Article published online: 2025-12-18

Interdigitating Dendritic Cell Sarcoma of the Rectum: A Case Report with Review of the Literature

Jitin Goyal[1] Palak Gandhi[1] Prerna Garg[1] Shivendra Singh[2] Anurag Mehta[3] Sunil Kumar Puri[1]

[1] Department of Radiology, Rajiv Gandhi Cancer Institute and Research Centre, New Delhi, India
[2] Department of GI Surgical Oncology, Rajiv Gandhi Cancer Institute and Research Centre, New Delhi, India
[3] Department of Pathology; Rajiv Gandhi Cancer Institute and Research Centre, New Delhi, India

Address for correspondence Jitin Goyal, MD, MBBS, DNB, Department of Radiology, Rajiv Gandhi Cancer Institute and Research Centre, Rohini Sector 5, New Delhi 110085, India (e-mail: jitin27031992@gmail.com).

Ind J Med Paediatr Oncol

Abstract

Interdigitating dendritic cell sarcoma (IDCS) is a rare cancer that arises from dendritic cells, which function as antigen-presenting cells within the immune system. The manifestation of IDCS in the gastrointestinal tract, particularly the rectum, is exceedingly rare. We describe a case of extranodal IDCS in the rectum of a young female patient who presented with hematochezia for 18 months. Her clinical assessment was performed in August 2023 at a tertiary oncology center at Rajiv Gandhi Cancer Institute and Research Centre in New Delhi. Clinical workup included digital rectal examination, sigmoidoscopy, pelvic magnetic resonance imaging, and positron emission tomography/computed tomography. Biopsy via sigmoidoscopy suggested a spindle cell tumor of neural origin. The patient underwent transanal tumor excision. Histopathology and immunohistochemistry confirmed IDCS with S100 positivity. The patient was placed on regular surveillance with follow-up every 3 months. A comprehensive literature review was conducted to assess existing reports of gastrointestinal IDCS, especially rectal involvement. Very few cases have been documented, with only two other confirmed rectal IDCS cases reported to date. This case emphasizes the accurate diagnosis and prompt management of rectal IDCS, adding to the limited existing literature.

Keywords
- case report
- interdigitating dendritic cell sarcoma
- rectum
- MRI
- PET/CT
- antigen-presenting cells

Introduction

Dendritic cells (DCs) are a class of antigen-presenting cells differentiated from hematopoietic stem cells and are essential to adaptive immunity. The four primary types of DCs include follicular, interdigitating, Langerhans, and fibroblastic cells. Interdigitating DCs (IDCs) are typically located in T cell zones of secondary lymphoid organs such as lymph nodes and spleen. IDCs are frequently observed in middle-aged males; however, variation in age group is observed, ranging from childhood to old age. Although predominantly reported in lymph nodes, IDC sarcoma (IDCS) has been documented in extranodal locations, including the liver (27%), spleen (18%), skin (15%), and rarely in the gastrointestinal (GI) tract. Rectal involvement remains a medical rarity. This case report highlights the rare occurrence of IDCS in the rectum, supported by radiological imaging and histological data. The report also presents a literature review to better understand the clinical, radiological, and pathological spectrum of the disease.

DOI https://doi.org/10.1055/s-0045-1814393.
ISSN 0971-5851.

Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India

Case Report

A 33-year-old adult female patient presented with complaints of bleeding per rectum and features of obstructed defecation syndrome. Digital rectal examination reveals a protruding intraluminal mass at a distance of 4 cm from the anal verge. Magnetic resonance imaging (MRI) was performed on 3-Tesla MRI Magnetom Vida, with torso coil and the post-contrast images were acquired after administration of gadolinium meglumine intravenous contrast agent.

MRI revealed a polypoidal pedunculated hyperintense altered signal intensity mass in the upper-mid rectum measuring 4.7 AP (anteroposterior) × 5.9 TR (transverse) × 4.9 CC (craniocaudal) cm with a peduncle hypointense stalk length of 2.8 cm in CC extent, showing diffusion restriction with low apparent diffusion coefficient and heterogeneous contrast enhancement (►**Fig. 1**).

Sigmoidoscopy with biopsy was performed. It revealed a large, round lesion of size 4 to 5 cm at the 7 o'clock position. Microscopic examination showed interwoven fascicles of slender spindle cells—suggestive of a spindle cell lesion. On immunohistochemistry (IHC), spindle cells were strongly positive for S100 and SOX10, and negative for SMA, CK, CD34, CKit, and HMB45. The proliferation K_i-67 index was found to be approximately 2%. Final diagnosis was confirmed as a spindle cell tumor of neural origin with low aggressiveness.

Whole-body positron emission tomography/computed tomography (PET/CT) was done for staging and revealed no distant metastases. The patient underwent transanal excision surgery after discussion with the medical tumor board. Histopathology sections (►**Fig. 2**) showed a relatively well-circumscribed submucosal tumor, infiltrating into the rectal muscularis propria. The tumor was lobulated and exhibited a storiform and fascicular growth pattern and invasive margins, with inflammatory cells predominantly lymphocytes and plasma cells in a perivascular location and foamy histiocytes. The tumor cells were spindled with ill-defined cytoplasmic borders, vesicular nuclei, and contained an inconspicuous nucleolus. Mitotic activity was scant. No necrosis/apoptosis was observed. On performing IHC (►**Fig. 2**), the tumor cells were immunopositive for S100, EMA, TLE (moderate), SOX10, and CD68 (speckled), and negative for CK, HMB45, MELAN A, NUT, SMA, SMMH, CD10, ER, STAT-6, CD34, CD21/23, and CXCL13. BRAFV600E mutant protein was not present, which rules out melanoma. INI-1 and H3K27Me3 were retained. Proliferation K_i-67 index of approximately 2% favored the spindle cell tumor of neural origin of low aggressiveness. Final diagnosis of IDCS of low aggressiveness was confirmed.

The postoperative course was unremarkable. The patient was followed up at 3 and 6 months with colonoscopy and ultrasonography abdomen with no recurrence to date. The patient was kept under surveillance and regular follow-up while this case was being reported.

Discussion

IDCs are found in the T-cell region of secondary lymphoid tissue, where they function as antigen-presenting cells.

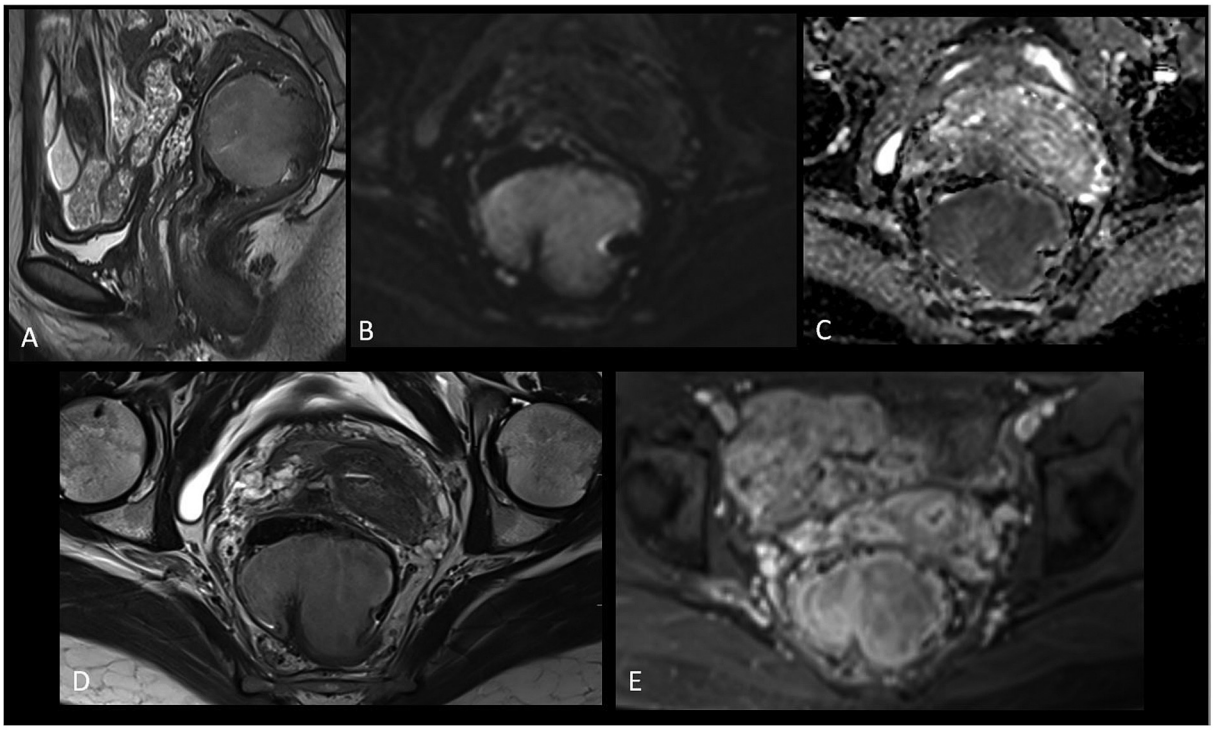

Fig. 1 CEMRI PELVIS: T2 sagittal (**A**), DWI and ADC (**B** and **C**), T2 axial (**D**), and post-contrast T1 axial (**E**). Polypoidal pedunculated hyperintense altered signal intensity mass in upper-mid rectum (A) measuring 4.7 AP × 5.9 TR × 4.9 CC cm with a peduncle hypointense stalk length of 2.8 cm approx. in CC extent., showing diffusion restriction with low ADC (B and C) and heterogeneous contrast enhancement (E). ADC, apparent diffusion coefficient; DWI, diffusion-weighted imaging.

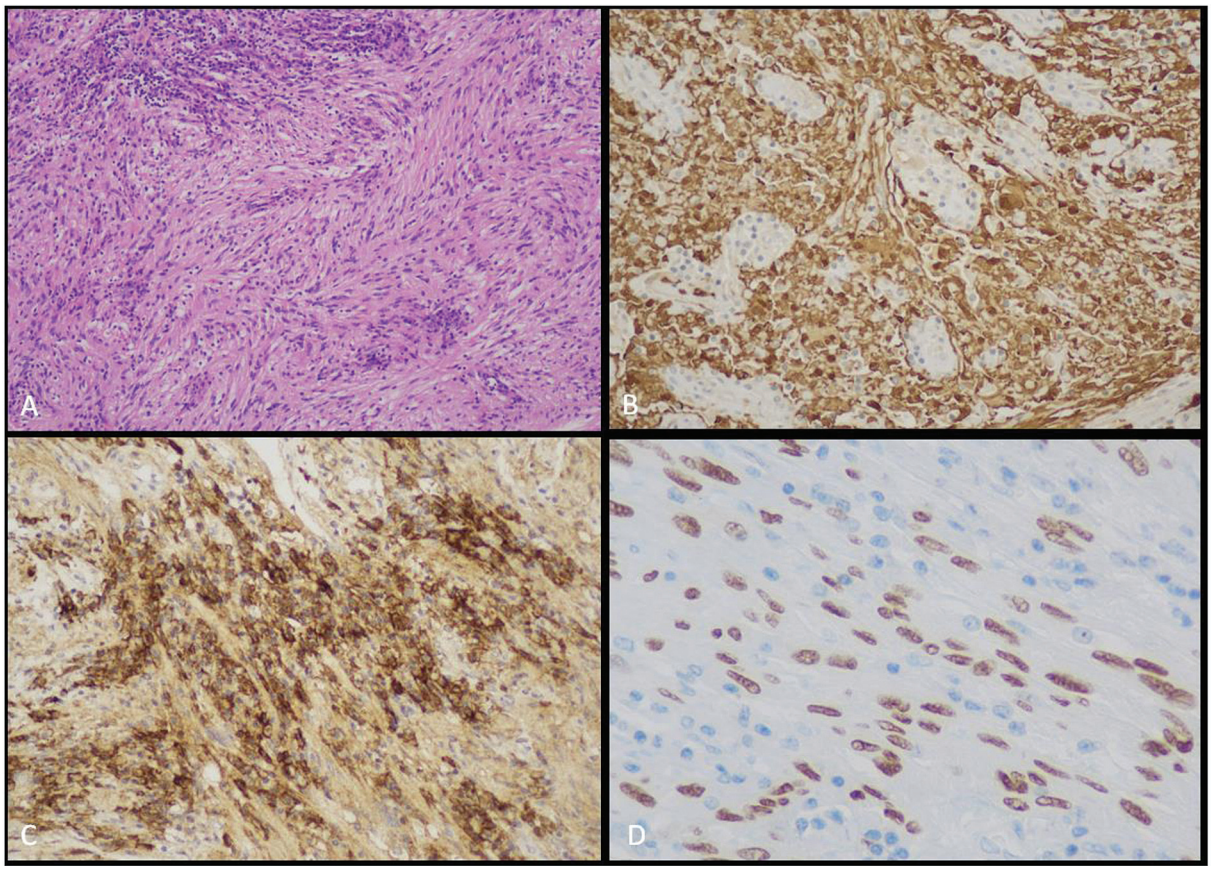

Fig. 2 H/E image HIGH power view: interdigitating dendritic cell sarcoma: tumor cells infiltrating into the rectal muscularis propria, exhibiting a storiform and fascicular growth pattern and invasive margins with inflammatory cells, predominantly lymphocytes and plasma cells in perivascular location, and foamy histiocytes. The tumor cells are spindled with ill-defined cytoplasmic borders, vesicular nuclei, and contain an inconspicuous nucleolus. Mitotic activity is scant, with no necrosis/apoptosis seen (**A**). On immunohistochemistry, tumor cells are immunopositive for S100, EMA, and SOX10 (**B–D**).

Proliferative disorders and neoplasms affecting IDCs are uncommon. IDCS is a rare neoplasm in the group of histiocytic and DC neoplasms.[1–3] It is often found in lymph nodes. Extranodal IDCS cases are very rare, with only a few cases reported in the medical literature. The most common site for extranodal IDCS is the liver (27%), followed by spleen (18%), skin (15%), lung (12%), and small intestine (11%).[4] The small intestine is the most frequent GI site for IDCS, where lesions are usually ulcero-proliferative.

All three cases reported in the literature, as well as the present case in the rectum, contain a polypoidal mass. Shibutani et al[5] reported the first rectal IDCS in a 76-year-old male patient with hematochezia and a mass protruding from the anus. Subsequently, in 2017, Hirji et al[6] reported another case presenting as rectal prolapse with a mass in a male patient, who presented at the age of 17 years. Yao et al in 2016[7] reported in a 64-year-old female patient with complaints of increased frequency of defecation and a protruding anal mass. The patient in our case report was a young female, approximately 33 years old, who presented with the chief complaint of blood per rectum. A substantial retrospective study involving 127 cases (including both nodal and extranodal diseases) conducted by Muhammed et al[8] in 2019 revealed that the median age of diagnosis was 58 years, with a higher prevalence among males (1.65:1) and a mortality rate of 36.4%. Out of 127 cases, 48.4% of tumors were primarily nodal, 31.7% primarily extranodal, and 19.8% with combined nodal and extranodal presentations. According to the Cox regression model used in the study, surgical resection was found to be the only viable option, with no additional survival benefits attributed to adjuvant chemoradiotherapy. The 1-year mortality rates for resected and nonresected diseases were significantly different at 17.8 and 63.2%, respectively ($p < 0.0001$).

When considering nodal cases, the head and neck (HNC) region is the most common location, with neck lymphadenopathy being the most common presentation. In the HNC region, extranodal cases are frequently seen originating in Waldeyer's ring. When considering extranodal cases, the liver is the most common site. However, for GI location, the terminal ileum is more common among patients. The exact etiology remains unidentified, and the pathogenesis is still not fully understood. Due to the infrequent occurrence of IDCS, there is a lack of knowledge of demographics, epidemiology, causation, pathogenesis, clinical presentation, radiological appearance, and management protocols. Consequently, further cases and literature reviews are necessary. Radiology would be helpful in differentiating IDCS from other differentials presenting as

Table 1 Review of IDCS case in the GI tract

References	Age	Gender	Location	Gross findings	Associated tumor	HPE and IHC	Radiological	Year
Daum GS, Liepman M, Woda BA. Dendritic cell phenotype in localized malignant histiocytosis of the small intestine. Arch Pathol Laboratory Med. 1985;109:647–50.[9]	43 y	Male	Jejunum	Proliferative mass	–	Positive for S100 protein	–	1985
Miettinen M, Fletcher CD, Lasota J. True histiocytic lymphoma of small intestine. Analysis of two S-100 protein-positive cases with features of interdigitating reticulum cell sarcoma. Am J Clin Pathol. 1993 Sep;100(3):285–92. doi: 10.1093/ajcp/100.3.285. PMID: 8379537.[10]	58 y	Male	Ileum	Proliferative mass	–	Stained for the histiocytic markers lysozyme, CD68, and HLA-DR and had positive results for S-100 protein and vimentin	–	1993
Banner B, Beauchamp ML, Liepman M, Woda BA. Interdigitating reticulum-cell sarcoma of the intestine: a case report and review of the literature. Diagn Cytopathol. 1997 Sep;17 (3):216–22. doi: 10.1002/(sici) 1097-0339(199709)17:3<216:: aid-dc10>3.0.co;2-i. PMID: 9285196.[11]	68 y	Female	Cecum	Proliferative mass	Prior history of diffuse large B-cell lymphoma	Positive for S100, HLA-DR, CD68, CD74, and CD45 and negative for CD3 (T cells), CD20 (B-cells), and α-1-antichymotrypsin	–	1997
Kanaan H, Al-Maghrabi J, Linjawi A, Al-Abbassi A, Dandan A, Haider AR. Interdigitating dendritic cell sarcoma of the duodenum with rapidly fatal course: a case report and review of the literature. Arch Pathol Laboratory Med. 2006 Feb;130 (2):205–8. doi: 10.5858/2006-130-205-IDCSOT. PMID: 16454564.[4]	36 y	Female	Duodenum	Ulcerative mass	Involving adjacent liver and peri-pancreatic lymph nodes	The tumor cells consistently expressed S100 protein and vimentin in the duodenum, liver, and lymph nodes. CD45 and CD68 were weakly positive. Smooth muscle actin was positive only in the duodenal lesion and negative in the liver and lymph nodes. Electron microscopy showed no evidence of Birbeck granules or lysosomes.	–	2006

Table 1 (*Continued*)

References	Age	Gender	Location	Gross findings	Associated tumor	HPE and IHC	Radiological	Year
Shibutani M, Teraoka H, Nakao S, et al. A case of interdigitating dendritic cell sarcoma of the rectum. The Jpn J Gastroenterol Surg 2010;43:857–62. 10.5833/jjgs.43.857.[5]	76 y	Male	Rectum	Polyp	–		–	2010
Nonaka K, Honda Y, Gushima R, Maki Y, Sakurai K, Iyama K, Sasaki Y. Narrow-band imaging of interdigitating dendritic cell sarcoma originating in the duodenum. Endoscopy. 2011;43 Suppl 2 UCTN:E113–4. doi: 10.1055/s-0030-1256146. Epub 2011 Mar 18. PMID: 21425001.[12]	50 y	Male	Ileum	Saucer-like elevated lesions	–	Oval- to spindle-shaped atypical cells, positivity with S100 protein and fascin. However, CD1a and CD21 were immuno-negative.	–	2011
Ishihara S, Honda Y, Asato T, Nonaka M, Nakagawa S, Hirashima K, Hayashi N, Baba H, Iyama K. Interdigitating dendritic cell sarcoma of the ileum recurred in multiple lymph nodes and duodenum three years after operation without chemotherapy. Pathol Res Pract. 2010 Jul 15;206 (7):514–8. doi: 10.1016/j. prp.2010.01.009. Epub 2010 Apr 15. PMID: 20399026.[13]	47 y	Male	Ileum; recurrence in the duodenum	Ulceration with mass	–	Oval to spindle-shaped atypical cells. Positivity of S100 protein.	CT suggestive of intussusception	2010
Yao ZG, Wang YK, Qin YJ, Zhao MQ, Li X. A rare case of rectal interdigitating dendritic cell sarcoma presenting with a polypoid appearance. Pathology. 2016 Dec 1;48(7):744–6.[7]	64 y	Female	Rectum	Polyp		IHC studies showed tumor cells were diffusely positive for vimentin, S100 protein, fascin, CD163, and Sox10, but uniformly negative for B- and T-cell markers, CD21, CD23, CD117, CD1a, langerin, Dog-1, CD34, SMA, MyoD1, myogenin, HMB-45, Melan A, lysozyme, and BRAFV600E. Index of K_i-67 proliferation rate was 50%.	No prior imaging done	2016

(*Continued*)

Table 1 (*Continued*)

References	Age	Gender	Location	Gross findings	Associated tumor	HPE and IHC	Radiological	Year
Hirji SA, Senturk JC, Hornick J, Sonoda T, Bleday R. A rare case of interdigitating dendritic cell sarcoma of the rectum: review of histopathology and management strategy. BMJ Case Rep. 2017 Aug 7;2017: bcr2017221754. doi: 10.1136/bcr-2017-221754. PMID: 28784918; PMCID: PMC5747786.[6]	17 y	Male	Rectum	Polyp	–	Sheets of relatively uniform spindle cells with numerous admixed lymphocytes. On IHC the tumor cells are strongly positive for S100 protein	CECT abdomen suggested a heterogeneous enhancing rectal polypoidal mass	2017
Karthikeyan R, Sakthivel H, Rajkumar N, Srinivasan K. Interdigitating dendritic cell sarcoma of the small intestine presenting as spontaneous hemoperitoneum - a rare case report. Niger J Surg. 2021 Jan-Jun;27(1):71–74. doi: 10.4103/njs.NJS_55_19. Epub 2021 Mar 9. PMID: 34012247; PMCID: PMC8112361.[14]	61 y	Male	Ileum		–			2019
Present case	33 y	Female	Rectum	Polyp	–	Spindle tumor cells were strongly positive for EMA, S100, SOX10, TLE (moderate), and CD68 (speckled). BRAFV600E mutant protein was not present. The tumor cells were also negative for CK, HMB45, Melan A, NUT, SMA, SMMH, CD10, ER, stat-6, CD34, D21/23, CXCL13. INI-1 and H3k27ME3 were retained and K_i-67 was ∼2%	MRI: well-defined pedunculated polyp (T2W-intermediate hyperintense and shows diffusion restriction)	2023

Abbreviations: CT, computed tomography; HPE, histopathological examination; IDCS, interdigitating dendritic cell sarcoma; IHC, immunohistochemistry; MRI, magnetic resonance imaging.

rectal polypoidal lesions, such as melanoma, GI stromal tumor, leiomyoma, and neuroendocrine tumors, and this correlation would further help pathologists and surgeons in early diagnosis, where a high index of suspicion persists.

After conducting a detailed review of the literature focusing on IDCS in the GI tract, it has been found that a total of 10 cases of extranodal GI IDCS have been documented thus far (►**Table 1**). The most frequently affected area was the ileum, particularly the terminal portion, likely due to the higher concentration of Peyer's patches in that region of the intestine. Only three cases of IDCS in the rectum have been previously reported. All four cases, including the present case, presented as rectal polypoidal lesions with bleeding per rectum and mild anemia. Two patients were male and two were female. Two cases were presented at a young age (17 and 33 years), while two were in old age (64 and 76 years). In this study, the fourth case of IDCS in the rectum in a young female patient was presented with a detailed discussion on radiological imaging appearance and findings on MRI and PET/CT. In the reviewed literature, there were limited findings and appearance of radiological modalities.

Due to the rarity of this disease, there is no specific imaging appearance that has been identified. It can be challenging to diagnose histopathologically, as it may be mistaken for neurogenic or other spindle cell tumors; thus, immunohistopathology is crucial.

IDCS expresses CD68 (variable, 50%), LYS (variable, 25%), S100 (strongly and diffusely, 100%), and vimentin, and is negative for CD1a, CD21/35, Desmin, and SMA—markers for follicular DC origin tumors.[1–3] There is no established standard treatment plan for treating IDCS due to its infrequency. Surgery is typically the primary approach. The risk of local recurrence is associated with factors such as the size, location, and margins of surgical excision. While there is no standardized follow-up protocol, it is generally recommended to monitor for local recurrence and pulmonary metastases through CT imaging or chest X-ray.

Strengths

This report highlights a rare rectal IDCS case with comprehensive MRI, PET/CT, and histopathologic correlation. The use of an extensive immunohistochemical panel enhances diagnostic accuracy. It adds valuable insight to the scarce literature on extranodal DC tumors.

Limitations

Being a single-case report with short follow-up and limited molecular testing, generalization is restricted.

Future Directions

Collaborative multicenter studies with genomic profiling and longer follow-up are needed to better define imaging characteristics, biological behavior, and optimal management strategies.

Conclusion

IDCS of the rectum is an extremely rare neoplasm. Accurate diagnosis depends on a combination of imaging and immunohistochemical analysis. Surgery is the cornerstone of treatment, and regular follow-up is crucial due to the potential risk of recurrence or metastasis. This case adds valuable clinical and radiological data to the limited literature on rectal IDCS.

Ethical Approval

Approved by the Institutional Review Board; informed consent requirement waived due to retrospective nature.

Funding

None.

Conflict of Interest

None declared.

Acknowledgments

The authors express their gratitude to the Departments of Radiology, Pathology, and Surgical Oncology for their support in the diagnosis and management of the case. All authors have contributed substantially to the preparation of this manuscript, reviewed it critically, and approved the final version.

References

1 Wright-Browne V, McClain KL, Talpaz M, Ordonez N, Estrov Z. Physiology and pathophysiology of dendritic cells. Hum Pathol 1997;28(05):563–579
2 Pileri SA, Grogan TM, Harris NL, et al. Tumours of histiocytes and accessory dendritic cells: an immunohistochemical approach to classification from the International Lymphoma Study Group based on 61 cases. Histopathology 2002;41(01):1–29
3 Rouillé E, Bilbault H, Levin C, et al. Interdigitating dendritic cell hyperplasia in a lymph node of a control C57BL/6 mouse. J Toxicol Pathol 2021;34(01):101–106
4 Kanaan H, Al-Maghrabi J, Linjawi A, Al-Abbassi A, Dandan A, Haider AR. Interdigitating dendritic cell sarcoma of the duodenum with rapidly fatal course: a case report and review of the literature. Arch Pathol Lab Med 2006;130(02):205–208
5 Shibutani M, Teraoka H, Nakao S, et al. A case of interdigitating dendritic cell sarcoma of the rectum. Jpn J Gastroenterol Surg 2010;43:857–862
6 Hirji SA, Senturk JC, Hornick J, Sonoda T, Bleday R. A rare case of interdigitating dendritic cell sarcoma of the rectum: review of histopathology and management strategy. BMJ Case Rep 2017; 2017:bcr2017221754
7 Yao ZG, Wang YK, Qin YJ, et al. Rectal interdigitating dendritic cell sarcoma presenting as a polypoid lesion. Pathology 2016;48(07): 744–746
8 Muhammed A, Ahmed ARH, Maysa H, et al. New insights into interdigitating dendritic cell sarcoma: pooled analysis and literature review. Ann Hematol 2019;98:2641–2651
9 Daum GS, Liepman M, Woda BA. Dendritic cell phenotype in localized malignant histiocytosis of the small intestine. Arch Pathol Lab Med 1985;109(07):647–650
10 Miettinen M, Fletcher CD, Lasota J. True histiocytic lymphoma of small intestine. Analysis of two S-100 protein-positive cases with features of interdigitating reticulum cell sarcoma. Am J Clin Pathol 1993;100(03):285–292

11 Banner B, Beauchamp ML, Liepman M, Woda BA. Interdigitating reticulum-cell sarcoma of the intestine: a case report and review of the literature. Diagn Cytopathol 1997;17(03):216–222
12 Nonaka K, Honda Y, Gushima R, et al. Narrow-band imaging of interdigitating dendritic cell sarcoma originating in the duodenum. Endoscopy 2011;43(Suppl 2 UCTN):E113–E114
13 Ishihara S, Honda Y, Asato T, et al. Interdigitating dendritic cell sarcoma of the ileum with recurrence in lymph nodes and duodenum post-surgery. Pathol Res Pract 2010;206(07):514–518
14 Karthikeyan R, Sakthivel H, Rajkumar N, et al. Interdigitating dendritic cell sarcoma of the small intestine with spontaneous hemoperitoneum. Niger J Surg 2021;27(01):71–74

