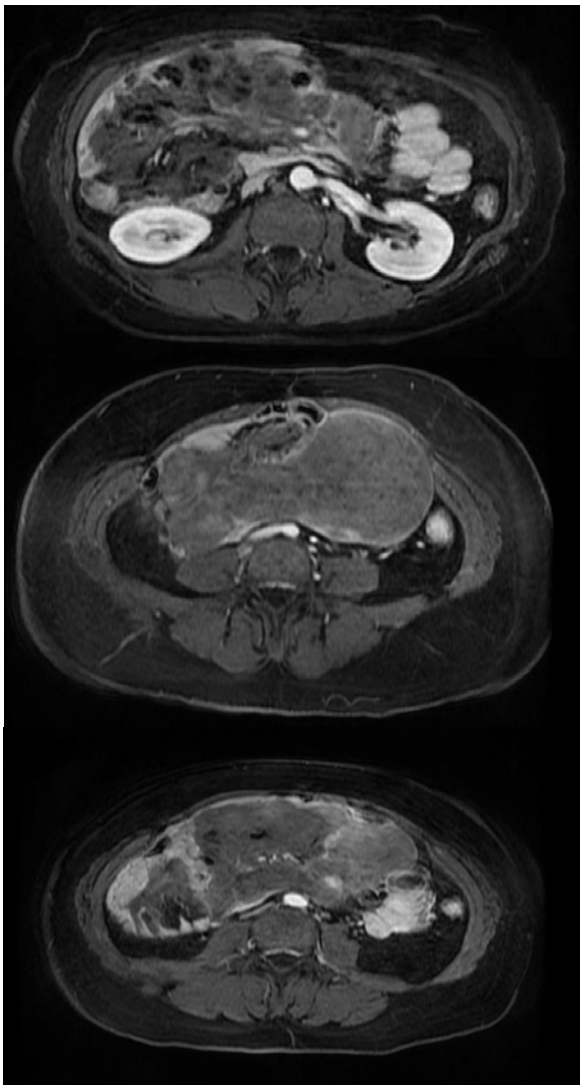


Fig. 3 – Magnetic resonance imaging scan. Transversal gadolinium enhanced liver acquisition with volume acquision images showing mild enhancement of the lesion with prominent central hypovascularity.

Tissue samples were acquired via ultrasound guided biopsy with histopathologic confirmation of DF with imunohistochemical analysis. After an abdominal surgeon was consulted, it was concluded that there was an increased risk of surgery due to the size of the lesion and increased possibility of bleeding, therefore the patient was planned for chemotherapy with potential further reevaluation for a possible surgical procedure.

Discussion

Desmoid tumors are rare tumors arising from musculoaponeurotic elements accounting for 0.03% of all tumors and 3.5% of all fibrous tissue tumors [5]. Patients with FAP, Gardner's syndrome, are especially predisposed to the development of mesenteric fibromatosis [6]. FAP is an autosomal dominant disease, caused by a germline mutation in the adenomatous polyposis coli tumor suppressor gene leading to the development of multiple colorectal adenomatous polyps. The modern management of FAP, incorporating predictive genetic testing and prophylactic surgery, has meant that extracolonic manifestations of FAP, particularly desmoid tumors and polyposis of the upper gastrointestinal tract are now the leading cause of mortality among patients having gone prophylactic colectomy [7–9].

Desmoid tumors develop in approximately 10% of patients with FAP and most are intraabdominal, following a more aggressive course with recurrence after resection being common. Aggressive mesenteric desmoids can lead to small bowel obstruction, ischaemia and perforation with surgical excision being difficult, due to their proximity to the superior mesenteric artery [4].

The most common imaging modalities used for detection and evaluation of DF are CT and MRI. Imaging characteristics of DF closely reflect the distribution of histologic components: spindle cells, myxoid matrix and collagenous stroma [10]. These tumors appear as soft tissue masses that are either sharply margined or with ill-defined infiltrative margins [11]. On CT images attenuation is variable, with the majority of masses demonstrating mild to moderate enhancement [12]. The signal intensity of the tumor on MRI is highly dependable on the proportion of collagen fibers, spindle cells and extracellular matrix present, with decreased signal intensity on T2 weighted images most likely resulting from dense collagen and hypocellularity, while increased T2 signal intensity reflects a high content of spindle cells [13–15]. Gadolinium enhancement is variable, with more prominent enhancement being present in the more cellular, less fibrotic areas.

Our mass demonstrated a significant signal hypointensity on T2 weighted images and mild gadolinium enhancement, which would suggest a very dominant presence of collagenous stroma.

Wide field surgical resection is the first line of treatment for most mesenteric fibromatosis with most cases requiring resection of the attached segment of the bowel [3]. Neurovascular structure encasement as well as invasion of the viscera should be detectable by imaging and included in

the radiologists report. Lesion location, multiplicity, infiltrative margins and relationship with mesenteric vessels and intraabdominal organs are important surgical considerations [2]. In our case, due to the localization of the tumor and its close proximity to the mesenteric vascular structures, the risk of surgery was deemed too high at the time, therefore chemotherapy was planned to achieve reduction in tumor size, which would make a future surgical procedure possible.

Conclusion

Mesenteric fibromatosis is a rare neoplasm that presents with a wide range of histologic and imaging features. CT and MRI are the imaging modalities of choice in the management of mesenteric fibormatosis. Defining lesion characteristics, localization, neurovascular and visceral affection are key in evaluating the potential for surgical resection, therefore playing a crucial role in planning an optimal treatment model.

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