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Case Report

Mesenteric desmoid tumor in its cystic form: Case report of a very rare variant ☆

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ABSTRACT

Desmoid tumors, also known as deep fibromatosis or desmoid-type fibromatosis, represent a rare subset of deep fibromatoses. It is a locally aggressive tumor, with no specific symptoms, and no metastatic potential. We report a case of a 38-year-old male patient with an abdominal mass. Radiological findings showed 2 tumors, the first was a solid inguinal mass of the left lateral iliac pedicle, and the second was a giant cystic mass in the abdominal cavity. An elective explorative laparotomy was performed to remove the 2 masses. Histopathological examination confirmed the desmoid tumor diagnosis of both lesions.

Introduction

Desmoid tumors, or desmoid-type fibromatosis, represent a rare subset of deep fibromatoses. It is a locally aggressive tumor, with no specific symptoms, and no metastatic potential.

Case presentation

A 32-year-old male patient presented with a left inguinal mass. Physical examination revealed a left inguinal mass, fix to the abdominal wall, with abdominal distension and significant weight loss. No fever or any another abdominal and extra-abdominal sign was associated. There was no family

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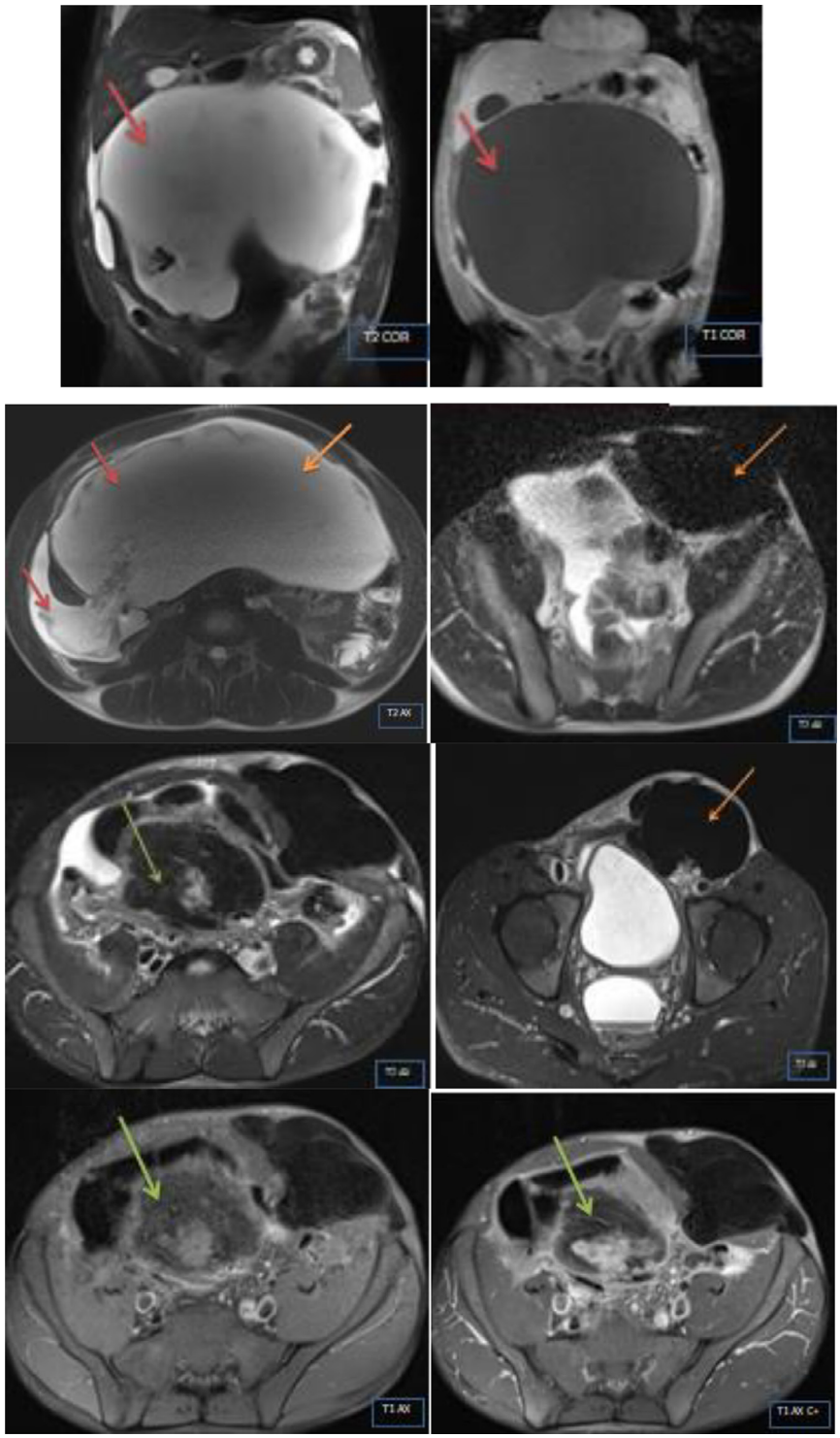

Fig. 1 – MRI in axial T2-weighted, coronal T2-weighted with fat saturation, and T1-weighted sequences with and without contrast injection reveals a sizable intra-abdominal mass occupying almost the entire abdomen, extending to the mesenteric root, displacing the loops and the liver, coming into contact with the aorta and iliac pedicle. This mass exhibits a dual cystic component, with hypointensity on T1 and hyperintensity on T2 (red arrow) for the cystic part, and hypointensity on T2 with heterogeneous enhancement postinjection for the solid tissue component (green arrow), measuring 14 x 16 cm. Additionally, there is another inguinal mass (orange arrow) with marked hypointensity on T2 and weak enhancement on T1 postinjection, measuring 106 x 82 x 176mm, without significant diffusion restriction. Moderate ascites is present.

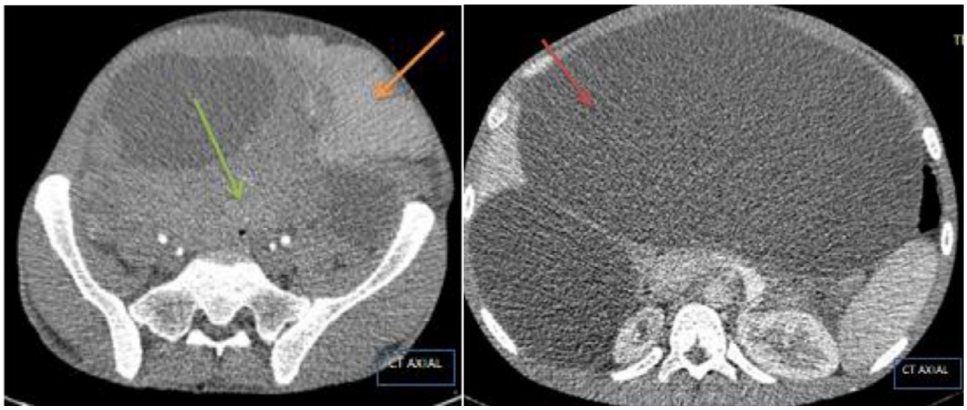

Fig. 2 – Axial CT scans depict the voluminous intra-abdominal mass with its dual components and the inguinal mass.

history of colon cancer, familial adenomatous polyposis (FAP), or personal history of abdominal trauma.

MRI (Fig. 1) reveals a sizable intra-abdominal mass occupying almost the entire abdomen, extending to the mesenteric root, displacing the loops and the liver, coming into contact with the aorta and iliac pedicle. This mass exhibits a dual cystic component, with hypointensity on T1 and hyperintensity on T2 (red arrow) for the cystic part, and hypointensity on T2 with heterogeneous enhancement postinjection for the solid tissue component (green arrow), measuring 14 × 16 cm. Additionally, there is another inguinal mass (orange arrow) with marked hypointensity on T2 and weak enhancement on T1 postinjection, measuring 106 × 82 × 176 mm, without significant diffusion restriction. Moderate ascites is present.

Axial CT (Fig. 2) scans depict the voluminous intra-abdominal mass with its dual components and the inguinal mass.

Surgical excision of the 2 masses was performed by laparotomy (Fig. 3).

Initially, a left inguinal incision was performed, dissection of the tumor after identification and release of the cord, (Fig. 4) the mass was confirmed to arise from the rectus muscle (Fig. 5), with no gross bowel wall involvement (Fig. 6).

Further dissection and release of the epigastric pedicles was carried out, along the tumor to the iliac bone (upper anterosuperior iliac spine) while respecting the peritoneum. the excision of the inguinal mass was enlarged to the abdominal wall to ensure free margins.

Then a midline sub umbilical laparotomy was performed showing a large cystic abdominal-pelvic tumor near the right colon and terminal ileum (Fig. 7), the tumor was in contact with the root of the mesentery and the abdominal aorta.

The cystic mass was punctured, approximately 8000 ml of intra-tumoral fluid was drained, tumor excision (Fig. 8) was carried out after freeing the adjacent organs (colon, small bowel and the aorta).

No pus, or hemorrhaging were found in the peritoneal cavity. Drainage at the end of the procedure was performed in order to drain the abdominal cavity.

To address the loss of significant muscle substance in the inguinal region, an abdominal binder « mesh » was installed (Fig. 9), ensuring intra-abdominal pressure and minimizing the risk of incisional hernia complications.

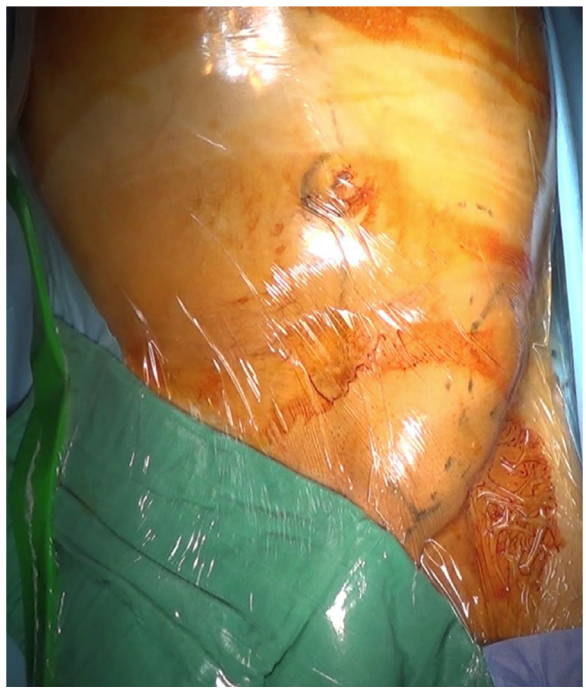

Fig. 3 – operative view of the Inguinal mass.

The postoperative course was uneventful, and the patient was discharged on postoperative day 10. The patient remained asymptomatic during a 3-month follow-up period.

The removed masses were sent to the department of pathology for histopathological study. On gross examination, the first tumor was cystic, measuring 17 × 11 × 8 cm, with tissular area measuring 9 cm in great diameter. The second mass was ill defined and irregularly shaped, with whitish fascicular cut surface and measured 19 × 11 × 8 cm. The latter was extremely firm at sectioning. Extensive sampling was performed. On histology, the first tumor consisted of elongated, slender, uniform spindled cells devoided of cytonuclear atypia; arranging in long sweeping fasciculs. Numerous thin walled vassels with perivascular oedema were seen. The second tumor showed variably sized bands of thick collagen fibers throughout the tumor; while neoplastic spindle cells

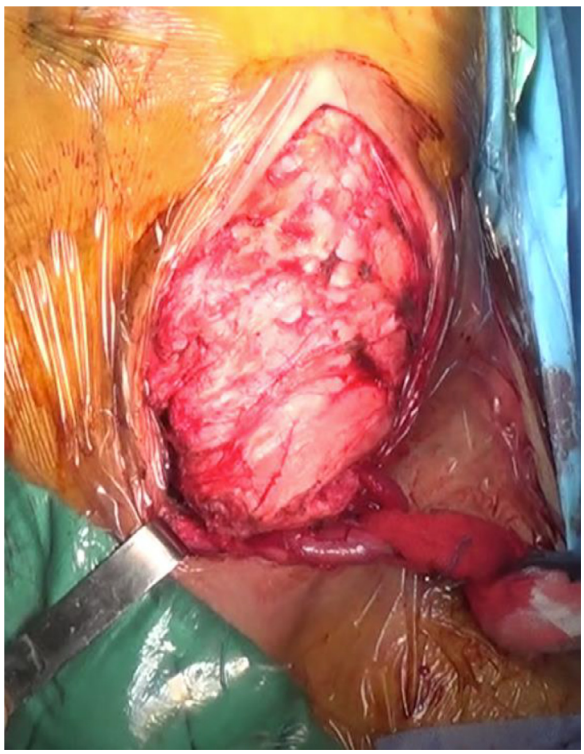

Fig. 4 – Dissection of the mass "M" after mobilizing the cord "C".

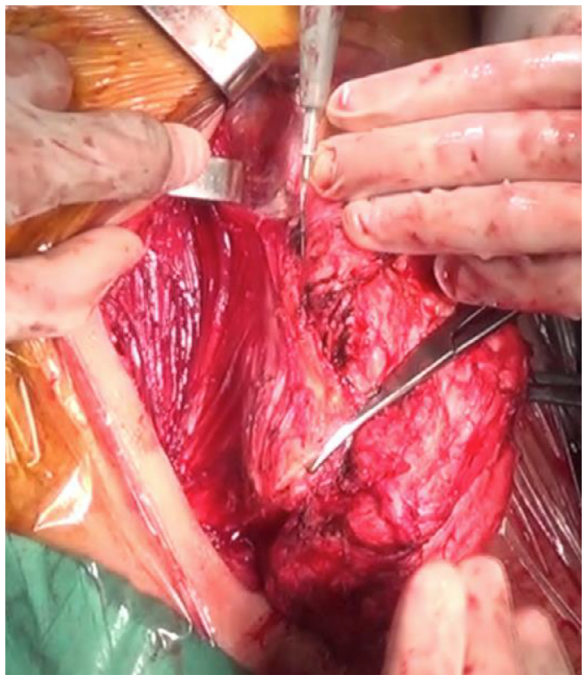

Fig. 5 – Completion of the dissection by freeing the rectus abdominis "RA" muscle.

were scarce and difficult to identify. There was no mitotic activity neither necrosis. At the periphery, both tumors had infiltrative margins. Immunohistichemistry was performed on both lesions, and showed diffuse positivity for: beta catenin (nuclear and paranuclear staining) and for smooth muscle actin. Neoplastic cells were negative for: desmin, S100 protein, CD34 and H-Caldesmon. On the basis of morphological and immunohistochemical findings, the definite diagnosis was: deep fibromatosis or desmoid type fibromatosis of conventional pattern (Fig. 10), and keloidal pattern (Fig. 11), for, respectively, the first and second tumor.

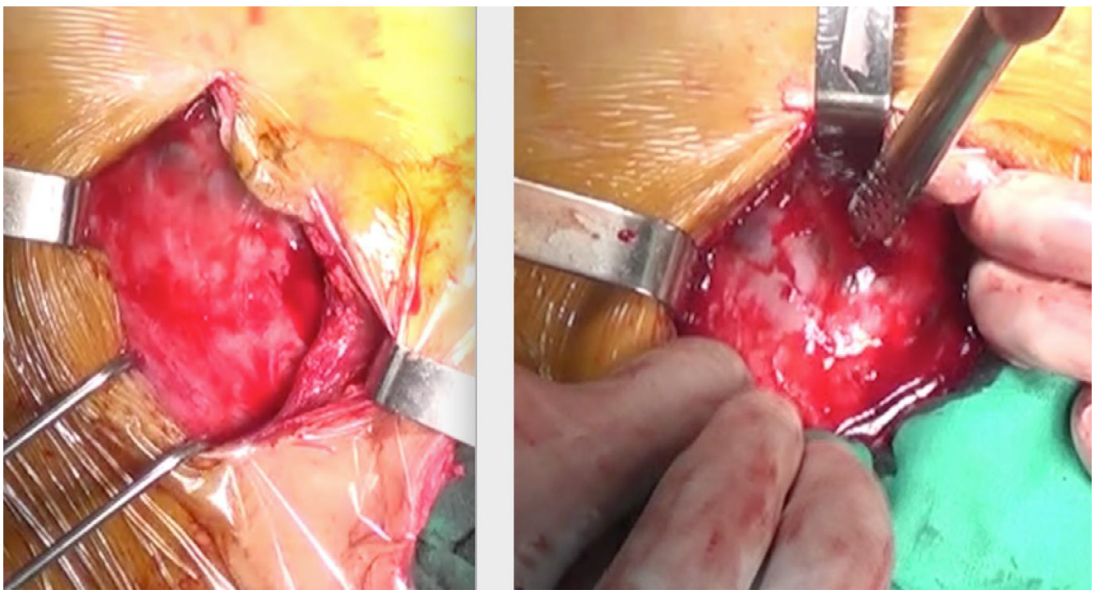

Fig. 6 – Operative view of the cystic abdominal mass and trocar puncture.

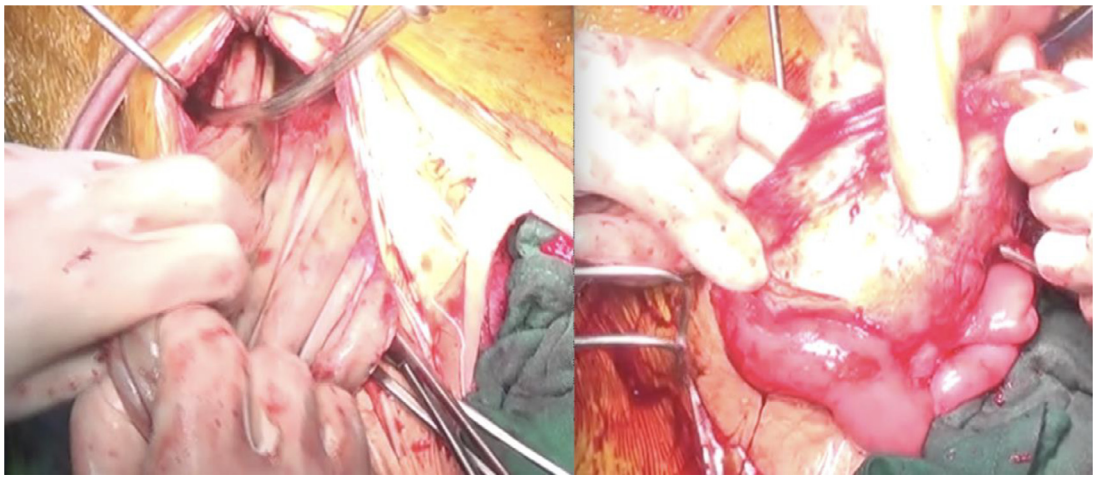

Fig. 7 – Dissection of the cystic mass "A", the mesentery and small bowel "B".

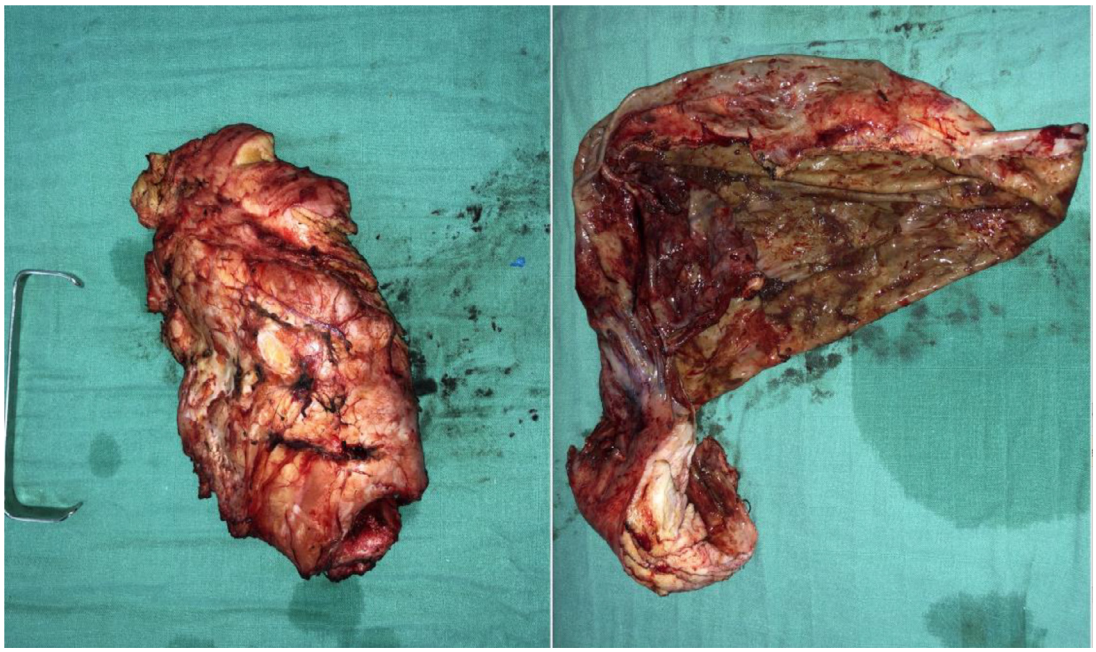

Fig. 8 – Macroscopic image of the specimen.

Discussion

Desmoid tumors, also known as deep fibromatosis (DT) or desmoid-type fibromatosis (DTF), represent a rare subset of deep fibromatoses, constituting less than 1% of retroperitoneal masses and less than 3% of all soft tissue tumors [1].

They represent less than 1% of retroperitoneal masses, 0.03% of all neoplasms and less than 3% of all soft tissue tumors. While locally aggressive, they exhibit no metastatic potential, although a few reports of spontaneous cystic degeneration in DF have been published, they however remain rare [2–4].

In the abdominal region, desmoid tumors can arise from either the deep abdominal wall soft tissue or intra-abdominally, the small bowel mesentery being the most common site.

Although frequently associated with polyposis syndromes, such as FAP and gardner's syndrom, most cases occur sporadically, with only 13% of patients having FAP, however it's incidence in FAP is approximately 850 times that of the general population [4,5,6,].

Their pathogenesis remains unknown, but hormonal, genetic and traumatic factors may be involved, causing a generalized defect in the growth regulation of connective tissue. Previous surgery is a less significant risk factor in sporadic cases compared to those with FAP (10% to 83%), and postresection recurrence appears to be higher in patients with FAP [1,6,7].

It is also classified on the basis of the anatomic location, such as abdominal wall, intra-abdominal, or extra-abdominal. In various studies, 28 to 69% of DF was intra-abdominal (mesenteric or pelvic) or located in the abdominal

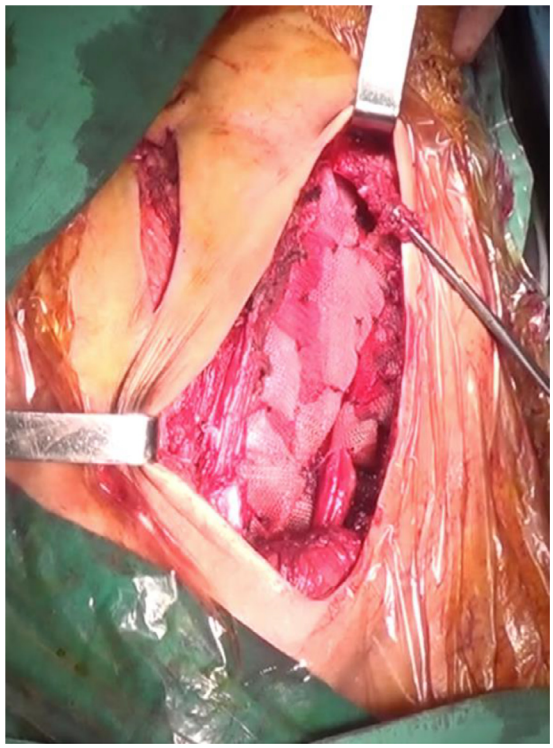

Fig. 9 – Abdominal wall reconstruction with "MESH".

wall. Retroperitoneal DF occurs in less than 1% of retroperitoneal masses [8].

They can occur at any age but are most prevalent in the third and fourth decade. Although both sexes may be affected, abdominal desmoids are more common in females, particularly those of childbearing age [7].

Desmoid tumors may present with a wide range of symptoms, and their clinical signs depend on the tumor's location. It ranges from incidental small lesions to rapidly-growing and aggressive abdominal masses, causing death within a few years or within months. Most patients present with an asymptomatic abdominal mass, but some have abdominal pain, paresthesia or obstructive symptoms (intestinal, vascular, ureteric, neural). Pain is not proportional to tumor size. Mesenteric DT can cause acute abdominal pain, hemorrhaging or peritonitis secondary to bowel perforation [3,6,8–10].

Imaging modalities like CT and MRI play a crucial The CT scan often reveals a substantial mass with lobulated contours , exhibiting spiculated contours creating a "whorled appearance" [11]. The appearance oftentimes resembles that of a solid lesion. However, we report an exceptionally rare presentation: a desmoid tumor in cystic form [12]. The absence of this solid component and the predominance of the cystic element may be secondary to a spontaneous regression of the tumor [13]. There is no evident organic abdominal origin [14]. The occurrence of calcifications, necrosis, or hemorrhage is uncommon [15]. In other cases described in the literature, a fatty component may be present [12].

Its presence poses a diagnostic challenge, particularly in the differential diagnosis with teratomas (which are the primary consideration when encountering a mass containing liquid and fat) and excavated lymph nodes.

The CT scan allows for the detection of complications such as intestinal obstruction and perforation, as well as mesenteric ischemia, fistulization [16], and abscess formation [17].

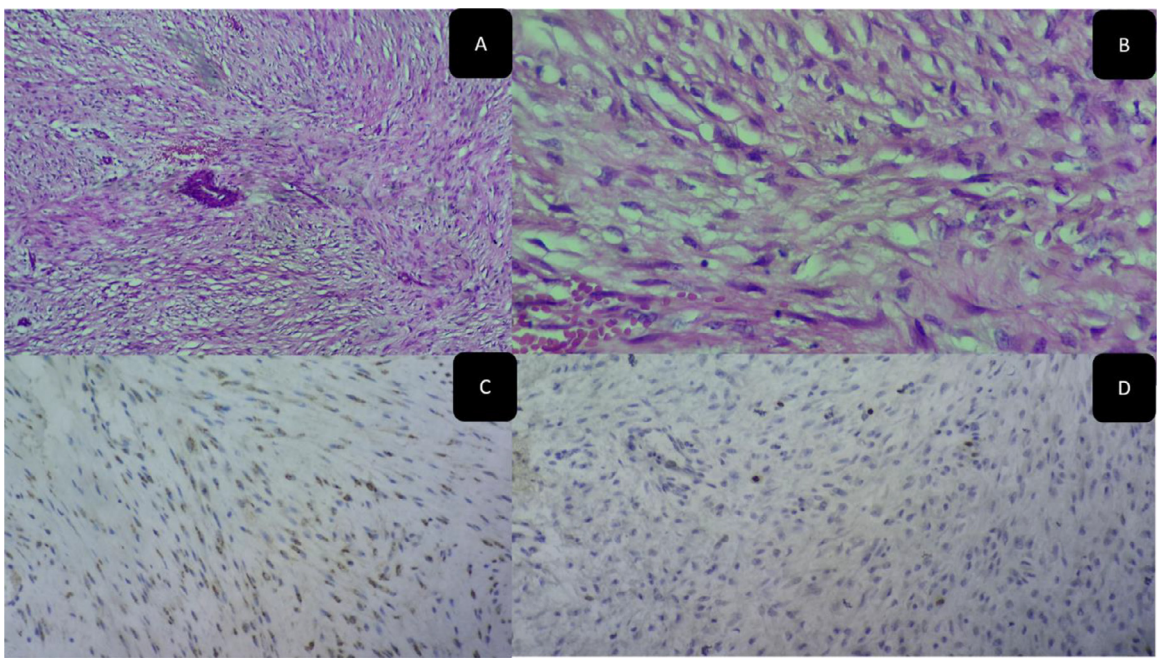

Fig. 10 – Desmoid-type fibromatosis conventional pattern histological and immunhistochemical aspects: fascicules of bland looking spindle cells, (A and B) (HE Stain) A: low power, B: high power, (C and D) (Immunohistochemistry stains) (C) positive Beta catenin, (D) low Ki 67 proliferation index.

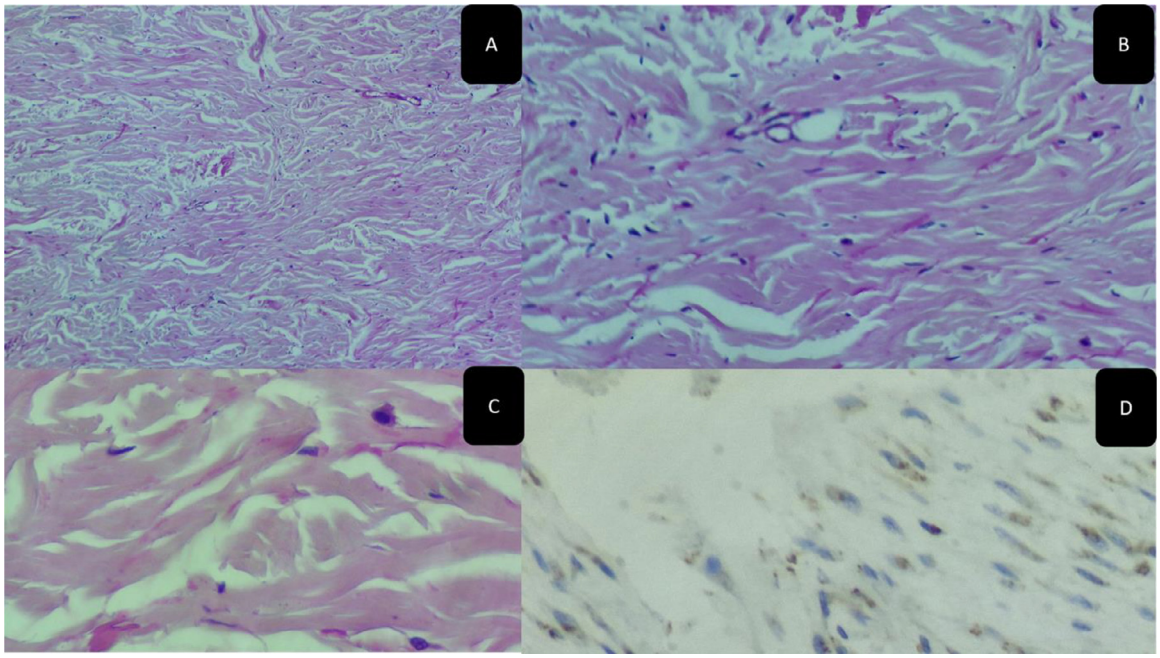

Fig. 11 – Desmoid-type fibromatosis keloid-like pattern type histological and immunhistochemical aspects: bands of thick collagen fibers throughout the tumor with scarce neoplastic spindle cells
(A-C) (HE Stain) (A) low power, (B) medium power, (C) high power, (D) (Betacatenin immunistain) positive Beta catenin.

On MRI, Tumors exhibit hypointensity on T1-weighted images and can manifest as hypo- or hyperintense on T2-weighted images. Reduced signal intensities on T2-weighted images indicate the dense collagen fibers and low cellularity of the tumor, while increased signal intensities on T2-weighted images suggest the presence of high spindle cells [18]. Small necrotic areas within the lesion are more discernible in MRI imaging. The diffusion-weighted sequence of MRI imaging (DWI and ADC) primarily aids in determining the dense hypercellularity within the tumor, as hypercellular tumors typically appear hypointense on ADC and hyperintense on DWI images [19,20]. The tumor can be either singular or multiple. The imaging appearance is not very suggestive.

Histological confirmation is mandatory for accurate diagnosis. Imaging aids in establishing a preoperative assessment, including the investigation of vascular involvement, the condition of the mesenteric pedicle, the degree of mesenteric invasion, and the size of the mass.

Upon radiological investigation, a variety of potential diagnoses is considered, including peritoneal carcinomatosis, peritoneal sarcoma, peritoneal lymphomatosis, mesothelioma, carcinoid tumor, peritoneal leiomyomatosis, intraperitoneal desmoplastic tumor, and peritoneal tuberculosis.

A multidisciplinary approach maybe be required, however surgical excision is the treatment of choice for intra-abdominal DT, although recurrence rate is high (15% to 30%) [1,8].

In an emergency setting (occlusion or peritonitis), treating only the mechanical complication and leaving the DT in place may be discussed, as the lumpectomy would lead to extensive digestive resection, especially in patients with FAP. The best indication for surgery remains patients who present parietal locations, since surgery can be performed with an acceptable morbidity [10].

Resection of the abdominal wall for desmoid tumors is safe, incomplete resection is associated with a high recurrence rate. Desmoid tumor should be resected in early small stages, otherwise abdominal wall repair could be technically demanding and challenging. Reconstruction can be achieved either by direct repair, or by using «MESH », or by myocutaneous flaps when the defect is extensive [1,4].

Surgery of Intraabdominal desmoid tumors is much more challenging, with a higher morbidity and mortality, which is due to hemorrhaging or extensive enterectomy. Most patients with these desmoid tumors have an unresectable disease (65%) [4,21].

Radical free margin excision is the treatment of choice, although it is not always a straightforward procedure on account of the tumor's extent and invasiveness [4].

Because of a high risk of local recurrence (30% to 50%) after surgical excision, there is a role for adjuvant treatment in patients with positive surgical margins. Improved progression-free survival rate of patients with unresected or partially resected tumors can be achieved with adjuvant radiotherapy [22].

Definite diagnosis requires pathological study [23]. Macroscopically, desmoid type tumors are ill defined, with infiltrative borders, showing whitish nodular cut surface. On histology, these tumors are composed of bland looking fibroblastic or myofibroblastic spindle cells arranging in a sweeping

fascicular pattern and showing numerous thin-walled blood vessels with characteristic perivascular oedema and infiltrative margins. The spectrum of desmoid tumors is broad, including different histological patterns such as: myxoid, nodular fasciitis-like, hypocellular and keloidal-like pattern, as described in the present case. In fact, the latter is reported in approximately 15% of the cases [23]. On immunohistochemistry, the hallmark of the tumor is the nuclear accumulation of beta-catenin [24]. Mutational analysis (mutations of the CTNNB1 gene and APC gene) might be required in challenging cases, especially beta catenin negative tumors and for hereditary cases as well [23,24]. The differential diagnosis is wide, including : solitary fibrous tumor, nodular fasciitis, synovial carcoma, myofibroma, gastrointestinal stromal tumor (GIST), low-grade fibromyxoid sarcoma, inflammatory myofibroblastic tumor and in rare instances rhabdomyosarcoma [23].

Conclusion

we report an extremely rare case of retroperitoneal and intraperitoneal DF, with 2 different components. Clinical signs depend on the tumor's location. Imaging modalities like CT and MRI are the key to diagnosis.

Ethics approval

Our institution does not require ethical approval for reporting individual cases.

Patient consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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