

DOC-20230112-WA0025..pdf

by

Submission date: 18-Jan-2023 09:18AM (UTC+0700)

Submission ID: 1994497528

File name: DOC-20230112-WA0025..pdf (232.14K)

Word count: 2162

Character count: 12918

Diagnostic and Therapeutic Challenges of Intra-abdominal Desmoid-Type Fibromatosis Mimicking Abscess: A Case Report

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Keywords:

Desmoid Fibromatosis Tumor, Intra-abdominal, chemotherapy, recurrent

ABSTRACT

Desmoid tumor is a rare disease that originates from the proliferation of fibrous tissue. Intra-abdominal desmoid tumors are sporadic or associated with specific familial syndromes, such as Familial Adenomatous Polyposis Syndrome (FAP). These tumors are often challenging to diagnose, and active surveillance is so essential given the high recurrence rate despite adequate treatment. We present the case of an 18-year-old female with a recurrent intra-abdominal desmoid fibromatosis tumor despite surgical resection combined with radiotherapy. The recurrences are treated with anthracycline chemotherapeutic regimens. There is a diagnostic delayed due to nonspecific presentations. Immunohistochemistry assay is a cornerstone for accurate histopathological diagnosis, which guides treatment. The mainstay of treatment is surgical resection; chemotherapy and radiotherapy have a role in managing both primary and recurrent lesions.



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Introduction

Desmoid tumor or aggressive deep-seated fibromatosis is a rare disease that originates from the proliferation of fibrous tissue [1]. In the general population, desmoid tumors are found in approximately 0.03% of all neoplasms and less than 3% of soft tissue tumors. Desmoid tumors usually occur in reproductive-age women and rarely occur after menopause. Several studies have shown that 37-50% of desmoid tumors occur intra-abdominal [2]. Intra-abdominal desmoid tumors are sporadic or associated with specific familial syndromes, such as Familial Adenomatous Polyposis Syndrome (FAP). These tumors are often considered chronic diseases and require active surveillance regardless of therapeutic strategy. They are potentially recurrent and lethal due to their locally aggressive invasion and ability to invade adjacent vital organs and structures [3]. The management of desmoid tumors is complex and requires a multidisciplinary team.

We present the case of an 18-year-old female with a recurrent intra-abdominal desmoid fibromatosis tumor despite surgical resection combined with radiotherapy. The recurrences are treated with anthracycline

chemotherapeutic regimens.

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2. Case Report

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An 18-year-old woman with no significant medical or family history was referred to our Department with complaints of left lower quadrant abdominal pain of a 1-month duration. During a physical examination, abdominal tenderness and scar from a previous exploratory laparotomy. Laboratory tests are within normal limits. Seven months before the presentation patient underwent consultation at the obstetric gynecology department with suspicion of an adnexa tumor. Abdominal computed tomography (CT) showed a left psoas abscess suggestive of TB that extends to the retroperitoneal and pelvic cavities, pushing the left kidney and left ureter cranially and abdominal aorta and left iliac artery to the right (Figures 1A and B).

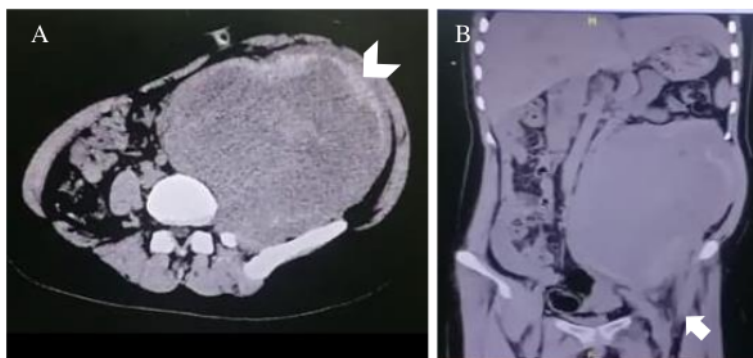


Figure 1. Abdominal computed tomography (CT) findings. Horizontal (A) and coronal (B) views by CT before surgical debulking (14/09/2020). Abdominal CT showed a left psoas abscess (white arrow) suggestive of TB that extends to the retroperitoneal and pelvic cavities (arrowhead), pushing the left kidney and left ureter cranially as well as abdominal aorta and left iliac artery to the right.

The patient was referred to the surgery department and underwent a biopsy that revealed a benign cyst (Figures 2A, B, and C). Three weeks after that, the patient underwent exploratory laparotomy and tumor debulking. The histopathology findings are the proliferation of spindle-shaped cells with collagenous stroma in the tumor tissue (Figures 3A and B). Hence, the final pathological diagnosis was intra-abdominal DF.

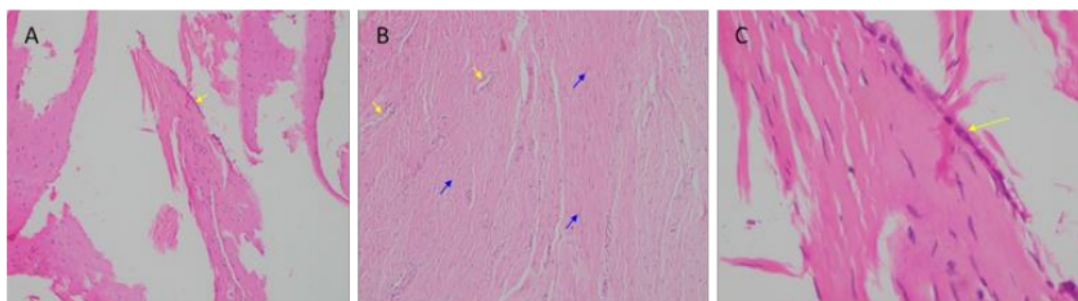


Figure 2. Histology findings of the biopsy specimen. Hematoxylin and eosin staining of the biopsy specimen cyst with one layer cuboid epithelial lining (yellow arrow) 4 times magnification (A), spindle-shaped fibrous tissue (blue arrow) surrounded with blood vessel (yellow arrow) 12 times magnification (B), cyst with one layer cuboid epithelial lining (yellow arrow) 100 times magnification (C)

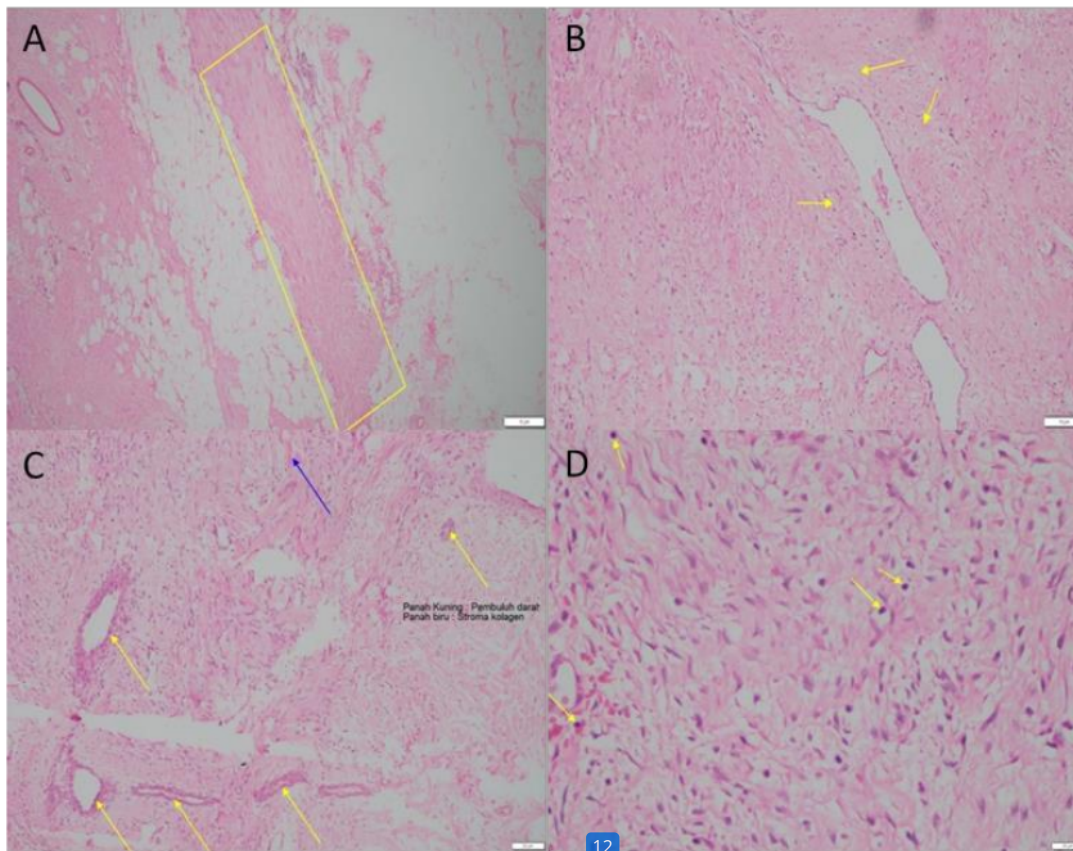


Figure 3. Histology findings of the resection specimen. Hematoxylin and eosin staining of the specimen confirmed a spindle cell morphology (yellow box) and focal invasion of the adipose tissue on low power (A), perivascular oedema (yellow arrow) 10 times magnification (B), collagen stroma (blue arrow) with blood vessel (yellow arrow) 10 times magnification (C), spindle cell morphology with lymphocyte infiltration (yellow arrow) 40 times magnification (D).

However, the boundary between the tumor and the average area was unclear, and no evident positive margin was observed. After debulking surgery, the patient continued radiotherapy 25 times. Despite radiotherapy, the patient still complains of abdominal pain dan progressive enlargement of abdominal mass. The patient was then referred to the hematology-oncology division of the internal medicine department and planned for immunohistochemistry and estrogen receptor assay. The immunohistochemical study showed that the tumor cells were positive for vimentin and negative for cytokeratin (Figures 4A and B). Estrogen receptors turn out to be negative. These featured consistent with a diagnosis of desmoid fibromatosis and underwent chemotherapy, with six cycles of doxorubicin and dacarbazine regimen. The patient did well post-chemotherapy. She continues to undergo surveillance for recurrence.

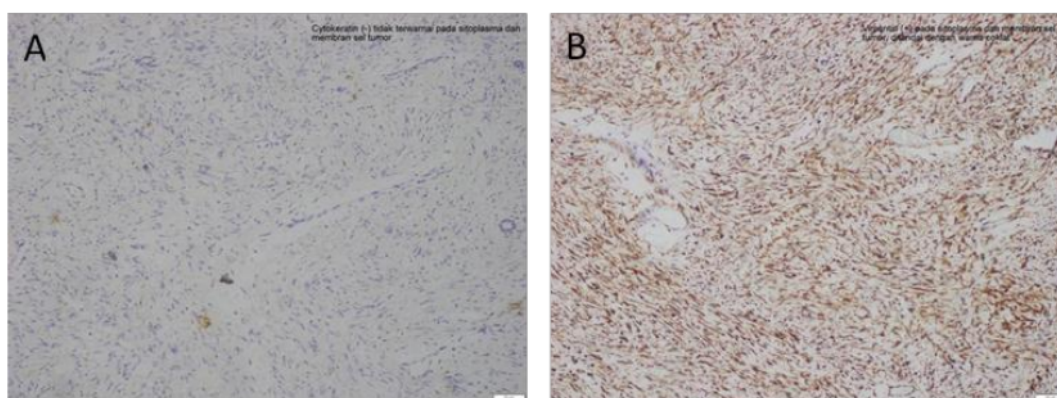


Figure 4. Immunohistochemistry findings of the resection specimen. Immunohistochemistry was negative for cytokeratin 40 times magnification (C) and positive for vimentin 40 times magnification (D).

3. Discussion

Desmoid tumors are common during puberty and the fourth decade [4]. The prevalence of cases is 3.7 per one million people yearly [5]. The etiology of desmoid tumors is unknown; it is multifactorial, with genetic, hormonal, and physical factors playing an important role in tumor development and growth [6]. Risk factors include high estrogen levels or germline predisposition such as FAP syndrome [2]. In this patient, FAP syndrome markers were not examined due to limited resources. There were no risk factors for desmoid tumors, such as a family history of colon cancer, history of abdominal trauma, and history of FAP syndrome. However, in some cases, desmoid tumors are found in patients without a history of colon cancer, abdominal trauma, or FAP syndrome [7].

Intra-abdominal desmoid tumors manifest as localized pain, palpable masses, or local complications, such as intestinal obstruction, bowel ischemia, fistulization, and urethral compression with obstructive nephropathy [8]. Although benign, the disease can cause death due to aggressive local invasion through compression or obstruction of the digestive system, urinary system, or blood vessels [9]. The patient has abdominal complaints; cross-sectional imaging reveals a locally advanced tumor.

Histopathologically, desmoid cells are spindle-shaped or edematous to some extent, caused by cell proliferation between massive collagen, small vessels, and spherical edema fibrous connective tissue. Opaque tumor cells have spindle-shaped to short nuclei with abundant cytoplasm. These cells are slightly dysmorphic, arranged in bundles, accompanied by a picture of nuclear mitosis, sometimes with mucoid degeneration and hyalinization [10]. In this case, histopathological results were obtained, consisting of spindle-core, non-atypical, entirely cellular, arranged in long fascicles. These grow infiltratively into muscle tissue and fat cells. Among them are visible collagen stroma and many vascular structures with perivascular edema accompanied by infiltration of surrounding lymphocytic inflammatory cells.

The immunohistochemistry assay was positive for vimentin and negative for cytokeratin and estrogen receptors. Vimentin is a type III filament protein encoded by the VIM gene, expressed in mesenchymal cells [11]. It is also expressed in certain carcinomas such as renal carcinoma, lymphoma, and melanoma. Vimentin positivity is of limited value in diagnosing soft tissue tumors; however, if the mesenchymal tissue is damaging for vimentin, this may indicate that the tissue is not differentiated from mesenchymal [12]. Cytokeratin is a keratin-containing intermediate filament protein found in the intracytoplasmic cytoskeleton of epithelial tissue, indicating carcinoma. However, it is also frequently found in several sarcomas such as

1 synovial sarcoma, epithelioid sarcoma, epithelioid hemangioendothelioma, angiosarcoma, and desmoplastic small round cell tumor [13]. Estrogen receptor negativity does not necessarily mean that the tumor is insensitive to estrogen and is not affected by antiestrogens [14].

There is no standard therapy for desmoid tumors due to their characteristics of local invasion and high recurrence rate. According to 2021 National Cancer Comprehensive Network guidelines, treatment of desmoid tumors includes surgery, radiotherapy, systemic therapy (i.e., chemotherapy, hormonal therapy, NSAID), and conservative management [15]. The mainstay of therapy for desmoid tumors is surgery with wide microscopic resection of the edges. However, there is a very high probability of local recurrence [9]. Adjuvant radiotherapy improves local tumor control; in patients undergoing surgical resection, the local tumor control rate at five years increases from 53% to 81% when combined with radiotherapy [7]. Despite surgical resection and radiotherapy, the patient showed recurrences. Then the patient was continued with a chemotherapy regimen of doxorubicin, dacarbazine, and celecoxib, and there is clinical improvement.

In addition to reducing swelling and pain, [15] nonsteroidal anti-inflammatory drugs have been suggested to treat desmoid tumors based on the high expression of cyclooxygenase (COX)-1 and COX-2 observed in desmoid cells in vitro [2]. Conventional dose chemotherapy is an option in cases where time to treatment response is critical (e.g., for intra-abdominal or head and neck desmoid tumors). The doxorubicin combination regimen with dacarbazine is incredibly effective and safe, even in desmoid tumor patients who are unresponsive to conventional hormone therapy [16]. Desmoid tumors respond better to anthracycline regimens (54%, non-anthracyclines 12%) [17].

8 A high level of clinical suspicion is required to diagnose desmoid tumors. The management of desmoid tumors is complex and requires a multidisciplinary team. In case of recurrences after surgical resection and adjunctive radiotherapy, chemotherapy with anthracyclines regimens showed a good response. Active surveillance is also essential, given the high recurrence rate despite adequate treatment.

16 Summary

We reported a case of intra-abdominal desmoid fibromatosis tumor in an 18-year-old woman with recurrence after surgical resection and radiotherapy; she was given NSAID therapy and chemotherapy with doxorubicin and dacarbazine regimen, with good clinical outcome.

5. References

- [1] Napolitano A, Mazzocca A, Ceruso MS, et al. Recent advances in desmoid tumor therapy. *Cancers (Basel)*. 2020;12(2135):1-13. doi:10.3390/cancers12082135
- [2] Sanchez-Mete L, Ferraresi V, Caterino M, et al. Desmoid Tumors Characteristics, Clinical Management, Active Surveillance, and Description of Our FAP Case Series. *J Clin Med*. 2020;9(12):4012. doi:10.3390/jcm9124012
- [3] Escobar C, Munker R, Thomas JO, Li BD, Burton G V. Update on desmoid tumors. *Ann Oncol*. 2012;23(3):562-569. doi:10.1093/annonc/mdr386
- [4] Otero S, Moskovic EC, Strauss DC, et al. Desmoid-type fibromatosis. *Clin Radiol*. 2015;70(9):1038-1045. doi:10.1016/j.crad.2015.04.015
- [5] Bui N, Kummar S. Molecular insights into desmoid tumors. *Oncotarget*. 2017;8(53):90608-90609.

doi:10.18632/oncotarget.21293

[6] Ma JH, Ma ZH, Dong XF, Yin H, Zhao YF. Abdominal wall desmoid tumors: A case report. *Oncol Lett.* 2013;5(6):1976-1978. doi:10.3892/ol.2013.1297

[7] Zenzri Y, Yahyaoui Y, Charfi L, et al. The Management of Desmoid Tumors: A Retrospective Study of 30 Cases. *Int J Surg Oncol.* 2020;2020. doi:10.1155/2020/9197216

[8] Ivanova I, Arif S, Dinev D, Kalchev K, Atanassova M, Kotzev I. Desmoid abdominal tumor - A clinical case report and brief literature review. *Gastroenterol Insights.* 2017;8(1):1-13. doi:10.4081/gi.2017.6665

[9] Jafri SF, Obaisi O, Vergara GG, Cates J, Singh J, Feedback J. Desmoid type fibromatosis: A case report with an unusual etiology. 2017;9(9):385-389. doi:10.4251/wjgo.v9.i9.385

[10] Wang Y fei, Guo W, Sun K kun, et al. Postoperative recurrence of desmoid tumors: Clinical and pathological perspectives. *World J Surg Oncol.* 2015;13(1):1-8. doi:10.1186/s12957-015-0450-8

[11] Eriksson JE, Dechat T, Grin B, et al. Introducing intermediate filaments: From discovery to disease. *J Clin Invest.* 2009;119(7):1763-1771. doi:10.1172/JCI38339

[12] Liu CY, Lin HH, Tang MJ, Wang YK. Vimentin contributes to epithelial-mesenchymal transition cancer cell mechanics by mediating cytoskeletal organization and focal adhesion maturation. *Oncotarget.* 2015;6(18):15966-15983. doi:10.18632/oncotarget.3862

[13] Wei S, Henderson-Jackson E, Qian X, Bui MM. Soft tissue tumor immunohistochemistry update illustrative examples of diagnostic pearls to avoid pitfalls. *Arch Pathol Lab Med.* 2017;141(8):1072-1091. doi:10.5858/arpa.2016-0417-RA

[14] Leithner A, Gapp M, Radl R, et al. Immunohistochemical analysis of desmoid tumors. *J Clin Pathol.* 2005;58(11):1152-1156. doi:10.1136/jcp.2005.026278

[15] von Mehren M, Kane JM, Agulnik M, et al. Soft Tissue Sarcoma, Version 2.2022, NCCN Clinical Practice Guidelines in Oncology. *J Natl Compr Canc Netw.* 2022;20(7):815-833. doi:10.6004/jncn.2022.0035

[16] Williams AD, Heightchew K, Siripirapu V. Diagnostic and therapeutic dilemmas in intra-abdominal desmoid tumors: A case report and literature review. *Int J Surg Case Rep.* 2016;26:150-153. doi:10.1016/j.ijscr.2016.07.044

[17] Garbay D, Le Cesne A, Penel N, et al. Chemotherapy in patients with desmoid tumors: a study from the French Sarcoma Group (FSG). *Ann Oncol Off J Eur Soc Med Oncol.* 2012;23(1):182-186. doi:10.1093/annonc/mdr051

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