Chronic Lymphocytic Leukaemia Complicated by Cranial Diabetes Insipidus - A Case Report

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ABSTRACT
Here we describe a case of 55-year-old lady who was admitted in hospital for evaluation of recurrent anaemia, polyuria and polydipsia with history of splenectomy 9 months back. Physical examination revealed anaemia, dehydration and scar mark of splenectomy. Initial laboratory tests were suggestive of chronic lymphocytic leukaemia (CLL) and further bone marrow examination & immunophenotyping confirmed the diagnosis. At the same time Polyuria & polydipsia was evaluated by water deprivation test and diabetes insipidus was diagnosed.

Keywords: Chronic lymphocytic leukaemia; Polyuria; Polydipsia; Diabetes Insipidus; Leukaemic infiltration; Water deprivation test.

Introduction
Chronic lymphocytic leukaemia (CLL) is a malignant tumour which arises from mature B lymphocytes and involves primarily the blood, bone marrow & lymphoid organs such as the lymph nodes and spleen. Central nervous system (CNS) involvement is not rare in acute leukaemia, particularly in acute lymphoblastic leukaemia (ALL).¹ But it is quite rare in chronic lymphocytic leukaemia. Diabetes insipidus (DI) is an unusual clinical manifestation of CNS involvement and occurs due to leukemic infiltration to the hypothalamus or neurohypophysis.² Our aim is to present a 55-year lady with chronic lymphocytic leukaemia with cranial diabetes insipidus with Hb. E trait which is a rare case.

Case Presentation
55-year old lady was presented with recurrent anaemia and huge splenomegaly about 9 months back. Splenectomy was done due to mechanical problem. This time patient again presented with anaemia, polyuria and polydipsia. On examination - patient was moderately anaemic and dehydrated. Pulse: 82 beats/min, blood pressure: 120/80 mm of Hg, temperature 98°F and respiratory rate 16 breaths/min. Patient was non-icteric, no lymphadenopathy or organomegaly present. Scar mark of splenectomy is present.

Investigation reveals Haemoglobin-6.9 gm/dl, WBC: 46×10⁹/L, Neutrophil 21%, Lymphocyte: 69%, Monocyte: 10%, Platelet: 250×10⁹/L, ESR: 20 mm in
1st hour. PBF shows Lymphocytosis possibly CLL (monotonous looking mature lymphocytes with presence of smudge cells) with possible immune haemolytic anaemia (plenty of nRBC, spherocytes, polychromatic cells & target cells). Reticulocyte count: 1.41%. Bone marrow with immunophenotyping: Suggestive of CLL (CD5+, CD19+, CD20+, CD23+, SMiG weak+, FMC7+, CD200+, CD79b+). In the meantime, polyuria and polydipsia was evaluated. Water deprivation test revealed cranial diabetes insipidus. MRI of brain showed mild thickening and enhancement of the pituitary stalk and tuber cinereum representing leukemic infiltration. But due to limitation, tissue diagnosis was not possible. As treatment criteria of CLL was met, treatment was given according to BR protocol (Bendamustine and Rituximab). Diabetes insipidus was treated with intranasal DDAVP (Desmopressin). After starting treatment, patient improved symptomatically & her polyuria & polydipsia was also improved. Patient is now on regular follow up.

Discussion
Diabetes insipidus is an uncommon disorder characterized by the persistent excretion of excessive quantities of urine and by thirst. It is classified into two types-cranial diabetes insipidus and nephrogenic diabetes insipidus. Cranial diabetes insipidus may be idiopathic or due to structural hypothalamic or high stalk lesion or due to genetic defect. It is a rare complication of leukaemia particularly it is very rarely seen in case of CLL.

The most marked symptoms are polyuria and polydipsia. The patient may pass 5-20 L or more of urine in 24 hrs. Urine is of low specific gravity and osmolality. Diabetes insipidus can be confirmed if serum ADH is undetectable or the urine is not maximally concentrated. water deprivation test is widely used. Assessment of other pituitary hormones & imaging of pituitary gland should also be done in case of cranial diabetes insipidus. Several reports showed a failure of the antileukemic therapy to alleviate the symptoms of DI in patients with leukaemia. This feature reflects that cranial DI associated with leukaemia results from destruction of cells rather than their dysfunction. Thus, the most useful therapy is standard hormonal replacement, although 15% of the patients are resistant to DDAVP therapy.

Conclusion
Although rare leukaemic infiltration of the pituitary gland should be evaluated in leukaemic patients presenting with visual disturbance, hypopituitarism or cranial diabetes insipidus. The diagnosis should be established, and appropriate treatment must be given immediately due to potential serious complications if left untreated.

References