

Primary mantle cell lymphoma complicated with cutaneous diffuse large B-cell lymphoma, leg type: a rare case report of composite non-Hodgkin lymphoma in different organs

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Case Report

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Abstract

Background

Composite lymphomas (CLs) are a kind of rare disease that two distinct categories of lymphomas occur in the same patient. Histologically, composite lymphomas can be composed of a Hodgkin's lymphoma and a non-Hodgkin lymphoma or two distinct non-Hodgkin lymphomas. So far, most of the cases have been reported to occur in a single anatomical site or mass.

Case presentation:

A 61-year-old man without any B-type symptoms complained of an enlarging mass in the abdomen for one month. A 10 × 10 cm abdominal mass could be touched in the hypogastric region. Through pathological biopsy, mantle cell lymphoma can be diagnosed. After one cycle chemotherapy regimen of FCD, red rashes and blisters came out on the patient's right lower extremity. Cutaneous diffuse large B-cell lymphoma (DLBCL) was diagnosed by skin biopsy. In this report, we describe a case of composite lymphoma occurring in different organs, which consisted of primary mantle cell lymphoma (MCL) and cutaneous DLBCL, leg type. The patient then received a series of chemotherapy regimens without rituximab then achieved partial response (PR).

Conclusions

To our knowledge, this is a rare case of CLs occurring in different anatomic sites that were treated by chemotherapy and achieved PR. As we learn more about the mechanisms and treatment of CLs, we look forward to more treatment options in the future for patients to give them a better prognosis.

Background

Composite lymphomas (CLs) are a kind of rare disease that two distinct categories of lymphomas occur in the same patient. Histologically, composite lymphomas can be composed of a Hodgkin's lymphoma and a non-Hodgkin lymphoma or two distinct non-Hodgkin lymphomas¹. Some cases involving two different subtypes of Hodgkin's lymphoma have been reported². True CLs are rare, and most CLs occur in a single anatomical site or mass. In this report, we describe a case of composite different lymphoma occurring in different organs, which involved primary mantle cell lymphoma (MCL) and cutaneous diffuse large B-cell lymphoma (DLBCL), leg type.

Case Presentation

A 61-year-old man without any B-type symptoms complained of an enlarging mass in the abdomen for one month. Physical examination revealed a firm, rubbery swollen lymph nodes in the bilateral neck and

axilla, of which the biggest one was about 4 × 2 cm. Furthermore, a 10 × 10 cm abdominal mass could be touched in the hypogastric region. He does not have any chronic diseases, and his family has no related genetic medical history. Complete blood count and other serum chemistries were normal except for LDH levels and serum CA125 concentration which were as high as 343U/L and 117.8 U/ml. Computed tomography (CT) of the chest, abdomen, and pelvis demonstrated multiple enlarged lymph nodes, the abdominal mass, and the thickening wall of the gastric antrum (Fig. 1A). Immunohistochemistry (IHC) of cervical lymph node biopsy showed strongly stained with CD20, CD79a, CD19, CD5, Bcl-2 and CyclinD1, which indicated mantle cell lymphoma (MCL). The Levels of Ki67 expression in tumors is 20%. Meanwhile, a bulge under the mucosa of the stomach was found by gastroscopy and intestinal lymphoma was discovered by colonoscopy (Fig. 2A, B, C). Microscopically, a large number of small lymphocytes infiltrated in the intestinal mucosa of the ileocecal area. IHC revealed CD20(+), CyclinD1(+), CD23(-), Bcl-6 (-) and Ki-67 (30%). MCL can be confirmed histopathologically. The IHC of the bone marrow biopsy demonstrated that the bone marrow cells were partially positive for CD20, CD79a and CyclinD1. In summary, we also found evidence of MCL involvement in both the bone marrow and ileocecal. The patient was diagnosed with MCL (Ann Arbor stage IV, International Prognostic Index (IPI) score of 4), with bone marrow and alimentary tract infiltration. In order to show the patient's immunohistochemical pictures, we went to the department of pathology in our hospital to choose the pathological slides. Unfortunately, they have deteriorated so we did not show them in this report.

Because of the patient's limited financial resources, he refused to use rituximab to treat the disease. After one cycle chemotherapy regimen of FCD (fludarabine 50 mg, d1-3 + cyclophosphamide 400 g, d1-3 + dexamethasone 20 mg, d1-5), red rashes and blisters came out on the patient's right lower extremity, with no pain and itching (Fig. 3A). The skin biopsy of right thigh revealed lymphoid hyperplastic lesions. IHC revealed that the skin cells were strongly positive for CD20, CD79a, CD10, Bcl-2 and Bcl-6 and negative for CyclinD1, CD3, CD21, CD23 and SOX-II, the expression of Ki-67(90%). To our surprise, diagnosis of diffuse large B-cell lymphoma, leg type, non-GCB was confirmed. After four cycles of chemotherapy regimen of FCD, the patient had no obvious benefit. We recommend that the patient be injected with rituximab, but he refused to follow our recommendation to use rituximab. Then two cycles of chemotherapy with CHOPE regimen (Cyclophosphamide 1.3 g, d1 + Pirarubicin 90 mg, d1 + Vinorelbine 40 mg, d1 + Dexamethasone 15 mg, d1-5 + Etoposide 100 mg, d1-3) and five cycles of TGDP regimen (Thalidomide 100 mg qd + Gemcitabine 1.8 g d1, 8 + Cisplatin 130 mg d1 + Dexamethasone 40 mg d1-4) were applied. The only side-effect during chemotherapy was neutropenia, which could be alleviated after treatment. After 8 months of chemotherapy, CT review showed swollen lymph nodes and retroperitoneal masses are significantly shrunk than before (Fig. 2B). An electronic colonoscopy review suggested a microscopic bulge of the mucosa (Fig. 2D, E, F). After six cycles of TGDP regimen chemotherapy, some of the rashes were dissipated. Meanwhile, no obvious side effects and new rash were observed, and the superficial lymph nodes all over the body were significantly shrunk (Fig. 3B). The partial remission (PR) was achieved because we did not see tumor performance through gastrointestinal endoscopy. The patient has been remaining asymptomatic and clinically stable until now.

Discussion And Conclusions

Our study reports a rare case with two different Hodgkin's lymphomas in different parts of the body, namely, mantle cell lymphoma and diffuse large B-cell lymphoma. After the patient benefited from a series of chemotherapy. To our knowledge, this is the first report of a CL patient occurring in different sites to achieve PR by chemotherapy. Most importantly, this case could provide clinical insight into CLs.

MCL is a rare, aggressive B-cell lymphoma that usually presents with lymph node and spleen disease. In rare cases, MCL can be used as an extranodal lymphoma, most commonly in the gastrointestinal tract. Diffuse large B-cell lymphoma (DLBCL) is the most common type of non-Hodgkin lymphoma that develops from the B cells of the lymphatic system. It accounts for approximately 40–50% of new cases of lymphomas³. The incidence of combined lymphoma is 1–4%⁴. This report presents a rare case with two B-cell lymphomas in different organs, which could be classified as composite B-cell lymphoma. CL composed of two different B-cell non-Hodgkin lymphomas, most often represent combined mantle cell lymphoma with CLL or follicular lymphoma. Here we report a rare case combine MCL with DLBCL, leg type.

Clonal relations of the two components of composite B-cell non-Hodgkin lymphomas are the key to study the pathogenesis mechanism. The clonal relationship can be determined by rearranged immunoglobulin or T cell receptor (TCR) V gene since each lymphocyte has a unique receptor and the rearrangement of the V gene remains stable during cellular division¹. The possibility of conversion from MCL to DLBCL cannot be ignored, although it is rare. In the case of transformation, large cells express cyclin D1 protein and display the same clonality as the small cells. More commonly, large cell MCL can occur simultaneously with small cell MCL⁵. In this case, cyclin D1 was not expressed in the DLBCL component which indicates that DLBCL was not converted from the MCL.

In CLs, the two disease components should be taken into the decision of treatment. When different lymphomas occur sequentially, each disease should be treated according to the corresponding principles^{6,7}. When the interval time between the first and second lymphomas is short and the treatment of the first lymphoma is strong, the first-line method of the second lymphoma may not be appropriate because the cells may have already been in the first treatment. It is present and proven to be resistant to treatment⁸. Andhavarapu S et al. reported a case of testicular composite lymphoma consisting of DLBCL and MCL. The patient was treated with six cycles of R-CHOP. Meanwhile, intrathecal chemotherapy with methotrexate was administered in every cycle. After treatment, PET and bone marrow biopsy confirmed that the patient had complete remission⁹. Albert K⁵ also described 2 cases of composite MCL and DLBCL which were well treated with rituximab. Rituximab is a specifically targeted drug for the treatment of B-cell lymphomas, but it is expensive. In this report, we didn't use the rituximab due to the poor economic conditions of the patient. Finally, the patient achieved PR after we have chosen the FCD, CHOPE, and TGDP regimen of chemotherapy regularly.

For the treatment of complex lymphoma at different sites, we need more clinical trials to explore. As we learn more about the mechanisms and treatment of CLs, we look forward to more treatment options in the future for patients to give them a better prognosis.

Abbreviations

CLs: Composite lymphomas; MCL:Mantle cell lymphoma; DLBCL:Diffuse large B-cell lymphoma; PR:Partial response; CT:Computed tomography; IHC:Immunohistochemistry; IPI:International Prognostic Index; TCR:T cell receptor

Declarations

Availability of data and materials

Stained and unstained slides of the case can be provided if required.

Ethics approval and consent to participate

Not applicable.

Consent for publication

We have solicited the patient's agreement before writing this case report. We confirm that written informed consent has been provided by the patient to have the case details and images published.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

The report was designed, written, and reviewed by YJ and ZZ. All authors contributed to the data collection, data analysis, and interpretation. The manuscript was approved by all authors.

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Figures

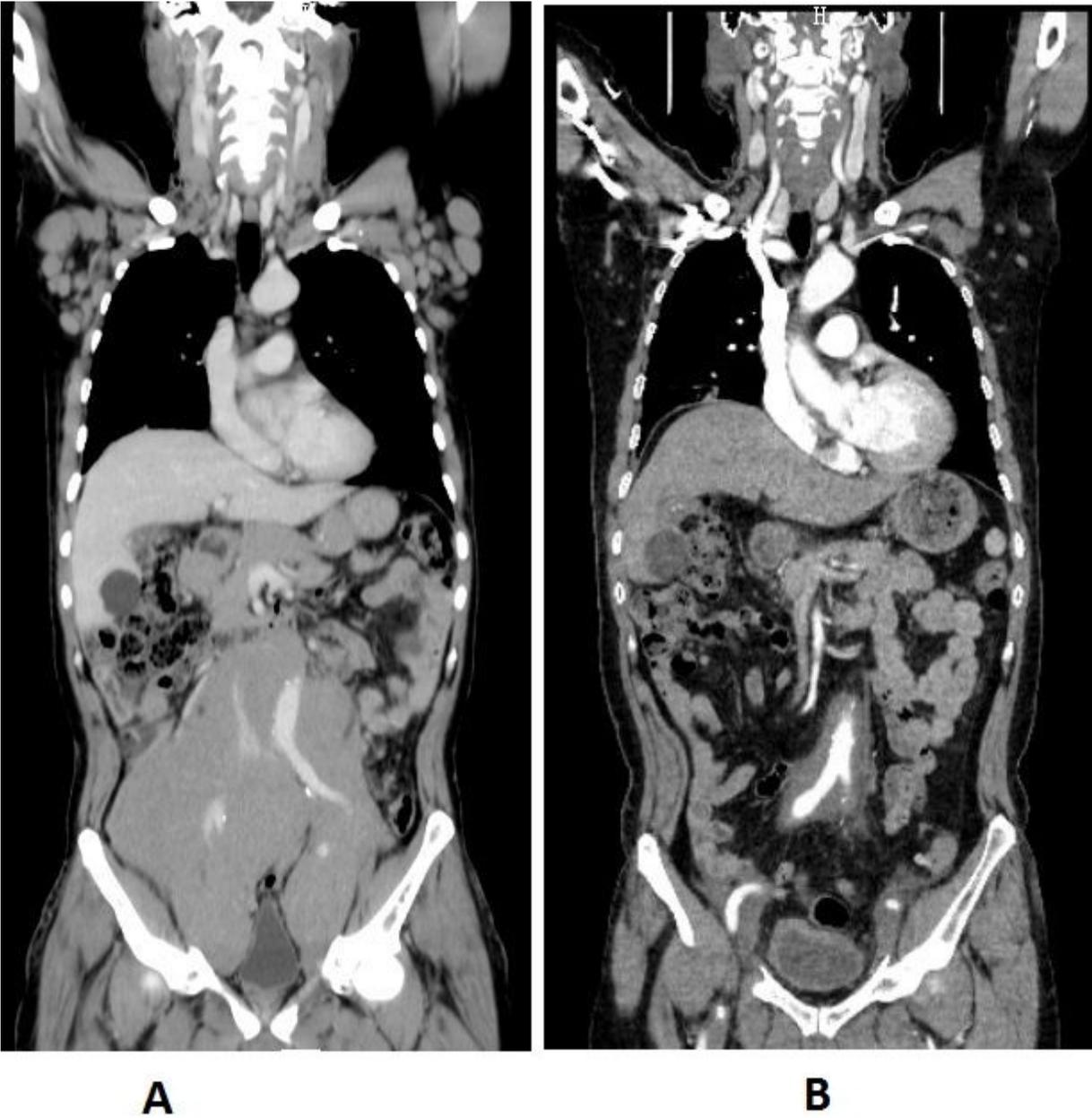


Figure 1

Abdominal Computed tomography images of the patient indicated the patient achieved partial response receiving chemotherapy without rituximab. A. Before treatment. The lymph nodes in the neck, chest, axilla, abdomen, pelvis, groin were swollen, and the gastric wall of the antrum was thickened in line with lymphoma swollen. The posterior ventral mass surrounded the abdominal aorta, inferior vena cava and bilateral iliac vessels. B. After chemotherapy. The original enlarged lymph nodes and masses were significantly smaller than before.

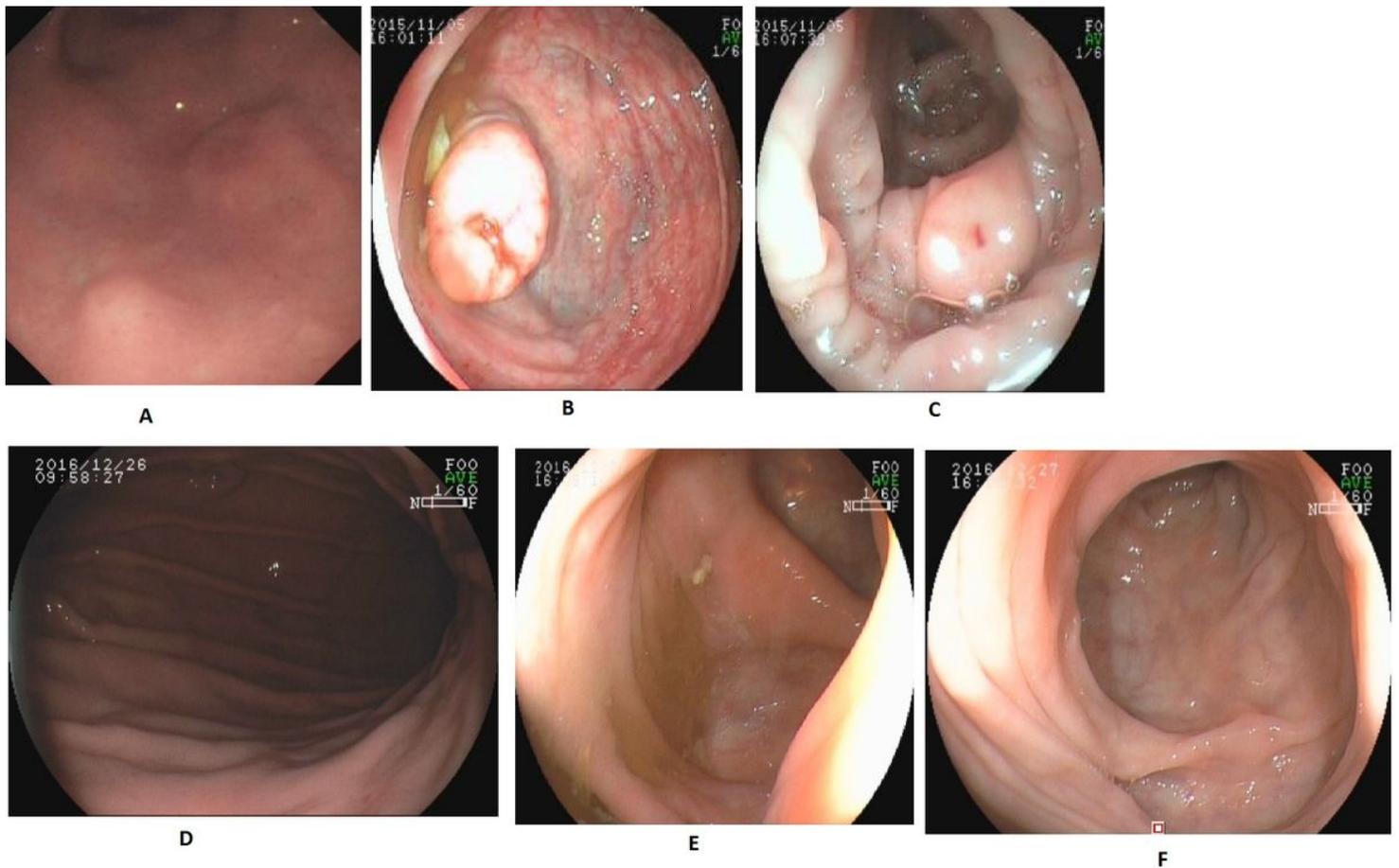


Figure 2

Electronic gastroscope and colonoscopy images before and after treatment. A. Nodular ridges were observed on the large curved side and the posterior wall of the middle part of the corpus, one of which was about 2.0cm×1.8cm. B. A mass was seen in the cecum, about 0.8cmX1.0cm in size, and the surface is erosive. C. A mucosal bulge was visible in the descending colon, the surface is smashed. D, E, F. After treatment, the masses in the corpus, ileocecal and descending colons almost disappeared.



Figure 3

A. After one cycle chemotherapy regimen, red rashes and blisters can be found on the patient's right lower extremity, without pain and itching. B. After six cycles of chemotherapy, some of the rashes were dissipated and no obvious new rashes were observed.

Supplementary Files

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