CASE REPORT

A Rare Case of Interdigitating Dendritic Cell Sarcoma of the Rectum: Review of Histopathology and Management Strategy

Sameer A Hirji,¹ James C Senturk,¹ Jason Hornick,² Toyooki Sonoda,³ Ronald Bleday¹

¹Department of Surgery, Brigham and Women's Hospital, Harvard Medical School, Boston, Massachusetts, USA

²Department of Pathology, Brigham and Women's Hospital, Harvard Medical School, Boston, Massachusetts, USA

³Department of Surgery, New York Presbyterian/Weill Cornell Medical Center, New York, USA

Correspondence to

Sameer A Hirji,
shirji@partners.org

Accepted 25 July 2017

SUMMARY

Interdigitating dendritic cell sarcoma (IDCS) is a rare neoplasm arising from a subclass of dendritic cells, known for their role in mediating various immunological functions, including T-cell mediated immunity. Although existing literature on IDCS is limited to scattered reports, extranodal manifestation in the gastrointestinal tract, and in particular, the rectum is extremely rare. To our knowledge, we report only the second case of IDCS arising in the rectum in a young 20-year-old man, successfully managed surgically and with a good oncological outcome. Existing literature on the incidence, pathophysiology and treatment strategies is also examined.

BACKGROUND

Interdigitating dendritic cell sarcoma (IDCS) is a rare neoplasm arising from a subclass of dendritic cells, which are well known for their role in mediating various diverse immunological functions, including initiation of T-cell-mediated immunity.^{1 2} While extranodal manifestation of IDCS is uncommon, the disease has been reported in the nasopharynx, skin, chest wall, bladder and abdominal viscera. IDCS arising in the gastrointestinal tract (GI) itself is extremely rare.^{3–5} Existing literature on IDCS in the GI tract is limited to only a few case reports, with only one reported case of IDCS of the rectum. Given the paucity of available data with regards to GI, and in particular, rectal IDCS demographics, presentation and clinical behaviours, evidence-based recommendations for early diagnosis and treatment of this disease process are lacking. We present a unique case of IDCS of the rectum in a 20-year-old man, diagnosed incidentally and successfully managed surgically with no recurrence. We will also review the current literature to examine the incidence, pathophysiology of various subtypes of dendritic cell sarcomas as well as management strategies.

CASE PRESENTATION

A 20-year-old man with an extensive history of rectal prolapse, diagnosed at the age of 17, initially presented to an outside hospital with a 2-year history of mild anaemia, frequent bowel movements and a 20-pound unintentional weight loss. During his workup, he developed acute appendicitis, which required surgical management with laparoscopic appendectomy. However, CT scan of the abdomen

and pelvis during his preoperative evaluation was also notable for an incidental rectal mass (figure 1). He subsequently underwent a flexible sigmoidoscopy, revealing a rectal mass 7cm from the anal verge that was non-diagnostic on biopsies. Given concerns for a possible malignancy in the setting of his recent weight loss and new symptoms of intermittent episodes of rectal bleeding, he underwent a transanal excision of the 8×8×6 cm posterior rectal mass. The mass was located on a broad stalk that measured approximately 3×3 cm in diameter (figure 2). Final pathology was consistent with IDCS (Stage III: G2, T2b, N0, M0) infiltrating the rectal wall and focally extending into mesorectum with an associated positive resection margin.

TREATMENT

Due to the nature of the pathology and location in the rectum, he presented to our institution for a second opinion. Following a multidisciplinary

Figure 1 CT scan images of the abdomen and pelvis with rectal mass.

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To cite: Hirji SA, Senturk JC, Hornick J, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-221754

Figure 2 Gross image showing relative size of the rectal mass.

evaluation resection was recommended, although preoperative radiation therapy was considered. He underwent a two-stage procedure, first involving a low anterior resection with hand-sewn side-to-end coloanal anastomosis and temporary diverting ileostomy followed by a take down of the ileostomy 2 months later. The coloanal anastomosis was handsewn because the scar from the previous transanal excision was about 1–2 cm above the dentate line. The surgery was uneventful and he recovered well without complications. His final pathology showed no residual tumour and negative mesorectal lymph nodes. With little data in the literature to demonstrate the utility of adjuvant treatments and because of clear surgical margins, postoperative adjuvant radiation or chemotherapy was not recommended. It was felt that there was minimal potential benefit and significant potential side effects of radiation and chemotherapy in a young otherwise healthy patient. Surveillance with physical exam along with CT scans was recommended.

Figure 3 (Top): Histology of interdigitating dendritic cell sarcoma. Sheets of relatively uniform spindle cells with numerous admixed lymphocytes. (Bottom): By immunohistochemistry, the tumour cells are strongly positive for S100 protein.

OUTCOME AND FOLLOW-UP

During his follow-up clinic visits over the last 18 months, he has been doing well, having regular bowel movements and appropriate weight gain. His repeat CT scans have been unremarkable with stable findings and no evidence of locally recurrent or distant disease.

DISCUSSION

IDCS is a very rare neoplasm arising from a subclass of dendritic cells, known for their role in mediating various immunological functions, including T-cell-mediated immunity.^{1 2} Despite its name, IDCS is a tumour of the haematopoietic system (bone marrow derived from the myeloid lineage), although its morphological appearance can mimic that of soft tissue sarcomas. This is the first extranodal IDCS treated at our institution. To our knowledge, this is only the second reported case of IDCS arising in the rectum that was successfully managed surgically. The localised nature of the tumour and lack of pathological evidence of lympho-vascular invasion or lymph node involvement led to our recommendation for surgery only for his treatment.

Fewer than 60 case reports of extranodal IDCS exist in the literature, with observed incidences including the nasopharynx, skin, chest wall, bladder and less commonly in the abdominal viscera. In fact, IDCS arising in the GI tract is extremely rare and limited to only a handful of case reports since 1985, including only one

previous reported case in the rectum. The majority of the GI cases underwent resection followed by adjuvant chemotherapy with mixed outcomes. For instance, Banner *et al* described a case of IDCS involving the caecum in a 68-year-old woman with a 5-year prior history of diffuse large B-cell lymphoma which had been treated with chemoradiation and felt to be in complete remission. For the IDCS, she underwent a right hemicolectomy but had disease recurrence (unknown location) at 3 months, necessitating additional adjuvant chemotherapy. She was alive with disease at 3 months although additional follow-up information was not provided.⁶ Ishihara *et al* documented a case of a 47-year-old male who presented with intussusceptions in the setting of two submucosal lesions consistent with IDCS of the ileum. The patient subsequently underwent an ileocolic resection without chemotherapy. He unfortunately had disease recurrence at 3 years in the duodenum with concurrent pelvic and abdominal lymphadenopathy necessitating chemotherapy. He was alive with disease at 8-month follow-up.⁵ A similar case of IDCS in the terminal ileum was reported by Miettinen *et al* with the patient being disease free at 12 months.⁷ Additionally, there were two other patients with IDCS of the jejunum and small intestine, respectively, who also underwent resection and adjuvant chemotherapy but who died of the disease.^{2,7} Patients undergoing only resection also had mixed outcomes. Kanaan *et al* presented a 36-year-old female who presented with IDCS involving the duodenum, with liver and peripancreatic lymph node involvement. She underwent a Whipple procedure due to the adjacent pancreatic head mass without adjuvant chemotherapy. Postoperatively, she deteriorated and died after 4 months.⁴ Having a similar treatment strategy, Nonaka *et al* presented a case of a 50-year-old man with a known diagnosis of IDCS of the terminal ileum who presented with multifocal lymphadenopathy and recurrence in the duodenum 3 years after an ileocolic resection without adjuvant chemotherapy. His prognosis was unknown.⁸

In our review of the literature, there is only one other reported case of IDCS of the rectum. The patient was a 76-year-old man who presented initially with haematochezia and a palpable mass 2 cm from the anal verge. His workup involved a colonoscopy that demonstrated a semicircular tumour with a central ulcer in the lower rectum. He subsequently underwent an abdomino-perineal resection but required adjuvant chemotherapy in the setting of positive regional and para-aortic lymphadenopathy, with overall interval improvement in the size of para-aortic nodes on follow-up axial tomography at 8 months.⁹

IDCS is typically composed of sheets of usually spindled but occasionally epithelioid cells with eosinophilic cytoplasm; when involving lymph nodes, IDCS shows a paracortical growth pattern. By immunohistochemistry, IDCS shows strong and diffuse expression of S100 protein; expression of CD45, CD4 and CD43 are variable, as is expression of histiocytic markers such as CD68 and CD163. The case reported herein expressed S100 protein, CD34, CD4, CD45, CD163, CD14, CD43 and CD68 but was negative for KIT, ALK, CD21, CD23, desmin and CD1a (figure 3). Likewise, in the previous case of the 76-year-old man, tumour cells expressed CD68, CD99, S100 protein and vimentin but were negative for CD1a, CD3, CD79a and KIT.^{7,9} Unfortunately, IDCS is often misdiagnosed as lymphoma, metastatic melanoma or various types of soft tissue sarcoma, which can delay treatment.¹⁰

In contrast, follicular dendritic cell sarcoma (FDCS) is relatively more common, with a slight female predominance. Moreover, FDCS are mesenchymal in origin, may arise in lymph nodes, mainly the axillary or cervical or at extranodal sites (similar to IDCS) including the GI tract, the stomach, liver, small bowel and rectum.¹¹

For instance, Shia *et al* reported a series of 46 patients with FDCS in which eight patients (17%) had lymph node involvement at the time of presentation. Treatment mostly involved surgery (41%) with adjuvant radiation (eight patients) or chemotherapy (three patients) or both (one patient). There was a 43% recurrence rate and 7% mortality rate over a median follow-up of 18 months, with a 5-year recurrence-free survival of 27.4%.¹¹ Although the pathogenesis of FDCS appears heterogeneous, unlike IDCS, some cases are associated with Epstein-Barr virus infection, mostly in the liver and spleen.¹¹ Clinically, definitive diagnosis is achieved through biopsies, although this can be challenging and discrepant at times as these tumours tend to mimic other tumour types. For instance, in a review of 46 cases of extranodal FDCS, around one-third of the cases were misdiagnosed at initial evaluation.¹¹ In contrast to IDCS, FDCS often shows a whorled growth pattern and expresses CD21, CD23, CD35 and clusterin, among other specific markers. While surgical management remains the mainstay treatment option, most patients require adjuvant treatment of some sort given the relatively high recurrence rate, although there is not yet a standard of care.¹¹

Treatment recommendation for IDCS of the GI tract can be varied given limited evidence-based studies.^{10,12} However, with localised disease, the literature and our experience suggest that surgery is the best treatment with a goal for a RO resection. The utility of adjuvant radiotherapy or chemotherapy in combination with complete surgical resection remains unclear with mixed outcomes based on previous reported cases. In patients with metastatic IDCS, chemotherapy is reasonable since the disease is of haematopoietic origin.¹⁰

Overall mortality for IDCS and FDCS has been reported in a pooled analysis from the Surveillance, Epidemiology and End Results database. Twenty patients with IDCS were compared with 54 patients with FDCS. FDCS had a better survival in patients with localised disease compared with IDCS patients. Two-year survival rates in IDCS patients with localised and disseminated disease were 68.1% and 15.8%, respectively.¹²

Through our case report, we present a review of existing literature on IDCS with a focus on the GI tract. Evidence-based recommendations for early diagnosis and treatment of gastrointestinal IDCS are lacking. Also, in such a rare disease, prospective studies are not possible, and case reports such as these continue to provide a framework to guide treatment decisions. Based on our experience, patients with IDCS of the rectum are best managed with resection for local/regional disease and with possible need for adjuvant therapies in cases with metastasis, lympho-vascular invasion or positive margins.

Learning points

- ▶ Interdigitating dendritic cell sarcoma (IDCS) is a very rare neoplasm arising from a subclass of dendritic cells.
- ▶ IDCS is a tumour of the haematopoietic system (bone marrow derived from the myeloid lineage), although its morphological appearance can mimic that of soft tissue sarcomas.
- ▶ Extranodal IDCS can exist but this is the second reported case of IDCS arising in the rectum.
- ▶ Patients with IDCS of the rectum are best managed with resection for local/regional disease.
- ▶ Adjuvant therapies are needed in cases of metastasis, lymphovascular invasion or positive margins.

Acknowledgements All authors contributed equally to the manuscript.

Contributors All authors have contributed significantly and equally to the manuscript. SH, JS and JH were involved in the drafting and obtaining images for

manuscript. RB, TS, SH were involved in extensive editing, review of literature and finalizing the draft.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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