


Case Report

Primary diffuse large B-cell lymphoma of the ileal conduit created after radical cystectomy

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Abbreviations & Acronyms

BCL = B-cell lymphoma
 CD = cluster of differentiation
 DLBCL = diffuse large B-cell lymphoma
 R-CHOP = rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone

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Introduction: Creation of an ileal conduit is associated with complications. A few cases have been reported on tumor development in an ileal conduit; diffuse large B-cell lymphoma originating from an ileal conduit is extremely rare.

Case presentation: A 62-year-old Japanese man who had undergone radical cystectomy and ileal conduit diversion 6 years previously presented with a whitish bulge that had developed on the surface of the ileal conduit during follow-up visit. Mass biopsy was performed and the histological diagnosis was diffuse large B-cell lymphoma. Positron emission tomography showed no metastatic lesions. We attempted chemotherapy because the tumor cells tested positive for CD20; after six courses of a regimen involving rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone; diffuse large B-cell lymphoma of the ileal conduit resolved gradually. The patient is alive and remains free from the diagnosed diffuse large B-cell lymphoma.

Conclusion: We reported an extremely rare case of diffuse large B-cell lymphoma originating from the ileal conduit created after radical cystectomy.

Key words: diffuse large B-cell lymphoma, ileal conduit.

Keynote message

We hereby report an extremely rare case of DLBCL originating from the ileal conduit created after radical cystectomy. Although its prevalence is rare, physicians should consider such a lymphoma when tumor formation at the stomal site is detected during long-term follow-up visits.

Introduction

Ileal conduit diversion is a popular technique for urinary diversion following radical cystectomy. Creation of an ileal conduit is associated with complications such as surrounding dermatitis, urinary obstruction, and acute pyelonephritis; however, a few cases have been reported on tumor development in an ileal conduit. DLBCL originating from an ileal conduit is extremely rare. DLBCLs occur in organs of the digestive tract, primarily in the stomach and ileum, but only a single case of DLBCL in the ileal conduit has been previously reported.¹

Gastrointestinal lymphoma usually arises secondarily from widespread nodal diseases, but primary gastrointestinal lymphoma is rare.² As far as we know, this is the second case of DLBCL on the surface of the ileal conduit, created following radical cystectomy.

Case presentation

A 62-year-old Japanese man presented to our institution with a whitish bulge in the stoma of his ileal conduit during his follow-up examination after radical cystectomy and ileal conduit diversion that had been performed 6 years previously. He faced difficulties while placing ostomy appliances because of an enlarged stoma. His medical history included hypertension, atrial fibrillation, esophageal cancer, stomach cancer, and bladder cancer. At the age of

55 years, he was diagnosed with bladder cancer and underwent radical cystectomy and ileal conduit diversion. The histological diagnosis was urothelial carcinoma (pT1). He remained free from the disease for 78 months post-operation. At the age of 62 years, he became aware of a whitish bulge that had developed on the surface of the ileal conduit. This newly developed mass was hard and necrotic (Fig. 1a). A total-body computed tomography scan revealed a contrast-enhanced mass in a limited area of the stoma but there was no distant metastasis or lymph node involvement (Fig. 2a). The differential diagnoses were considered to be a neoplasm or inflammatory changes due to a mismatch between the size of the stoma and that of the ostomy flange. Tissue biopsy was performed. Histopathological analysis using hematoxylin and eosin staining revealed an increased number of diffuse-type, large-sized carcinoma cells with high nucleus-cytoplasm ratio (Fig. 3a). On immunohistochemical staining, the tumor cells tested positive for B-cell markers such as the CD20 (Fig. 3b) and BCL6 but tested negative for CD5 and CD10 as well as for Epstein–Barr virus-encoded small ribonucleic

acid *in situ* hybridization. The final histopathological diagnosis was DLBCL. No lesions were detected by positron emission tomography. Moreover, bone marrow examination revealed no abnormal findings. According to the Lugano classification for gastrointestinal tract lymphoma, the patient was classified as stage 1.³ When six courses of R-CHOP regimen were administered, DLBCL on the ileal conduit resolved gradually (Figs 1b,2b). The patient is alive and remains free from DLBCL 30 months after diagnosis.

This study was approved by the institutional review board and was performed in accordance with the Declaration of Helsinki guidelines.

Discussion

In our hospital, we encountered the second case of DLBCL originating from an ileal conduit created after radical cystectomy.

The most common extranodal site of lymphoma is the gastrointestinal tract, but the occurrence of primary gastrointestinal lymphoma is uncommon.² From the viewpoint of

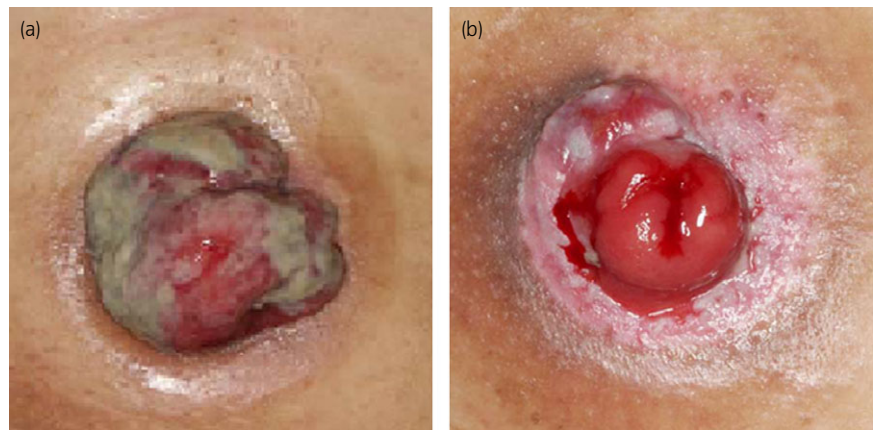


Fig. 1 An image of the stoma. (a) Enlarged stoma accompanied with a whitish mass at diagnosis, (b) normal colored stoma after six courses of R-CHOP regimen.

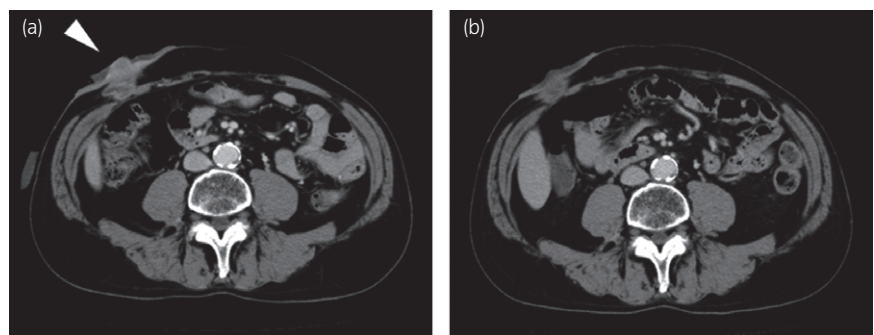


Fig. 2 Horizontal view of the contrast-enhanced computed tomography scan. (a) A mass (arrowhead) on the surface of the ileal conduit, (b) the mass distinguished after six courses of R-CHOP regimen.

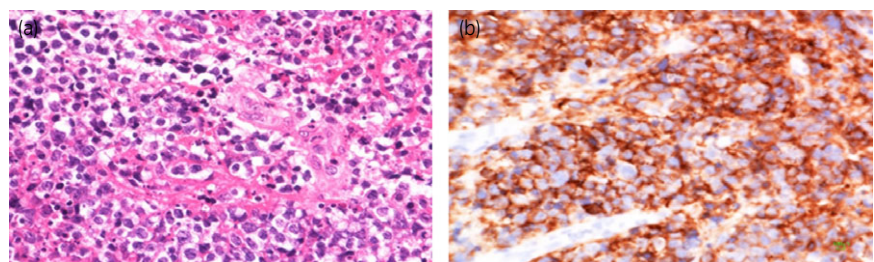


Fig. 3 (a) Hematoxylin and eosin staining (high-power field, original magnification $\times 400$) showing large-sized carcinoma cells with a high nucleus-cytoplasm ratio; (b) immunoperoxidase staining (high-power field, original magnification $\times 400$) revealing CD20-positive tumor cells.

histopathology, 90% of primary gastrointestinal lymphomas are of B-cell lineage; T-cell lymphomas or Hodgkin lymphomas are extremely few.² Lymphomas represent 15–20% of all small intestinal neoplasms and 20–30% of all primary gastrointestinal lymphomas; the ileum is the most common site for small intestinal lymphoma.² Several factors such as age, stage of disease, lactate dehydrogenase level, and use of chemotherapy are independently and significantly associated with survival.⁴

A previous study reported positive associations between DLBCL and rheumatoid arthritis, Sjogren syndrome, systemic lupus erythematosus, and celiac disease.⁵ Inflammatory bowel disease,⁶ prolonged use of azathioprine or 6-mercaptopurine,⁷ human immunodeficiency virus infection,⁸ and congenital or acquired immunodeficiencies⁹ have been reported to be the risk factors for lymphoma development at the stoma of the digestive tract. Muta *et al.* reported that long-term inflammation or immunodeficiencies might be the underlying risk factors for the occurrence of lymphomas,¹ such as the one mentioned in our case. The mucosal layer of the stoma remained wet owing to the presence of urine, which led to a change in the microbial environment of the stoma.¹ Continuous stimulation of close contact between the stoma and the flange may be one of the causes for lymphoma development. Moreover, Ota *et al.* reported a case of DLBCL of the ileal neobladder, in which the patient had no medical history of inflammatory bowel disease or immunosuppressive therapy; they mentioned that the cause of DLBCL in their case might be associated with chronic infection.^{10,11} The cause of DLBCL in our case was unclear because the patient had a past medical history of esophageal cancer, stomach cancer, and bladder cancer but did not use chemotherapy or have diseases that lead to immunodeficiencies. However, it can suffice to say that continuous chronic inflammation due to the flange and urine is the most reasonable cause of DLBCL in this case.

Preoperative biopsy, performed at an early stage, is a feasible way to decide disease management.¹¹ Generally, chemotherapy, surgery, and radiation therapy are considered for treating DLBCL. In this case, we selected chemotherapy because the tumor cells tested positive for CD20, and the R-CHOP regimen was expected to be efficacious. The advantages of avoiding surgery were compensated with the risks of toxicity and septicemia after chemotherapy because of the necrotic tissues.¹¹ Radiation therapy was not our first choice of treatment because adhesion of the tumor with surrounding

tissues would render surgeries, if required, difficult, following radiation therapy. Although, radiation therapy is considered to be successful in the down-staging of the local lesion, it poses the risk of causing radiation-induced enterocolitis.¹¹

Conclusions

We reported an extremely rare case of DLBCL originating from the ileal conduit created after radical cystectomy. Although its prevalence is rare, physicians should consider such a lymphoma when tumor formation at the stomal site is detected during long-term follow-up visits.

Conflict of interest

The authors declare no conflict of interest.

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