



Evans' Syndrome in Hodgkin's Lymphoma: A case report

Mona M. Taalab¹

¹Oncology center Mansoura University (OCMU), Mansoura city, Egypt.



Abstract

Autoimmune phenomenon has been described in lymphoproliferative disorders; however, rarely seen in patients with Hodgkin's lymphoma (HL). Autoimmune hemolytic anemia and thrombocytopenia are infrequently observed in HL at time of diagnosis or relapse. We hereby report a case of a classic HL presented with Evan's syndrome at time of relapse.

background

Hodgkin's lymphoma is a type of malignancy derived from germinal center B –lymphocytes. HL is divided into two main variants: classic Hodgkin's lymphoma (CHL) which constitutes approximately 95% of cases, and rare nodular lymphocytic predominance (NLPHL) subtype¹. Supradiaphragmatic lymphadenopathy are commonly involved; cervical, anterior mediastinal, supraclavicular, and axillary lymph nodes involvement, while the inguinal areas are less frequently seen. Systemic symptoms occur in about one third of the cases HL. Extra nodal presentation is unusual. The most common sites to be involved are the spleen, liver, lungs, pleura, and bone marrow². Several autoimmune disorders have been observed in lymphatic malignancies, including autoimmune hemolytic anemia and immune thrombocytopenia; they frequently complicate the course of NHL though seldom occur in HL. Few case reports have described the association of HL and autoimmune abnormalities³ particularly Evans' syndrome.

Objectives

We report a case of relapsed classic HL stage IIB co-presented with Evans' syndrome. We describe the clinical course and the treatment modalities that led to satisfactory outcome.

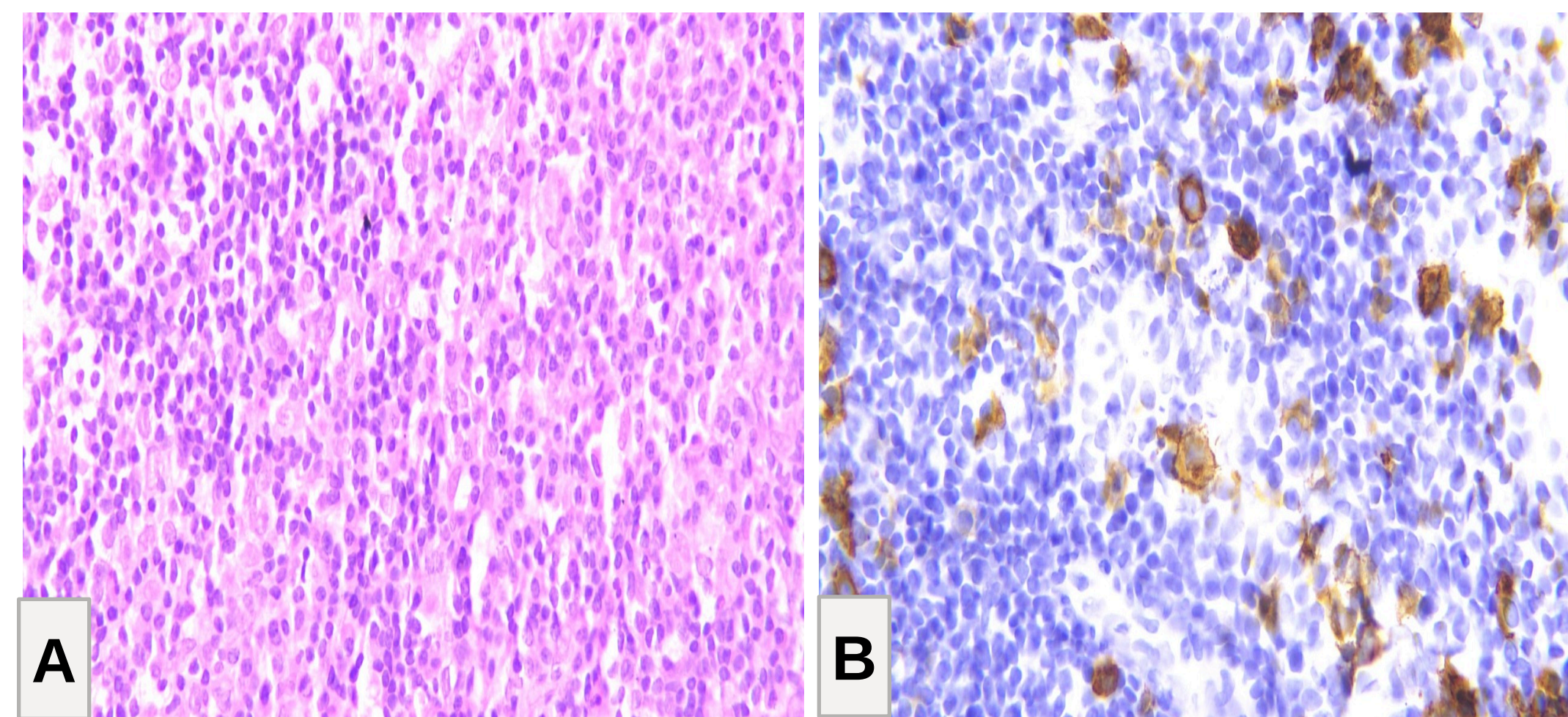
Case report

CHL patient was on follow up til 11/2020 when she complained of right cervical swelling that showed on examination: Right upper anterior cervical LN 3x3 cm, non tender and firm in consistency. Lymph node biopsy revealed classic HL, IHC showed positive CD15 and CD30.A positron emission tomography (PET) scan showed malignant lymphadenopathy in the cervical region. Bone marrow examination was free of lymphomatous deposits. CBC:TLC: 5.4, Hb 4.0 g/dl, MCV 90,3 fl, reticulocytic count 25%, platelets count 7x10⁹/L, T.bilirubin 5.9 mg/dl, direct bilirubin 0.9 mg/dl, direct Coombs' test: positive, LDH :1630 u/L. Our patient received high dose Dexamethasone 40 mg daily for 4 days, repeated every 2 weeks for 4 cycles plus Rituximab 500 mg weekly for 4 wks. CBC at end of rituximab therapy: TLC: 6.8x10⁹/l, Hb: 11 g/dl MCV: 81.89 fl, platelets count: 282 x10⁹/total bilirubin 0.8 mg/dl, direct bilirubin: 0.2 mg/dl. The patient received salvage chemotherapy; modified DHAP (dexamethasone, Cytarabine, cisplatin) every 3 weeks for 2 cycles. CBC: TLC: 5.5 x10⁹/l. Hb: 11.9 g/dl MCV:84 fl, platelets count:132x10⁹/l.

Follow up PET/CT scan was negative for residual viable lymphomatous lesion. Bone marrow illustrated 80% cellularity with no abnormal deposits. Patient still in remission while the article is written and is scheduled for consolidation by autologous stem cell transplantation.

Summary

A 25 year old woman diagnosed with classic HL, stage IIB, unfavorable. She was treated with 6 cycles of ABVD (D1,D15 regimen plus involved site radiotherapy (ISRT) which induce complete remission. After 2 years of follow up she presented with cervical nodal recurrence of classic HL with concurrent development of Evans' syndrome. Patient was treated successfully with high dose dexamethasone plus weekly 4 weeks Rituximab followed by 2 two cycles salvage therapy with modified DHAP. Patient achieved and continued in second complete response till the article is written.



A: Lymph nodes showed large atypical cells with large vesicular nucleoli on background of lymphocytes, plasma cells and eosinophils. **B:** Photomicrograph showed Reed sternberg cells stained strongly positive for CD15 with evident Golgi dot staining.

Discussion

Among the postulated mechanisms of AIHA in HL is that the antibodies are directly produced by the tumour cells or are related to an immune regulatory phenomenon⁴. The antibody associated with AIHA in some patients with HL has been identified as an anti transition (It) antibody⁴. Patients with HL are known to have an impaired cell mediated immune response due to decrease in number as well as function of T lymphocytes. Decreased number of cytotoxic T cells could lead to excessive autoantibody production. This appears to be partly due to hyper activation of B-cells⁵. Our case raised concern that Hodgkin's lymphoma should be ruled out in autoimmune disorders in particular the refractory and resistant cases.

References

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