CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 46 (2018) 50–55

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Detecting a rare composite small bowel lymphoma by Magnetic Resonance Imaging coincidentally: A case report with radiological, surgical and histopathological features

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ARTICLE INFO

Article history:
Received 31 January 2018
Received in revised form 3 April 2018
Accepted 8 April 2018
Available online 16 April 2018

Keywords:
Composite lymphoma
Follicular lymphoma
Magnetic Resonance Imaging
Lymphoma radiological features
Lymphoma surgery
Case report

ABSTRACT

INTRODUCTION: Diagnosing lymphoma continues to prove challenging in the clinical practice. Composite lymphoma (CL) is defined by the coexistence of different lymphoma subtypes in the same anatomical location. This condition has seldom been witnessed in the gastrointestinal (GI) tract. We weren't able to find previous cases in the literature about small bowel CL with follicular lymphoma (FL) and classical Hodgkin lymphoma (CHL). Surgery is the treatment of choice to obtain accurate histology, to manage and prevent acute complications. We state that this work has been reported in line with the SCARE criteria.
CASE PRESENTATION: We describe an extremely rare case of small bowel CL, presenting as an intestinal bulky mass with circumferential infiltration of bowel loops. The small bowel tumor was incidentally detected by abdominal Magnetic Resonance Imaging (MRI) in a 64-year-old man who suffered from rectal discomfort and non-specific clinical symptoms. After this radiological finding, the patient underwent multiphase contrast computed tomography (MDCT) for initial staging and to study vascular involvement. Surgery was recommended to obtain an accurate diagnosis both due to initial symptoms of the intestinal obstruction and to avoid small bowel complications. The histopathological examination revealed a small bowel CL composed mainly of B cells FL with also CHL components.
CONCLUSION: It is important to note that involvement of the proximal ileal loops is very rare in small bowel lymphoma. MRI represents a precious diagnostic tool to evaluate the intra and extramural extent of the tumor.

1. Introduction

Small bowel (SM) tumors are relatively rare and can represent a diagnostic conundrum for both clinicians and radiologists. This is mainly due to vague symptoms and the overlapping of imaging features which may present as ghost tumors. Tardy diagnosis with poor prognosis is common [1–3]. Gastrointestinal (GI) lymphoma accounts for about 1–4% of GI malignancies [2]. Almost all the primary GI lymphomas are of B cell lineage with very few T-cell lymphomas and classical Hodgkin lymphoma (CHL). Its most frequent sites of occurrence are the stomach followed by the small bowel and the ileocecal region [2,4,5]. The coexistence of classical (CHL) and non-Hodgkin lymphoma (NH-L) in the same anatomic location is a very unusual condition known as composite lymphoma (CL). Local coexistence of a HL and NH-L in the GI tract has been reported very rarely [6–8]. We present an extremely rare case of small bowel lymphoma composed by follicular lymphoma (FL) and CHL. This case sheds light the heterogeneous variety in terms of clinical presentation, radiological features and histological subsets of GI lymphoma. We state that this work has been reported in line with the SCARE criteria [9].

2. Case presentation

A Caucasian 64-year-old man was admitted to the Gastroenterology Department of our Institution for rectal discomfort and a mild epigastric pain, which started two months earlier. He also reported a stabbing perianal pain. He had only graduated from primary school and suffered from clinical depression.

Abbreviations: ADC, apparent diffusion coefficient; CL, composite lymphoma; CHL, classical Hodgkin lymphoma; CT, computed tomography; DWI, diffusion weighted imaging; FL, follicularlymphoma; FSE, fast spin eco; GI, gastrointestinal; HB, haemoglobin; MALT, mucosa-associated lymphoid tissue; MCV, mean cells volume; MRI, Magnetic Resonance Imaging; NH-L, Non-Hodgkin Lymphoma; True-Fisp, true fast imaging steady state precession; w, weighted.

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https://doi.org/10.1016/j.ijscr.2018.04.005

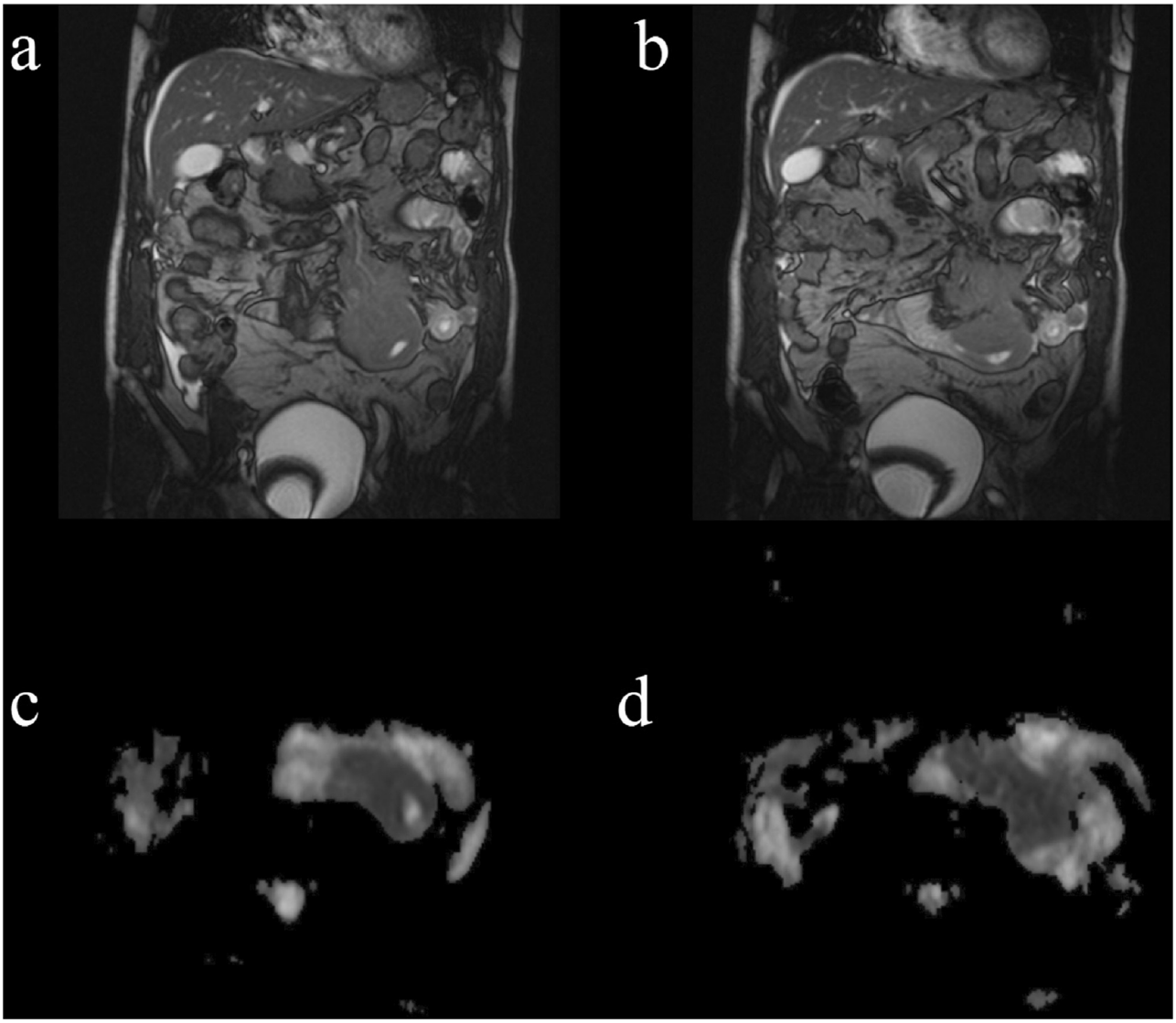

Fig. 1. The bulky mass on True-FISP characterized by mesenteric pattern (a) with circumferential extension of small bowel loop (b). Diffuse low apparent diffusion coefficients (d,c).

He had been operated two years before for haemorrhoids by means of a Longo hemorrhoidopexy. An esophagogastroduodenoscopy and colonoscopy had already been performed at another Institution, which could not find any tumor. The esophagogastroduodenoscopy described only a mild gastritis. Routine blood examination showed normal laboratory values with hemoglobin (Hb) 12.8 g/dL, mean cell volume (MCV) 85.8FL, normal white blood cell counts 6400/mm^3 with normal platelet counts 172,000/mm^3. The lactate dehydrogenase level was 130 IU/L in the normal range, β2-microglobulin was mildly elevated. Other laboratory values, including electrolytes, creatinine, and liver enzymes, were normal. Serological testing showed negativity for hepatitis B virus and human immunodeficiency viral infections. Carcinoembryogenic antigen, α-fetoprotein and carbohydrate antigen were in the normal range. An upper abdominal ultrasound showed only liver steatosis and a double renal district on the left side.

Therefore, clinicians requested an MRI due to the patient complaining about rectal discomfort and perianal pain and in order to rule out with certainly any perianal pathologies and also to exclude a pudendal nerve entrapment. The study was performed by oblique axial T1-weighted (w)Fast-Spin-eco (FSE), oblique axial T2w FSE, oblique axial and oblique coronal fat-suppressed T1w FSE, completed by gadolinium-based contrast material. This study did not show perianal pathologies but found a consistent amount of peritoneal ascites. Therefore, as the non-specific clinical symptoms, it was decided to extend the study to all abdomen. Coronal and axial True fast imaging steady state precession (True- FISP), axial Diffusion weighted imaging (DWI), axial and coronal fat suppressed T1w images after gadolinium-based contrast material were also carried out. These latter sequences revealed a large bulky mass in the mesenteric region, with thickening and mild dilatation of the involved small bowel walls. It displayed hypointense signal on T2w, with low apparent diffusion coefficient (ADC) (mean ADC value 0.6591×10^{-3} mm^2/s) (Fig. 1) and homogeneous enhancement (Fig. 2). Small bowel lymphoma was given as first diagnosis, adenocarcinoma and carcinoid tumors as other differential diagnosis. A total body computed tomography (CT) was required for initial staging, which confirmed the abnormal mass with a medium size of $9.71 \times 8.64.12.62$ cm involving also superior mesenteric artery (Fig. 3), thought still patent. No lung, brain and bone metastasis were observed.

Twelve days after the staging CT, the patient showed symptoms of intestinal obstruction, which led to the patient's hospitalization for supportive care without the need of surgery at first because the

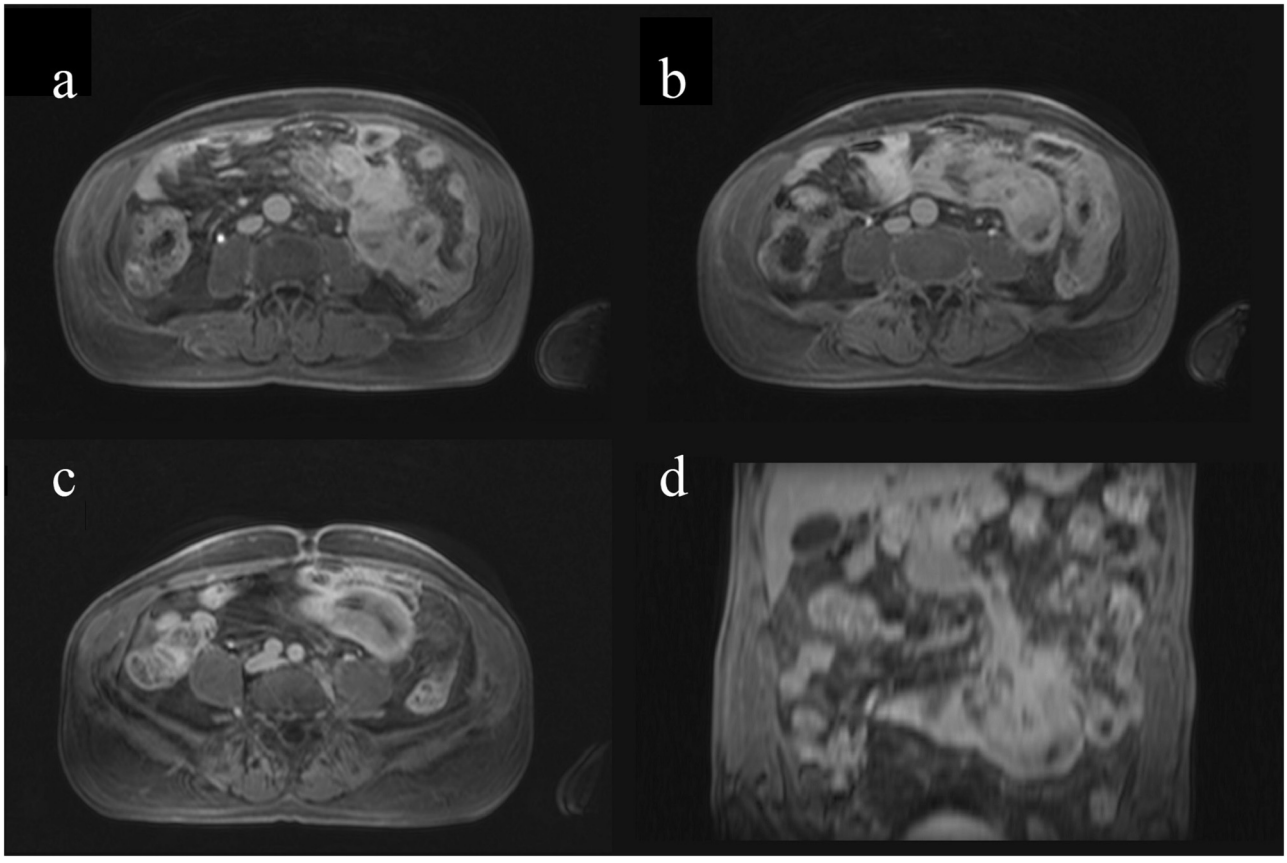

Fig. 2. MRI examination after contrast administration. The bulky mass showed few hypointense areas (a), though the enhancement was homogeneous (b,c) with focal widening of the interested small bowel loop; (d) coronal reconstruction.

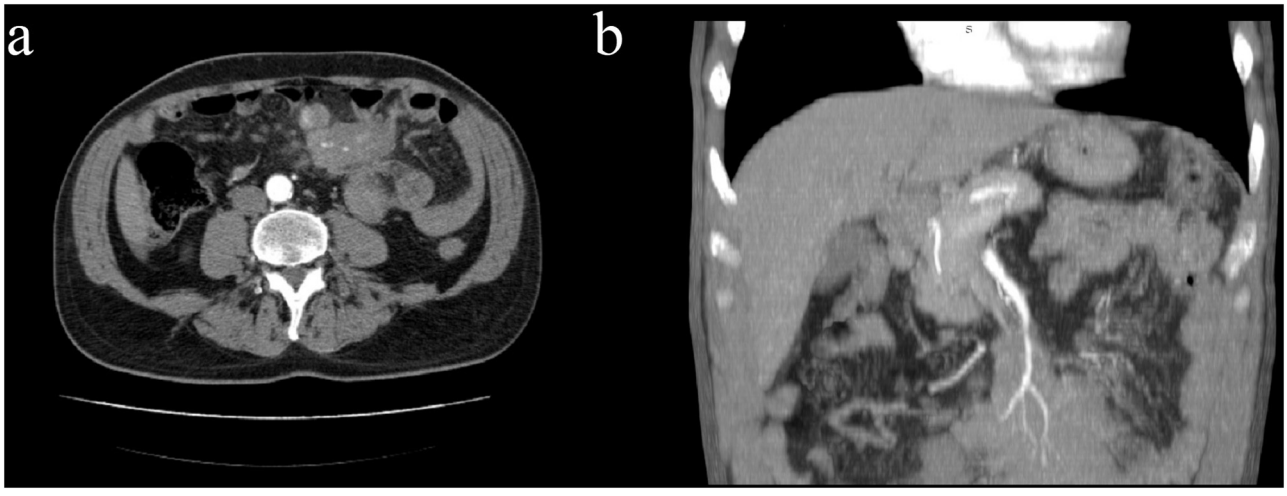

Fig. 3. various mesenteric lymphadenopathy and mass enveloping of secondary branch mesenteric vessels(a); superior mesenteric artery involvement, though with patent appearance(b).

obstruction had dissolved on its own. However, in order to establish a correct histological diagnosis and to prevent further intestinal obstructions, surgery was, in our opinion, necessary.

We started with a laparoscopy but switched to an exploratory laparotomy due to the lack of mobility of the intestinal segment and the size of the tumor. A 10-cm sclerotic and whitish plaque incorporated the first ileal loops and tended to extend to the mesentery (Fig. 4a). This had led to a retraction of some adjacent loops (Fig. 4b). Therefore, an approximately 75-cm ileal resection was performed, including the adjacent mesentery and the mesenteric lymph nodes. It was not possible to carry out a complete resection of the tumor in order to avoid a massive necrosis of the small intestine, so a residual pathology can still be found along the superior mesenteric artery wall. The intestinal continuity was ensured by a mechanical latero-lateral entero-enteric anastomosis and a perianastomotic drainage was positioned (Fig. 4c).

The patient was discharged on the sixth post-operative day and the subsequent course was characterized by a dehiscence of the abdominal wound which was treated through second intention over the course of a month. A large amount of tissue from this

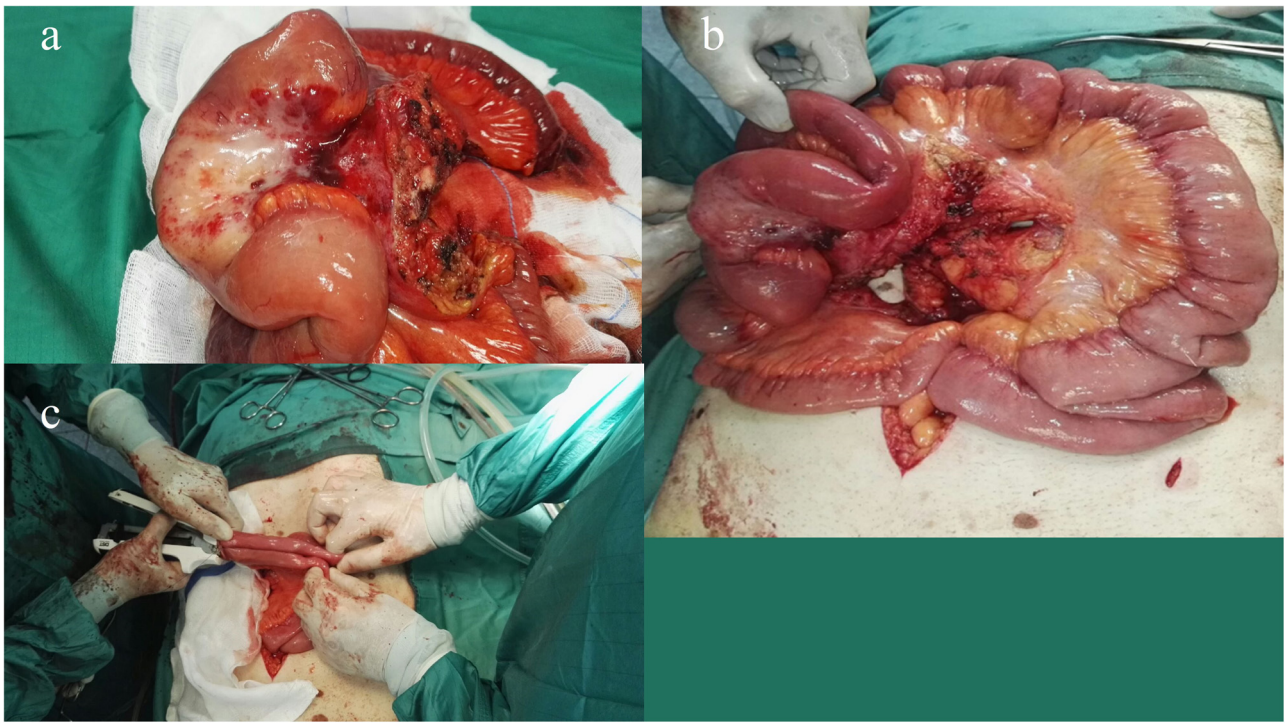

Fig. 4. A 10-cm sclerotic and whitish plaque (Fig. 4a) incorporated the first ileal loops and tended to develop towards the mesentery (a); ileal resection extended for approximately 75 cm, including adjacent mesentery and the mesenteric lymph nodes (b); latero-lateral entero-enteric anastomosis (c).

tumor was submitted for microscopic evaluation. At histopathology examination, the tumor displayed two distinct morphological components. The larger of these was characterized by centroblasts cells and neoplastic lymphoid follicles in the mucosa, submucosa with nodular appearance, resulting in a low grade (G1-G2) B-cells FL with cluster of differentiation BCL2+,CD20+,BCL6+/−, CD3-, CD15-, CD30-,EBER-,cyclinD1-,CD38-,CD168-. Furthemore, the other area (about 10%) presented occasional cells morphologically compatible with mononuclear Hodgkin cells with immunochemistry BCL2+,CD30+,MUM+,CD20+/−,PAX5+/− and CD10-,BCL6-,EBER-,CD138-,cyclin D1- and elevated proliferation rate index (Ki67**).** No secondary localizations were found on the mesenteric lymphonodes. No evidence of bone marrow involvement was found. After 6 months of chemotherapy the patient showed no signs of systemic dissemination.

3. Discussion

Follicular lymphoma (FL) accounts for 1–3.6% of GI-NH-L and it is usually a low-grade lymphoma which develops very slowly [10]. It represents a subtype of indolent NH-L, therefore patients may be frequently asymptomatic. However clinical symptoms are related to bowel thickening and bowel obstruction. This neoplasm originates in germinal center B cells and is characterized by centrocytes and centroblasts forming follicular patterns of various size [10–13]. The age range of the diagnosis is from 26 to 81 years with a predominance of females. GI-FL more frequently involves the duodenum, around the ampulla of Vater, the terminal ileum, and the ileocecal region FL [10]. It usually appears as a white-tone polyposis with unifocal or multifocal distributions. Such large mass as our case is less common [10–14]. Although GI lymphoma histology can be varied. CL with GI primary development represents less than 5% of GI lymphomas [6]. This rare condition may explain the potential genetic relationship between two phenotypically different lymphomas, suggesting a common precursor. The literature reports about 21 cases of GI diffuse large B cell lymphoma with CHL [6], but we didn't find previous cases of small-bowel lymphoma with FL and CHL.Many tools are available to diagnose lymphoma [13–18], including MRI that, in our case, helped us to diagnose this malignancy. This feature highlighted the crucial importance of radiological techniques in the investigation and work up of intestinal tumors [13,15–19]. Five different patterns CT/MRI of GI lymphomas have been reported in the literature: polypoid/nodular pattern, infiltrative pattern, aneurismal pattern, exotic mass and stenosing mass [16–18]. The polypoid pattern usually causes bowel intussusceptions and shows solid nodules with homogeneous signal density/intensity developing as polypoid mass. It is more frequently seen in GI FL and presents no wall thickening. The infiltrative form is characterized by circumferentially infiltrating lymphoma and involves a variable length of small bowel with thickening and later effacement of folds. The aneurysmal pattern usually coexists with the infiltrative form and it can represent its evolution. Esofitic mass growth or mesenteric pattern is characterized by a single large mass formation (bulky appearance), usually more than 5 cm. Mesenteric small bowel involvement and lymphoid tissue outside of the intestinal wall through the adventitia, are typical features. This finding is more typical of Burkitt lymphoma or in advanced state disease [2,16,19,20]. In the larger masses, larger ulcerative complications, tissue necrosis, perforation, and enteroenteric fistula formation could be common. A stenosing form is a rare pattern of small bowel lymphoma growth with concentric fibrotic stenosis of intestinal loops and it typically occurred in CHL. In our case, the radiological features corresponded to the mesenteric pattern with circumferential tumor extention and a focal widening of the involved small-bowel loop due to the tumor taking the place of the muscularis propria [21]. The tumor showed hypointense signal in T2w, restricted diffusivity and low values on ADC maps and homogeneous enhancement. DWI represents a precious sequence and may help in detection of GI-L [16]. However, ADC values for rare GI lymphomas have not been studied. Over the years the role of surgery in the treatment of small bowel lymphomas has gradually decreased in importance,

more often giving way to chemotherapy, radiotherapy and biological therapy [20,22]. Nowadays, surgery can be an option in the early stage of mucosa-associated lymphoid tissue (MALT) bowel lymphomas and attain the same effectiveness as radiotherapy. In locally advanced lymphomas of the small bowel, surgical resection is only indicated during laparotomy or laparoscopy to define the accurate histology of the neoplasia or to manage complications such as intestinal occlusion, bleeding, and perforation [22,23]. Prevention surgery could also be advocated before chemotherapy in bulky lesions in order to prevent bowel perforation caused by rapid necrosis of the cells and to reduce the high incidence of mortality due to an emergency laparotomy. As segmental intestinal resection with its mesentery containing 12 lymph nodes is recommended.

4. Conclusion

Lymphoma histology can be multifaceted. Nowadays, MRI has been proved a highly accurate method both for early diagnosis and for local staging of the disease. Gross resection is the recommended surgical procedure for patients with small bowel lymphoma, improving the progression-free survival (PFS) [23]. Surgery can be beneficial in bulky mass, though caution should be exercised especially in patients with compromised nutritional status, immunity and poor general conditions. In these cases surgery may be accompanied by a higher risk of post operative complications (leakage of anastomosis, abscess formation, prelonged peritonitis) and early mortality. However unlike other GI malignancy, emergency surgery for complications of small bowel lymphoma does not lead to poor prognosis.

Conflict of interest

Authors have no conflict to disclose.

Funding

The research did not receive any specific grant from funding agencies in the public, commercial, or profit sectors.

Ethical approval

Ethical approval has been exempted by the institution, since it is not considered a risky investigation for the patient or for the researchers.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Pezzella M: made the surgical intervention, partecipated substantially in conception, design, and execution of the study and in the analysis and interpretation of data; also participated substantially in the drafting and editing of the manuscript. Made the surgical images in the manuscript.

Brogna B: followed the MRI examination, participated substantially in conception, design, and execution of the study and in the analysis and interpretation of data; also participated substantially in the drafting and editing of the manuscript;made the radiological images in the manuscript.

Romano A: partecipated to the surgical intervention; participated substantially in conception, design, and execution of the study.

Torelli F: partecipated to the surgical intervention; participated substantially in conception, design, and execution of the study.

Esposito G: participated to the surgical intervention; participated substantially in conception, design, and execution of the study.

Petrillo M: participated to the surgical intervention; participated substantially in conception, design, and execution of the study.

Romano F.M.: participated to the surgical intervention; participated substantially in conception, design, and execution of the study.

Reginelli A: followed the MRI examination, participated substantially in conception, design, and execution of the study and in the analysis and interpretation of data.

Di Martino N:made the surgical intervention, participated substantially in conception, design, and execution of the study and in the analysis and interpretation of data; also participated substantially in the drafting and editing of the manuscript.

Grassi R: followed the MRI examination. participated substantially in conception, design, and execution of the study and in the analysis and interpretation of data; also participated substantially in the drafting and editing of the manuscript.

Registration of research studies

The paper is not a research study.

Guarantor

For the surgical part: Professor Di Martino N, for the Radiological part Professor Grassi R.

Acknowledgement

None.

References

[1] R.M. Gore, U.K. Mehta, J.W. Berlin, V. Rao, G.M. Newmark, Diagnosis and staging of small bowel tumours, Cancer Imaging 6 (1) (2006) 209–212.
[2] P. Ghimire, G.Y. Wu, L. Zhu, Primary gastrointestinal lymphoma, World J. Gastroenterol. 17 (6) (2011) 697–707.
[3] B. Li, Y.K. Shi, X.H. He, S.M. Zou, S.Y. Zhou, M. Dong, J.L. Yang, P. Liu, L.Y. Xue, Primary non-Hodgkin lymphomas in the small and large intestine: clinicopathological characteristics and management of 40 patients, Int. J. Hematol. 87 (2008) 375–381.
[4] M.A. Bautista-Quach, C.D. Ake, M. Chen, J. Wang, Gastrointestinal lymphomas: Morphology, immunophenotype and molecular features, J. Gastrointest. Oncol. 3 (3) (2012) 209–225.
[5] S. Nakamura, M. Takayuki, Gastrointestinal lymphoma: recent advances in diagnosis and treatment, Digestion 87 (3) (2013) 182–188.
[6] G. Goyal, A.H. Nguyen, K. Kendric, G.C. Caponetti, Composite lymphoma with diffuse large B-cell lymphoma and classical Hodgkin lymphoma components: a case report and review of the literature, Pathol. Res. Pract. 212 (12) (2016) 1179–1190.
[7] Q. Huang, S.P. Wilczynski, K.L. Chang, L.M. Weiss, Composite recurrent Hodgkin lymphoma and diffuse large B-cell lymphoma: one clone, two faces, Am. J. Clin. Pathol. 126 (2) (2006) 222–229.
[8] H.W. Wang, W. Yang, L. Wang, Y.L. Lu, J.Y. Lu, Composite diffuse large B-cell lymphoma and classical Hodgkin's lymphoma of the stomach: case report and literature review, World J. Gastroenterol. 19 (37) (2013) 6304–6309.
[9] R.A. Agha, A.J. Fowler, A. Saetta, I. Barai, S. Rajmohan, D.P. Orgill, SCARE Group, The SCARE statement: consensus-based surgical case report guidelines, Int. J. Surg. 34 (2016) 180–186.
[10] S. Yamamoto, H. Nakase, K. Yamashita, M. Matsuura, M. Takada, T. Chiba, et al., Gastrointestinal follicular lymphoma: review of the literature, J. Gastroenterol. 45 (4) (2010) 370–388.
[11] J. Shia, J. Teruya-Feldstein, D. Pan, A. Hegde, D.S. Klimstra, D.A. Filippa, et al., Primary follicular lymphoma of the gastrointestinal tract: a clinical and pathologic study of 26 cases, Am. J. Surg. Pathol. 26 (2) (2002) 216–224.

[12] K. Takata, T. Miyata-Takata, Y. Sato, T. Yoshino, Pathology of follicular lymphoma, J. Clin. Exp. Hematopathol. 54 (1) (2014) 3–9.
[13] M. Iwamuro, E. Kondo, K. Takata, T. Yoshino, H. Okada, Diagnosis of follicular lymphoma of the gastrointestinal tract: a better initial diagnostic workup, World J. Gastroenterol. 22 (4) (2016) 1674–1683.
[14] C. Vetro, G. Bonanno, G. Giulietti, A. Romano, C. Conticello, F. Di Raimondo, et al., Rare gastrointestinal lymphomas: the endoscopic investigation, World J. Gastrointest. Endosc. 7 (10) (2015) 928–949.
[15] S. Ghai, J. Pattison, S. Ghai, M.E. O'Malley, K. Khalili, M. Stephens, Primary gastrointestinal lymphoma: spectrum of imaging findings with pathologic correlation, Radiographics 27 (5) (2007) 1371–1388.
[16] G. Lo Re, V. Federica, F. Midiri, D. Picone, G. La Tona, M. Midiri, et al., Radiological features of gastrointestinal lymphoma, Gastroenterol. Res. Pract. 6 (1–9) (2016) 2498143.
[17] M. Anzidei, A. Napoli, C. Zini, M.A. Kirchin, C. Catalano, R. Passariello, Malignant tumours of the small intestine: a review of histopathology, multidetector CT and MRI aspects, Br. J. Radiol. 84 (1004) (2011) 677–690.
[18] A. Faggian, M.R. Fracella, G. D'Alesio, M.E. Alabiso, D. Berritto, R. Grassi, et al., Small-bowel neoplasms: role of MRI enteroclysis, Gastroenterol. Res. Pract. (1–6) (2016) 9686815.
[19] R.B. Lewis, A.K. Mehrotra, P. Rodríguez, M.A. Manning, M.S. Levine, From the radiologic pathology archives: gastrointestinal lymphoma: radiologic and pathologic findings, Radiographics 34 (7) (2014) 1934–1953.
[20] A.D. Zelenetz, L.I. Gordon, W.G. Wierda, J.S. Abramson, R.H. Advani, L.E. Fayad, Non-Hodgkin's lymphomas, version 2.2014, J. Natl. Comp. Cancer Netw. 12 (6) (2014) 916–946.
[21] M.S. Levine, S.E. Rubesin, L. Pantongrag-Brown, J.L. Buck, H. Herlinger, Non-Hodgkin's lymphoma of the gastrointestinal tract: radiographic findings, AJR Am. J. Roentgenol. 168 (1) (1997) 165–172.
[22] R. Cirocchi, E. Farinella, S. Trastulli, D. Cavaliere, P. Covarelli, G.M. Verdecchia, et al., Surgical treatment of primitive gastro-intestinal lymphomas: a systematic review, World J. Surg. Oncol. 9 (1) (2011) 145.
[23] Y.W. Hong, I.M. Kuo, Y.Y. Liu, T.S. Yeh, The role of surgical management in primary small bowel lymphoma: a single-center experience, Eur. J. Surg. Oncol. 43 (10) (2017) 1886–1893.