CASE REPORT

Metastatic anaplastic large cell lymphoma (ALCL) presenting with small bowel perforation

Novinth Kumar Raja Ram, Siti Fareeda Anuar, Tuan Nur' Azmah Tuan Mat
Department of General Surgery,
Hospital Sultanah Aminah, Jalan Persiaran Abu Bakar Sultan, Johor, Malaysia

Keywords: ALCL; small bowel perforation; lymphoma

Introduction

Lymphoma ranks third in the incidence of small bowel malignancy, with adenocarcinoma being the commonest (1). Anaplastic large cell lymphoma (ALCL) constitutes about two percent of adult non-Hodgkin lymphoma and is a subset of T cell lymphoma histology. It has a bimodal age distribution with regard to incidence with the first peak in adolescence and the second after the sixth decade , with a male predominance. Four cases of primary small bowel ALCL presenting with perforation have previously been reported (1). However, literature regarding metastatic ALCL to the small bowel presenting with perforation is scarce. We report a case of metastatic ALCL which presented as jejunal perforation.

Case report

A 65 year old Malay gentleman presented with fever and sudden onset of abdominal pain, most pronounced over the epigastrium for two days. He had a history of ALK negative anaplastic large cell lymphoma, which was diagnosed 5 months prior to the presentation. He had completed two cycles of chemotherapy with Cyclophosphamide, Doxorubicin, Vincristine and Prednisolone (CHOP). He was later switched to Chlorambucil + Prednisolone due to intolerance. He also suffered from diabetes mellitus and hypertension. Examination revealed tenderness over the epigastric region with peritonism. Computed Tomography (CT) of the abdomen showed fluid in perihepatic, subhepatic and pelvic regions with air pockets within. Patient underwent an emergency laparotomy with a provisional diagnosis of a perforated viscous. Intra operatively, he was found to have a jejunal perforation, approximately 25 cm from duodeno-jejunal flexure with an ulcerative lesion within. There were multiple enlarged mesenteric lymph nodes, largest measuring 3x3 cm. Peritoneal cavity was grossly contaminated with succus entericus. A wedge resection with primary anastomosis and peritoneal lavage was done. Perioperative dissection showed an ulcerative lesion, occupying about 60% of the jejunal lumen. Patient was in intensive care for 48 hours post-opperatively with respiratory support. He had an uneventful perioperative recovery and was discharged home on postoperative Day 9.

Histopathological examination revealed ulcerative exudate surrounded by neoplastic lymphoid infiltrates. Hallmark cells were also seen; hence the interpretation of ALCL (ALK negative) of small bowel with clear proximal and distal

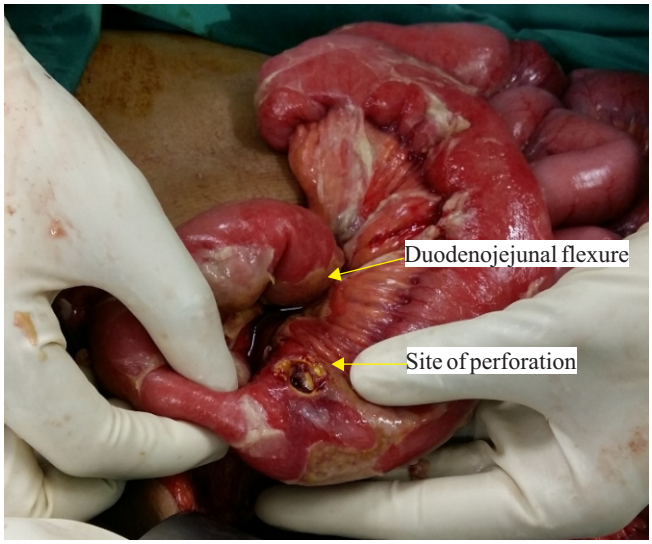

Figure 1. Intraoperative image showing the site of perforation at the mesenteric border of proximal jejunum

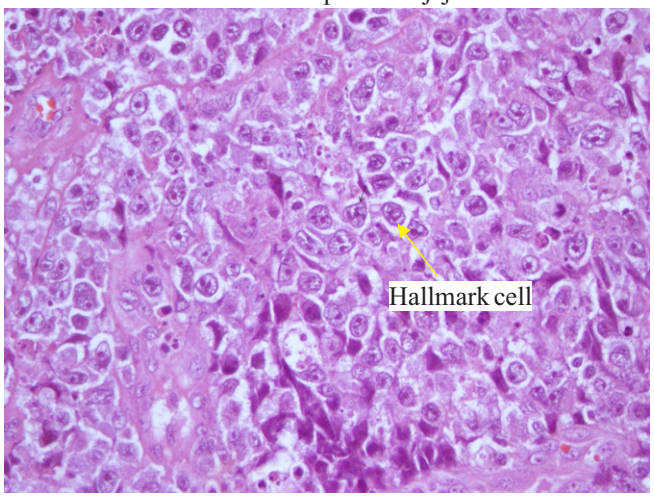

Figure 2. Hemotoxylin and eosin staining showing hallmark cells

Correspondence: Novinth Kumar Raja Ram
E-mail: novinth@gmail.com
Received: 03-03-2018 Accepted: 09-07-2018
https://orcid.org/0000-0001-8604-8918
DOI: http://doi.org/10.4038/sljs.v36i2.8518

margins was made. Unfortunately he passed away 2 weeks later due to respiratory failure.

Discussion

ALCL belongs to a rare group of peripheral T cell lymphoma with 4 distinct forms, namely primary systemic ALK+, primary systemic ALK-, breast implant associated and primary cutaneous ALCL.

The diagnosis is established by histopathological examination where the 'hallmark cell' is pathognomonic. It has an eccentric nucleus, and a prominent, pale Golgi region or a paranuclear hof. Systemic ALCL can further be sub classified according to the presence or absence of ALK (anaplastic lymphoma kinase), which has an impact on the prognosis.

This report highlights a rare occurrence of a metastatic perforation secondary to ALCL, the pathogenesis of this complication and the choice of management.

Presence of gastrointestinal disease plays a huge impact in the course and outcome of lymphoma(1).Bowel perforation manifests in advanced stage of the disease. Small bowel is the commonest region to perforate owing to its relatively thinner wall. In this case, the perforation occurred in the jejunum.

The pathogenesis of bowel perforation secondary to metastatic carcinoma is consequent to spread of the malignant cells to the intramural portion of the bowel. When the cells become non-viable, perforation takes place, whereas if they remain viable and grow into the lumen, obstruction occurs (2).The use of chemotherapy and steroids accelerates the process of tumour necrosis with the intestine (3). Even though systemic chemotherapy along with steroids had been administered to this patient, the histopathology revealed ulceration rather than tumour necrosis. Hence, the probable pathogenesis of the bowel perforation in this case is infiltration of lymphoma cells into the bowel wall through the lymphatics. Subsequently, mucosal ulceration ensued, slowly increasing in depth combined with increased intraluminal pressure leading to focal rupture of bowel.

Although emergency surgery for bowel perforation secondary to metastatic lymphoma is inevitable, the outcome is generally dismal, as gastrointestinal perforation secondary to metastasis from extra-abdominal primary is regarded as a terminal event in the course of the disease. Majority of the patients succumb to the illness within 6 months of the bowel perforation. There is only one report citing a patient who survived up to 5 years post bowel resection for a metastatic lymphoma (4) .

Keeping this in mind, the best operative course for patients with bowel perforation resulting from a metastatic tumor, would be a resection of devitalised bowel along with the metastatic lesion, and fashioning of an end stoma with a mucus fistula (5). This would eliminate the possibility of anastomotic leak and the complications associated with it, aiming to provide the patient with an acceptable quality of life. We adhered to this principle in our case with the exception of stoma, due to the proximal location of the perforation.

Conclusion

Small bowel perforation secondary to metastatic malignant lymphoma is a rare phenomenon. However, it is prudent to consider this differential when we approach a patient with spontaneous perforation of the hollow viscous. Due to the poor survival associated with this condition, the surgical management should be one that subjects the patients to the least amount of stress which would promote a faster perioperative recovery and in turn, a better quality of life.

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

References

1. Ara C., Coban S, Kayaalp C. et al. Spontaneous Intestinal Perforation Due to Non-Hodgkin's Lymphoma: Evaluation of Eight Cases. *Dig Dis Sci* 2007; 52: 1752-56 doi: 10.1007/s10620-006-9279-x
2. Leidich RB, Rudolf LE. Small bowel perforation secondary to metastatic lung carcinoma. *Ann Surg* 1981 Jan;193:67–69 PMCID: PMC1345004
3. Wada M, Onda M, Tokunaga A, Kiyama T et al. Spontaneous gastrointestinal perforation in patients with lymphoma receiving chemotherapy and steroids. J Nippon Med Sch 1999;66:37–40 PMID: 10097589
4. Ise N, Kotanagi H, Morii M, Yasui Oet al.Small bowel perforation caused by metastasis from an extra-abdominal malignancy: report of three cases.Surg Today. 2001;31(4):358-62. doi: 10.1007/s005950170161
5. Shiraishi M, Hirayasu S, Nosato E, Shimoji H et al.Perforation due to metastatic tumors of the ileocecal region.World J Surg 1998; 22:1065–68. PMID: 9747168

Learning Points:

- To consider spontaneous hollow viscous perforation as a differential in a patient who presents with acute peritonitis
- In the surgical management of a patient with metastatic small bowel perforation secondary to lymphoma, to perform a procedure that subjects the patient to the least surgical stress, individually tailored according to location of perforation and extent of peritoneal contamination