CASE REPORT

Richter's Syndrome (Diffuse Large B Cell Lymphoma) : Case Report

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BACKGROUND

Richter's syndrome (RS) is a complication and disease transformation of chronic lymphocytic leukaemia (CLL)into a more aggresive lymphoma, where clinical features, laboratory findings and treatment clearly differ from that of CLL.[1] Approximately 2-9% of patients with CLL progress to RS. The transformation rate is 0,5-1% per year.[2,3] The Median time from the diagnosis of CLL to transformation has been in the range 2 to 4 years. RS is rapidly progressive disease with a median survival of months.[1,3]

CASE REPORT

A 34-years-old male was referred by a surgeon in February 2018 with complaints neck swelling for 2 months and abdominal swelling for 1 months,weight loss and night sweats. His complete blood count showed leucocytosis (WBC of 185.000/dl) with mature like lymphocyte was 85%. Abdominal ultrasound scanning confirmed hepatosplenomegaly, multiple intra-abdominal lymphadenopathy paraaortici, and multiple nodules in spleen.Thorax X-ray confirmed noduler lession in paracardial dextra et causa pulmonal metastase. Bone marrow aspiration showed

hypercellular marrow with heavy infiltration of abnormally large lymphoid cells having abundant basophilic cytoplasm confirmed Chronic Lymphocytic Leucemia . A few smudge cells were also present .The biopsy result showed Diffuse Large B cell Lymphoma, CD 20 + . The patient was put on a 21-day-cycle RCHOP chemotherapy for six cycles. After six cycles chemotheapy, there was 90% regression in the size of the masses(the cervical lymphadenopathy was gone, no abdominal swelling, no splenomegaly, no lung metastase nor parailica lymphonodes but still small multiple nodules in spleen). The laboratory investigations revealed normal. We will plan second line chemotherapy with R-ICE regiment.

DISCUSSION

Approximately 2-9% of CLL transforms into lymphoma high grade called Richter's syndrome.The most frequent histologic type is DLBCL, which has the same clonal origin as CLL in 50% of the cases. It corresponds to diffuse proliferation of neoplastic large B cell lymphoid cell (CD20+,CD 79A+). Richter's syndrome is a very difficult disease to treat and the prognosis is said to be very poor.

CONCLUSION

Richter's syndrome occurs in about 5% of patients with CLL and its clinical outcome is poor. Extranodal involvements of RS may be present in up to 40% of patients and central nervous system manifestation was found to be the most common. People with RS develop aggressive disease with rapidly enlarging lymph nodes, enlargement of the spleen and liver, and elevated levels of a marker in the blood known as serum lactate dehydrogenase, or LDH. The improvement of survival in RS patients is achievable with intensive anti-lymphoma chemotherapy and subsequent allogeneic stem cell transplantation (alloHSCT) performed in complete remission.

Key Words

Chronic lymphocytic leukemia; Diffuse Large B Cell Leukemia; Richter's syndrome transformation

A Rare Case of A young Male with Richter's Syndrome: Diagnostic Aspects and Treatment

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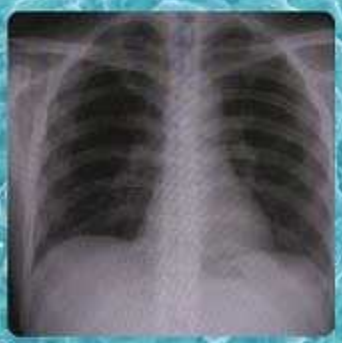
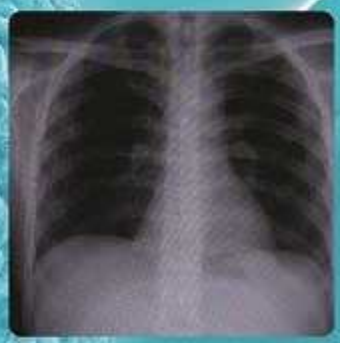
CASE REPORT

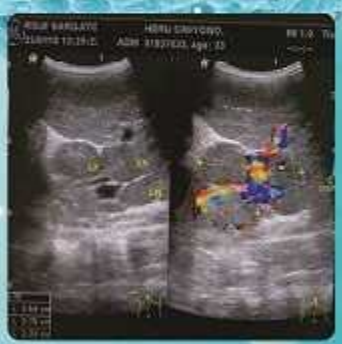
A 34-years-old male was referred by a surgeon in February 2018 with major complaints of neck and abdominal enlargement, weight loss and night sweats for the last 2 months. His complete blood count showed leucocytosis (WBC of 185.000/dl) with mature like lymphocyte of 85%. Abdominal ultrasound confirmed hepatosplenomegaly, multiple intra-abdominal paraaortic lymphadenopathy, and multiple nodules in the spleen.Thorax X-ray confirmed noduler lession in paracardial dextra and sinistra suspected pulmonal metastase. Bone marrow aspiration showed hypercellular marrow with heavy infiltration of abnormally large lymphoid cells with abundant basophilic cytoplasm confirmed the diagnosis of CLL. The biopsy of neck lymphonode showed Diffuse Large B Cell Lymphoma(DLBCL) and CD 20 +. All these data confirmed a diagnosis of Richter's Syndrome, i.e transformation from CLL to DLBCL. The patients underwent chemotherapy with R-CHOP three weekly for six cycles. Clinical and images evaluations showed 90% regression of the systemic nodal and the remaining lesions was only small localized nodal in the spleen. The blood test showed normal result.



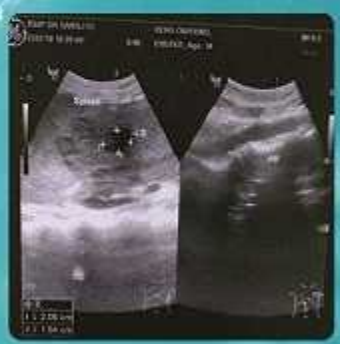
Before chemotherapy



After six cycles RCHOP chemotherapy



Before chemotherapy



After six cycles RCHOP chemotherapy

DISCUSSION

The most frequent histologic type is DLBCL, which has the same clonal origin as CLL in 50% of the cases.[1] It corresponds to diffuse proliferation of neoplastic large B cell lymphoid cell (CD20+,CD 79A+).[3] Richter's syndrome is a very difficult disease to treat and the prognosis is poor. The improvement of survival in RS patients is achievable with intensive anti-lymphoma chemotherapy and subsequent allogeneic stem cell transplantation (alloHSCT) performed in complete remission.[2]

CONCLUSION

We presented a rare case of Richter's Syndrome occur in a young male. A succesfull treatment with six cycles of R-CHOP was proved by 90% of systemic nodal masses regression leaving a localized small nodules in the spleen. Close monitoring is required to prevent relapse.

KEY WORDS

Chronic Lymphocytic Leukemia; Diffuse Large B Cell Leukemia; Richter's Syndrome

REFERRENCE

1. Matolcsy A, Inghirami G, Knowles DM: Molecular genetic demonstration of the diverse evolution of Richter's syndrome (chronic lymphocytic leukaemia and subsequent large cell lymphoma). Blood 1994; 83: 1363–1372.
2. Tsimberidou AM, Keating MJ: Richter syndrome: biology, incidence, and therapeutic strategies. Cancer 2005; 103: 216–228.
3. Tsimberidou AM, Keating MJ: Richter's transformation in chronic lymphocytic leukemia. Semin Oncol 2006; 33: 250–256.