

## Ulcerative form of small intestinal lymphoma causing massive lower GI bleeding : a case report.

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*A case of an 18-year-old man with primary small intestinal lymphoma (PSIL) is reported. The patient's initial presenting symptom was massive hematochezia, which is uncommon for PSIL. A barium enema revealed an unusual location of the terminal ileum, which was directed upward and located medial to the ascending colon. Conventional upper GI and small bowel follow-through study with compression technique demonstrated a broad-based ulcer with well-defined margin, situating about 10 cm proximal to the ileocecal valve. A single ulcer of the terminal ileum is an unusual radiological feature for lymphoma. Histopathology of the lesion revealed poorly differentiated lymphocytic lymphoma.*

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ลินดา พานธงรักษ์-บรวาน์, บุญเที่ยง ศีตีสาร. เลือดออกในทางเดินอาหารส่วนปลาย สาเหตุจากлимโฟมาของลำไส้เล็ก : รายงานผู้ป่วย 1 ราย. จุฬาลงกรณ์เวชสาร 2533 สิงหาคม ; 34 (8) : 607-612

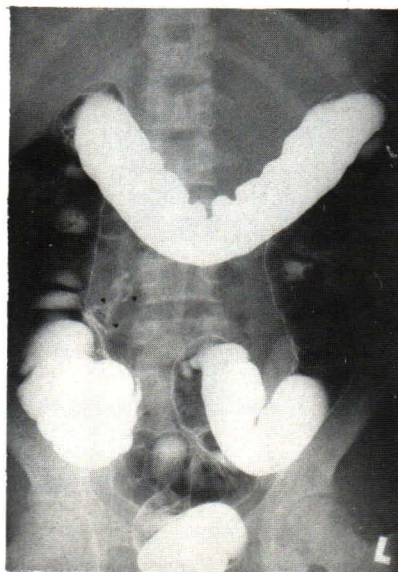
รายงานผู้ป่วยชาย อายุ 18 ปี ได้รับการวินิจฉัยว่าเป็น *primary small intestinal lymphoma (PSIL)* อาการนำของผู้ป่วย คือถ่ายเป็นเลือดสดๆ จำนวนมาก ซึ่งเป็นอาการนำที่พบได้ไม่บ่อยนักใน *PSIL* การตรวจ *barium enema* พบว่าส่วนของ *terminal ileum* อยู่ในตำแหน่งที่ผิดปกติ คือชี้ขึ้นด้านบน และวางตัวอยู่ด้านในต่อลำไส้ใหญ่ส่วนต้น การตรวจ *upper GI* และ *small bowel follow-through* พร้อมทั้งการ *compression* ลำไส้เล็ก พบแผล 1 แผล ที่มีฐานแผลกว้าง และมีขอบเขตชัดเจนอยู่ที่บริเวณ *terminal ileum* ห่างจาก *ileocecal volve* ประมาณ 10 เซนติเมตร ลักษณะแผลที่พบเพียงแผลเดียวในลำไส้เล็กโดยที่ไม่พบความผิดปกติอย่างอื่น เป็นลักษณะภาพทางรังสีที่พบได้ไม่บ่อยนักสำหรับлимโฟมา การตรวจทางพยาธิสภาพ พบว่าเป็น *poorly differentiated lymphocytic lymphoma*.

Small intestinal lymphoma exhibits a broad spectrum of radiological appearances. Typical radiological features are aneurysmal dilatation, nodular filling defects, constrictive segments, and large extraluminal masses.<sup>(1)</sup> Occasionally, an ulcerative form is seen, consisting of multiple ulcerated lesions with relatively normal intervening mucosa.<sup>(1)</sup> A single ulcer is, in contrast, an uncommon finding in intestinal lymphoma. We, therefore, report such an unusual radiological feature of lymphoma involving the terminal ileum.

## CASE REPORT

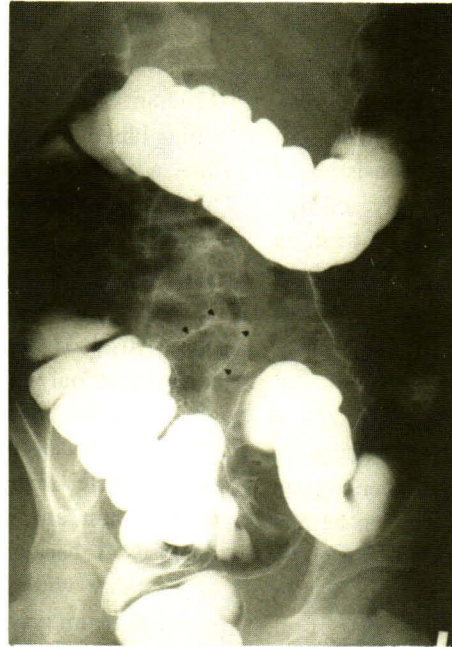
An 18-year-old man presented to Chulalongkorn Hospital with massive hematochezia. He had been in good health until 7 months prior to admission when he had developed massive rectal bleeding without preceding abdominal pain. He had been hospitalized in a district hospital where blood transfusion (2 units) was given. Peptic ulcer had been diagnosed at that time without any further investigation. Twenty days later, the patient had a second episode of rectal bleeding and was admitted to another district hospital. Gastroscopy was performed and no peptic ulcer was detected. He received another blood transfusion and was discharged home. One month prior to admission, he had a few more episodes of rectal bleeding. He presented to a provincial hospital where a barium enema and upper GI with small bowel follow-through were performed. No abnormality was discovered within the gastrointestinal tract. The patient was then referred to Chulalongkorn Hospital. His weight decreased about 3 kilograms over the 7 months.

Physical examination revealed a weak and pale young man with good consciousness. His heart rate was 120/min. Supine blood pressure was 90/50 mm Hg, while sitting blood pressure was 80/30 mm Hg. Body temperature was 38°C. Conjunctiva was markedly pale. No lymphadenopathy was palpable. The abdomen was soft with no organomegaly. The physical examination was otherwise within normal limits. His hemoglobin was only 6.2 gm% and the red blood cell morphology was microcytic and hypochromic. Flexible sigmoidoscopy was remarkable only for of a hemorrhagic spot about 40 cm from the anus which the clinician believed was traumatic and secondary to the procedure. A <sup>99m</sup>Tc-pertechnetate scan was done to rule out Meckel's diverticulum and showed no ectopic area of gastric mucosa. A barium enema was done and showed no abnormality within the large intestine. However, the terminal ileum was in an unusual position (Fig. 1,2). It pointed upward, located medial to the ascending colon, instead of its normal downward or horizontal direction. This finding indicated that there probably were adhesions to the terminal ileum. An upper GI and small bowel follow-through was then performed. Compression of the distal part of the small intestine revealed evidence of a broad-based, and well-defined ulcer with edema of the surrounding mucosa, situating about 10 cm proximal to the ileocecal valve (Fig. 3). The overhead radiograph of the study did not show any abnormality (Fig. 4). An exploratory laparotomy was then performed, based on the result of the upper GI and small bowel follow-through study. The ileum was resected with end-to-end anastomosis. No intra-abdominal lymphadenopathy was observed.

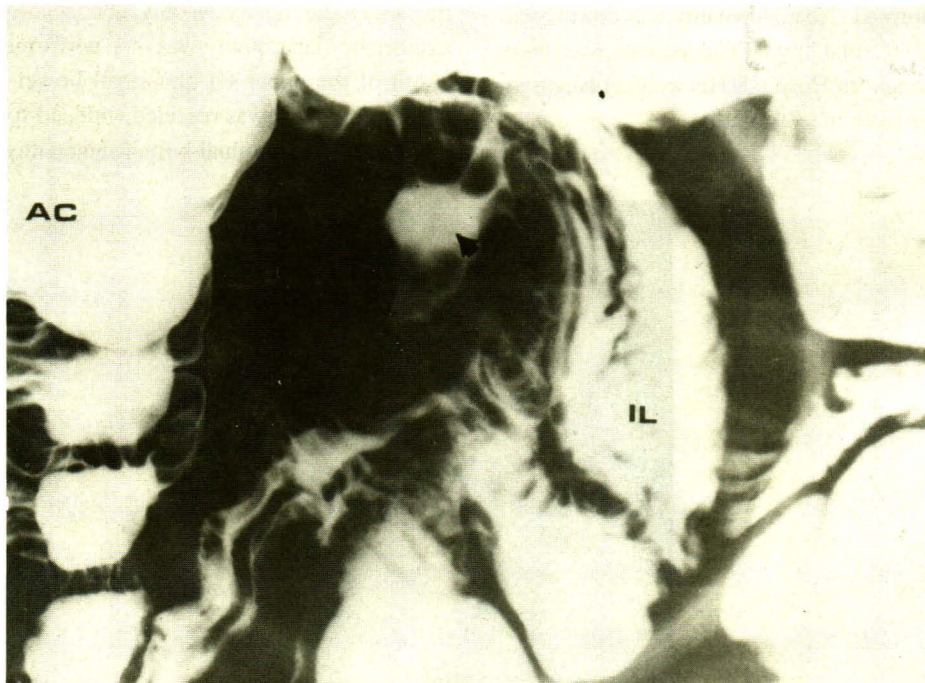


**Figure 1.** Barium enema, PA view, with barium outlining the contracted terminal ileum. The terminal ileum situates in an unusual location, which is directed upward and located medial to the ascending colon (arrowheads).





**Figure 2.** Barium enema, oblique view, with barium outlining the distended terminal ileum. Note the unusual location of the terminal ileum (arrowheads).



**Figure 3.** Compression radiograph of the terminal ileum, following a conventional upper GI and small bowel follow - through study, revealing a broad - based and well-defined ulcer (arrow) with edema of the surrounding mucosa. (AC = ascending colon, IL = ileum).



**Figure 4.** Overhead radiograph of the conventional upper GI and small bowel follow - through study showing no abnormality.

Gross pathology revealed a large ulcer, about 3 cm in diameter, with a well-defined border. Microscopic section through the ulcer showed evidence of poorly differentiated lymphocytic lymphoma. Post-operation, CT scan of the whole abdomen was performed and no lymphadenopathy or other organ involvement by lymphoma was observed. A course of chemotherapy was then administered. The patient recovered uneventfully and was finally discharged home.

## DISCUSSION

Intestinal lymphoma is considered to be "primary" if 1) the predominant lesion is in the intestine, 2) the initial presenting symptoms are related to intestinal involvement and 3) there is no evidence of a generalized disease or intestinal predisposing disorder.<sup>(2,3)</sup> Although the investigation for other sites of lymphoma was limited, this case is consistent with primary small intestinal lymphoma (PSIL). PSIL is not common and represents about 2-4% of all non-Hodgkin's lymphoma.<sup>(4,5)</sup> Although PSIL is not common, it is still one of the most common primary malignant neoplasms of the small intestine.<sup>(6)</sup> PSIL is common in adolescence<sup>(7)</sup> and has a male to female ratio of approximately 2:1.<sup>(1)</sup> PSIL may present as a surgical emergency due to perforation or obstruction or, rarely, massive hemorrhage,<sup>(7)</sup> as

presented in our case.

The small intestine is one of the most difficult areas of the gastrointestinal tract to evaluate. Endoscopic visualization of the small intestine is not possible and barium contrast studies remain the primary diagnostic tool. A barium enema (BE) with reflux into the terminal ileum may be adequate in some patients with suspected small bowel disease. However, the ability to achieve reflux is unpredictable, and it is difficult to distend the terminal ileum enough to maximize the diagnostic accuracy in this area.<sup>(8)</sup> As shown in our case, a BE could be interpreted as normal if the study was not scrutinized carefully. Although the barium did not adequately fill the terminal ileum, the unusual location of the terminal ileum was an indirect sign of possible pathology within the distal part of the small intestine (Fig. 1,2). The upper GI and small bowel follow-through with compression technique was able to demonstrate the lesion in our case, which was a broad-based, well-defined ulcer, situated approximately 10 cm proximal to the ileocecal valve (Fig. 3). This conventional small bowel study has been described as having high false negative rates and small bowel enema or enteroclysis may be more sensitive and accurate.<sup>(8,9)</sup> The patient was very fortunate that the lesion was detected by the conventional method, since enteroclysis is not a regular practice at our institution. Angiography would be the next procedure of choice if

the lesion is not demonstrated by any non-invasive barium contrast study. However, angiography is invasive and the bleeding point is unlikely to be localized unless there is active bleeding during performance of the procedure. The alternative non-invasive imaging techniques to localize the bleeding point are the  $^{99m}\text{Tc}$ -sulfur colloid scan or the  $^{99m}\text{Tc}$ -red blood cells scan. However, it also may be difficult to distinguish the site of bleeding by the isotope scanning techniques since extravasated blood is rapidly spread by intestinal propulsion and tends to accumulate in the right colon.<sup>(10)</sup>

The principal radiological manifestations of intestinal lymphoma are the aneurysmal, constrictive, nodular, ulcerative, mesenteric, and sprue forms.<sup>(1)</sup> A combination of different forms is a frequent finding. The ulcerative form of intestinal lymphoma usually presents as multiple ulcerated lesions with relatively normal intervening mucosa. A single ulcer, as presented radiologically in our case, is unusual. The site of the lesion, the terminal ileum, is a clue to correct diagnosis, since the terminal portion, near the ileocecal valve, is

where lymphoid tissue is normally most abundant.<sup>(11,12)</sup>

The differential diagnosis of a single ulcer of the terminal ileum includes Meckel's diverticulum, tuberculosis, typhoid fever, Crohn's disease, and isolated non-specific ulcer.

## CONCLUSION

This report described a case of PSIL, unusual in its initial presentation which was massive hematochezia, and in its radiological appearance which was a single ulcer of the terminal ileum. The conventional upper GI and small bowel follow-through with compression technique verified the site of the lesion.

## ACKNOWLEDGEMENT

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