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Follicular Lymphoma presenting as B-ALL - A aggressive tumour with poor prognosis, in a 4 yr old child

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Abstract
There is mounting evidence that leukemic presentation portends a worse prognosis in patients with FL in leukemic phase.The presence of circulating lymphoma cells in FL is a rare event and is associated with shorter PFS Median progression-free survival independently of FLIPI score and β2-m level. It can be safely said that circulating lymphoma cells >4.10^9/L have a poorer outcome. Although ~1/3 of the patients experience long term PFS, these should be monitored carefully during and after first line treatment to consider HSCT. Follicular Lymphoma presenting as B-ALL at diagnosis is a independent risk factor & has a poor prognostic outcome because it is highly aggressive tumour. The very rare nature of this entity & its grave prognosis merits its reporting.

Keywords: follicular lymphoma, ALL, leukemic phase, survival rate, prognosis

Introduction
Follicular lymphoma (FL) is a lymphoproliferative neoplasm of follicle center B-cells, which have at least a partially identifiable follicular pattern. FL is the second most frequent type of B-cell lymphoma and accounts for approximately 25–30% of all the cases of non-Hodgkin lymphomas (NHL) among adults [1].
The Follicular Lymphoma International Prognostic Index (FLIPI) are commonly used to predict patients outcome, Follicular Lymphoma presenting as B-ALL at diagnosis is a independent risk factor& has a poor prognostic outcome [2]

Case report
A 4 years old girl presented with painless generalized lymphadenopathy & fever.
On examination she had pallor, pleural effusion & abdominal distension. CT scan showed cervical, axillary, mediastinal, para-aortic, iliac, and inguinal lymphadenopathy. A biopsy of the cervical lymph node showed features of FL- grade 3. It was confirmed on immune-histochemistry.
CBC report: - WBC = 18.2 $x10^3$/ microL, HGB = 7.7 g/dl, MCV=74.1 fl, PLT =32 $x10^3$/ microL. LDH & B2 microglobulin were increased. LFT & RFT are normal.
Peripheral blood analysis revealed 74 % Blasts & 1% neutrophis with the remainder being lymphocytes. The Blasts were predominantly small sized cells with irregular cleaved nuclei & scant cytoplasm (Buttock cells, notched-nucleus cell"). Immunophenotyping by Flow cytometry favoured a diagnosis of B- ALL.

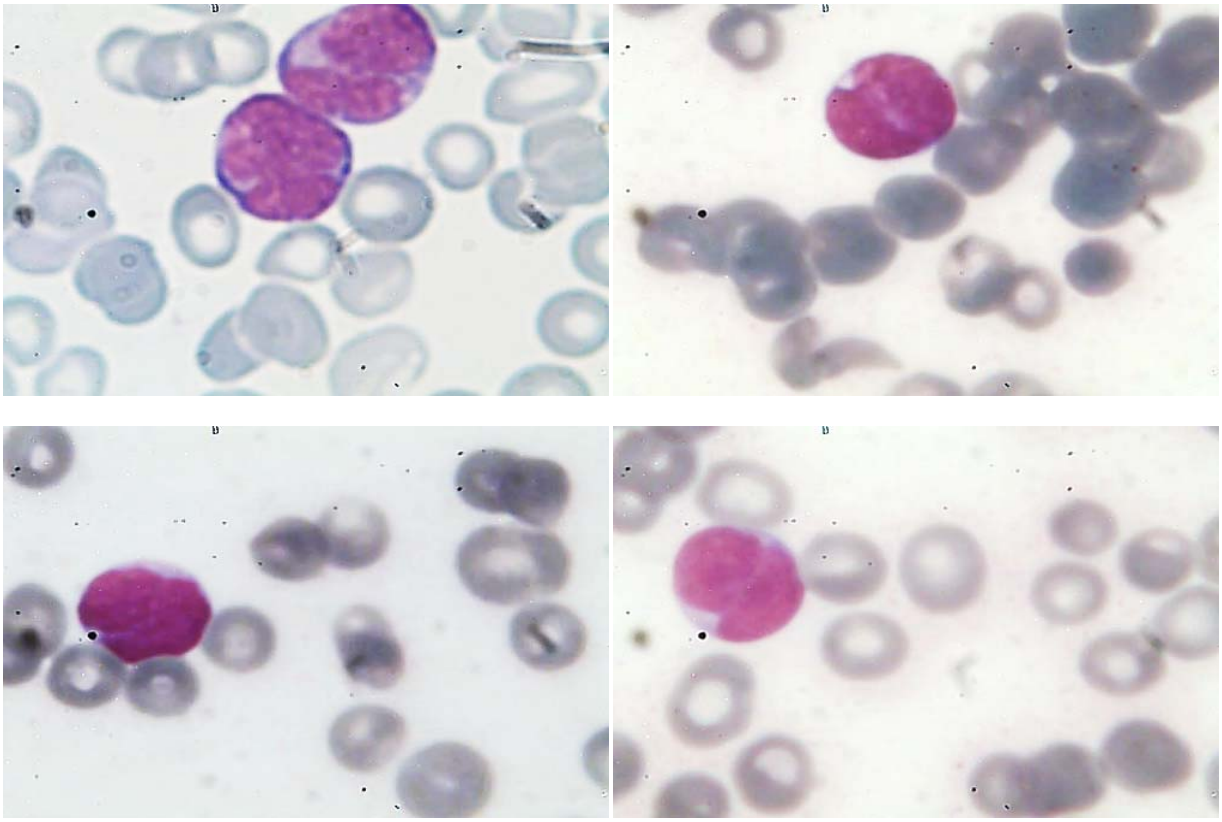
Discussion
Morphology of FL on histopathology. – shows at low magnification, a predominantly nodular or nodular and diffuse growth pattern in involved lymph nodes. Two principal cell types are present in varying proportions: (1) small cells with irregular or cleaved nuclear contours and scant cytoplasm, referred to as centrocytes (small cleaved cells); and (2) larger cells with open nuclear chromatin, several nucleoli, and modest amounts of cytoplasm, referred to as centroblasts. In most follicular lymphomas, small cleaved cells are in the majority [3] Similar findings were noted in this case from the cervical lymph node biopsy.
FL may evolve to or present with a leukemic component. In the leukemic form of FL, a variable proportion of the peripheral blood lymphocytes exhibit a distinct appearance that has been previously described as "notched-nucleus cell" or buttock cells with cleaved nucleus. The

lymphocyte count varies from moderately to severely elevated; in the present study, it ranged between 3,000 - 5,000. cells/mm [3]. The true incidence of such an event is unknown but varies in different series, ranging between (4–23%). A Japanese group reported leukemic phase at presentation in FL patients as high as (45%) [4]

In a recent report from Japan, Kodaira and colleagues showed that a leukemic presentation, which accounted for 21% of the cases, portends a worse progression-free survival (PFS) & more recently, Sarkozy and colleagues identified a leukemic phase in 7% of patients with FL, which was associated with a shorter PFS, independent of the FLIPI score and beta-2-microglobulin levels [3, 4]

Some unanswered questions remain

Should leukemic FL be diagnosed based on the solely presence of circulating FL cells? Or should a diagnosis of leukemic FL be rendered based on an actual number of circulating FL cells? Should all patients with a clinical diagnosis of FL undergo flowcytometry to detect a leukemic component? What is the best regimen to treat patients with leukemic FL? Should we recur to ASCT in specific cases?



(Buttock cells/ Notched nuclei cells in PBS)

Conclusion

The presence of circulating lymphoma cells in FL is a rare event and is associated with shorter PFS Median progression-free survival independently of FLIPI score and β2-m level. It can be safely said that circulating lymphoma cells >4.10^9/L have a poorer outcome. Although ~1/3 of the patients experience long term PFS, these should be monitored carefully during and after first line treatment to consider HSCT

Follicular Lymphoma presenting as B-ALL at diagnosis is a independent risk factor & has a poor prognostic outcome because it is highly aggressive tumour.

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