# Emergency Operation for Non-Hodgkin Lymphoma of the Small Intestine

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We report 5 cases of non-Hodgkin lymphoma of the small intestine (S-NHL) treated with emergency operation. These cases showed three reasons for emergency operation: [1] massive hemorrhage with shock before diagnosis of S-NHL (Case 1), [2] obstruction of the small intestine before and during chemotherapeutic treatment for S-NHL (Cases 2 and 3), and [3] spontaneous perforation with peritonitis before diagnosis of S-NHL or iatrogenic perforation following chemotherapy (Cases 4 and 5). For tumor discovery, double-balloon endoscopy of the small intestine was employed in 3 cases. Three tumors were histologically diagnosed before treatment, while 2 were histopathologically diagnosed using the resected specimens after emergency operation. An advanced stage of NHL was frequently observed. No surgical mortality accurred. We always consider the possibility of emergency operation before, during, and after the diagnosis and treatment of patients with S-NHL. Shinshu Med J 60: 21—25, 2012

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I Introduction

Non-Hodgkin lymphoma (NHL) is the third most common malignant neoplasm of the small intestine1). The small intestine is the second most common site for NHL of the gastrointestinal (GI) tract<sup>2)</sup>, and NHL of the small intestine (S-NHL) is observed in approximately 20 % of GI-NHL in Japan<sup>3)</sup>. Previously, surgery was frequently adopted for GI-NHL patients for both the treatment and histologic diagnosis4). However, initial chemotherapy has recently been performed in GI-NHL patients3)5). Recently, new endoscopic techniques, including capsule-endoscopy and DBE, have been made available for several tumors and disorders of the small intestine<sup>6)7)</sup>. These devices have contributed to the diagnosis of S-NHL, including not only the detection of S-NHL but also the sampling of biopsy specimens<sup>8)9)</sup>. The role of surgery in S-NHL could change after the development of these endoscopic techniques and devices. However, surgery, especially emergency operations due to acute abdominal conditions, should still be considered for S-NHL patients as perforation or obstruction of the small intestine has been reported in S-NHL patients with or without chemotherapy<sup>10)</sup>. We report 5 cases of S-NHL that required emergency operation.

## II Case Reports

Case 1, a 75-year-old woman, was operated on for massive hemorrhage of the GI tract, clinically showing dynamic shock. Before surgery, no cause of massive intestinal hemorrhage had been identified. At laparotomy, a jejunal tumor was detected and removed. The resected tumor was histopathologically diagnosed as diffuse large B-cell lymphoma (DLBCL) of the jejunum after surgery. Three months after the operation, the patient died of the systemic involvement of NHL, although the postoperative course was uneventful.

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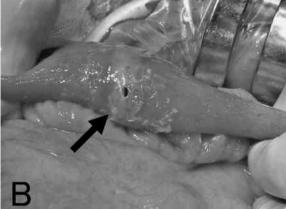


Fig. 1 Operative findings

- A Case 3: Intestinal obstruction caused by repeated chemotherapy. The arrow shows that the obstruction originated from the intestinal lymphoma with degenerative and fibrous changes.
- B Case 4: Spontaneous perforation of the intestinal lymphoma. The arrow shows the perforation.

Case 2, a 68-year-old man, underwent an emergency operation involving partial resection of the ileum. Intestinal obstruction due to rapid enlargement of the ileal tumor was observed when the patient was admitted for chemotherapy for DLBCL of the ileum detected by double balloon endoscopy (DBE). The postoperative course was uneventful. After surgery, chemotherapy using cyclophosphamide, doxorubicin hydrochloride, vincristine, and methylpredonisolone with rituximab (R-CHOP) was performed. The patient is well without any recurrence 42 months after surgery.

Case 3, a 52-year-old man, showed intestinal obstruction after 4 cycles of CHOP chemotherapy for T-cell lymphoma of the jejunum diagnosed by DBE. Although conservative treatment for the intestinal obstruction using a long intestinal tube was performed, it did not improve. The patient underwent an emergency operation (Fig. 1A). The postoperative course was uneventful. Although further chemotherapy was performed, the patient died of central nervous system involvement of S-NHL 18 months after surgery.

Case 4, a 78-year-old man, was operated on due to diffuse peritonitis with sudden onset. At lapar-otomy, a perforated tumor was detected in the jejunum and removed (Fig. 1B). The resected tumor was histopathologically diagnosed as DLBCL after surgery. The postoperative course was uneventful,

and the patient received systemic chemotherapy using R-CHOP. The patient is well without any recurrence 36 months after surgery.

Case 5, a 78-year-old woman, had diffuse peritonitis due to a perforation of the duodenum, when balloon dilation was endoscopically performed for stenosis caused by repeated chemotherapy using CHOP for duodenal DLBCL. The patient surgically underwent wide drainage of the abdominal cavity. Although infection of the postoperative surgical site was observed, the patient survived for 61 months after surgery without recurrence. The clinicopathologic features of the 5 S-NHL cases treated by emergency operation are summarized in Table 1. All patients satisfied the diagnostic criteria for primary lymphoma of the gastrointestinal tract described by Lewin et al111. The histologic classification of NHL was carried out according to WHO criteria<sup>12)</sup>. The NHL stage was determined according to the Lugano International Conference Classification<sup>13)</sup>.

# III Discussion

Emergency surgery has occasionally been necessary for S-NHL patients. There are a few recent reports concerning limited investigation of this specific situation regarding S-NHL<sup>14)15)</sup>, although several reports concerning emergency surgery for small intestinal tumors, including gastrointestinal

Table 1 Cases of small intestinal non-Hodgkin lymphoma treated by emergency operation

(		Creaman	Do+00+1	Reason	C	Ninicopathologic features	gic features		Before	Before surgery	After Surgery	urgery	Surgical	Outcome after
Cases		Age/ Sex Symptom Detection	Detection	for surgery	Site	Size (mm)	Size (mm) Histology Stage* Cx Rx	Stage*	Cx	Rx	Cx Rx	Rx	morbidity	surgery (months)
-	75/F	bleeding	surgery	shock	jejunum	54	DLBCL	IV	I	ı	I	ı	ISS	died (3)
2	M/89	pain	DBE	ileus	ileum	09	DLBCL	IV	I	I	+	I		alive $(42)$
3	52/M	pain	DBE	ileus	jejunum	150	T-cell	$\Pi_{\mathrm{E}}$	+	I	+	+	Ι	died (18)
4	78/M	pain	surgery	perforation	jejunum	40	DLBCL	$\Pi_{\mathrm{E}}$	I	I	+	I	1	alive (36)
2	$78/\mathrm{F}$	pain	DBE	perforation	duodenum	48	DLBCL	$\Pi_1$	+	I	+	I		alive $(61)$

\*, Lugano staging classification; Cx, chemotherapy; Rx, radiotherapy.

DBE, double balloon endoscopy; DLBCL, diffuse large B-cell lymphoma; SSI, surgical site infection.

stromal tumor and other tumors, are available<sup>16)17)</sup>. The patients were urgently operated on for various reasons such as massive bleeding and dynamic shock (Case 1), obstruction of the small intestine without and with chemotherapy (Cases 2 and 3), and spontaneous or iatrogenic perforation of the small intestine (Cases 4 and 5). These emergency situations were encountered in every period around the treatment of S-NHL. We should always consider the possibility of emergency surgery before, during, and after treatment in patients with S-NHL.

Intestinal hemorrhage has been clinically observed in approximately 10 % of S-NHL: 6.3 % reported by Koch et al2) and 14.3 % by Daum et al18). Massive hemorrhage caused by S-NHL has rarely been observed in patients involving the lower GI distal to the ligament of Treitz, and its frequency was approximately 1-2 % of patients with small intestinal bleeding<sup>19)20)</sup>. Massive hemorrhage of S-NHL is a fatal complication, although Case 1 with dynamic shock was cured by emergency surgery and intensive care. The patient did not receive chemotherapy after surgery, because the general condition worsened. It was expected that the clinical outcome would be poor. The emergent state of S-NHL patients is an issue concerning the clinical outcome after surgery.

Regarding intestinal obstruction, NHL was the second-most common tumor of the small intestine<sup>17)</sup>, and it was frequently observed in S-NHL patients<sup>2)21)</sup>. This complication can affect the diagnosis and treatment of S-NHL. Usually, elective surgery was performed for intestinal stenosis caused by S-NHL before chemotherapy, and chemotherapy was subsequently performed. Emergency surgery would be required for S-NHL patients with intestinal obstruction, and two modes of obstruction should be considered for S-NHL patients: chemotherapeutic complications of S-NHL, such as Case 3; and optimal surgery for an unknown tumor with obstruction of the small intestine.

Perforation of the small intestine has sometimes been observed in patients with S-NHL. Koch et al<sup>2)</sup> reported that it was observed in 9.4 % of S-NHL patients. Daum et al<sup>18)</sup> reported that it was observed in 25 % of S-NHL patients, and there was a high rate (37 %) in T-cell lymphoma. Free perforation of the small intestine or tumors occurs spontaneously or after chemotherapy in patients with S-NHL<sup>14)22)</sup>. When NHL is treated by effective chemotherapy, perforation of the GI tract may be a potential complication caused by tumor degeneration. However, as in Case 5, iatrogenic perforation due to balloon dilation for stenosis after chemotherapy for S-NHL has been rarely reported. New technical devices for endoscopy have been developed and archived for small intestinal disorders including S-NHL. We considered these complicated perforations even though they are unlikely in S-NHL patients, involving not only chemotherapeutic effects but also iatrogenic complications.

Surgery for malignant tumors of the small intestine, including S-NHL, has often been associated with a high mortality rate; previous reports demonstrated a rate of 5 % or more<sup>15)17)23)</sup>. In our series, no surgical mortality was observed. Furthermore, intestinal perforation has been associated with a poor prognosis in patients with S-NHL<sup>15)18)</sup>. Two of the 5 patients showed a poor outcome, with lower-level morbidity and morbidity after emergency

operation. As a reason for this, emergent situations such as perforation and obstruction of the small intestine are often characterized by systemic inflammatory response syndrome, and an immunosuppressed state may be observed. However, in S-NHL patients, no information has been made available regarding these emergent situations, and further studies may be needed. Careful management of S-NHL patients undergoing emergency surgery is required for a safe postoperative course, and promoting effective postoperative treatment and prolonged survival.

In conclusion, the recent role of surgery for S-NHL patients has changed after the clinical presentation of new endoscopic techniques and devices, such as DBE and capsule-endoscopy, for the small intestine. We should consider the possibility of emergency surgery before, during, and after management and treatment in patients with S-NHL.

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