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RESEARCH ARTICLE

A RARE CASE OF HAIRY CELL LEUKEMIA IN A15-YEAR-OLD BOY

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ABSTRACT

Hairy-cell leukemia (HCL) is a rare slow-growing mature B cell neoplasm of the blood (2% of lymphoid leukaemias) (1). The median average age lies between 50 to 55 years. Can occur at any age from 20-80 years (1). Children are very rarely affected (1). Male to female ratio is 5:1(2). Diagnosis of HCL is based on clinical symptoms, morphologic findings in peripheral blood and bone marrow, flowcytometric immunophenotyping of blood cells and cytogenetics studies.

INTRODUCTION

Hairy-cell leukemia (HCL) is a rare slow-growing mature B cell neoplasm of the blood(2%of lymphoid leukaemias) (Madeleen, 2018). The median average age lies between 50 to 55 years. Can occur at any age from 20-80years (Madeleen, 2018). Children are very rarely affected (Madeleen, 2018). Male to female ratio is 5:1 (Dores *et al*., 2018). Diagnosis of HCL is based on clinical symptoms, morphologic findingsin peripheral blood and bone marrow, flowcytometric immunophenotyping of blood cells and cytogenetics studies.

CASE REPORT

A 15 - year - old male was admitted to a tertiary care hospital, with the history of fever of 4 days. He was previously healthy. No family history of malignancy. Physical examination revealed right cervical lymph nodes of 1x1 cm in the posterior tangle and a few ecchymotic patches on the arm and massive splenomegaly. A pancytopenia on the FBC noted. Bio chemistry was normal, viral studies and the monospot test was negative. No coagulopathy was observed. CT abdomen revealed massively enlarged spleen (25x10 cm), hepatomegaly (20 cm in MCL), homogenously enlarged celiac and superior mesenteric lymph nodes. Ultrasound scan of neck. revealed multiple bilateral level III and 1V cervical lymph nodes. Blood picture confirmed pancytopenia.

Few atypical lymphoid cells characterized by medium to large cells with villous projections were noted in the blood picture. 60% of atypical lymphoid cells with hairy projections suggestive of hairy cell leukaemia were seen in the bone marrow aspirate The trephine biopsy architecture was characteristic. The immunophenotypic results of "BCLPD" and "BCLPD HAIRY" panels showed bright positivity of CD19, CD20, FMC7, CD103, CD123, CD 11c, CD200, smIg Kappa and positive CD 25. Negative results were CD5, CD10, CD79b and CD43. Cytogenetics revealed positive BRAF – V600 E mutation.

DISCUSSION

Hairy cell leukemia (HCL) is a rare type of blood cancer occurring in B lymphocytes. The uncontrolled proliferation of these cells result in (pancytopenia). Affected individuals usually exhibit fatigue, weakness, fever, weight loss, and/or abdominal discomfort due to massive splenomegaly, hepatomegaly, increasedsusceptibility to bruising and/or severe infection. The aetiologyof hairy cell leukemia is not known.The pathogenesis of HCL was determined by the discovery of its underlying genetic cause, the BRAF-V600E kinase-activating mutation, which is somatically and clonally present in almost all patients through the entire disease spectrum and clinical course. [(3)] By aberrantly activating the RAF-MEK-ERK signaling pathway, BRAF-V600E shapes key biologic features of HCL, including its specific expression

signature, hairy morphology, and anti-apoptotic behavior. Accompanying mutations of the KLF2 transcription factor or the CDKN1B/p27 cell cycle inhibitor are recurrent in 16% of patients with HCL and likely cooperate with BRAF-V600E in HCL pathogenesis (Emmanuel, 2008). HCL behaves like a chronic leukemia. With treatment, most patients achieve clinical remissions and, sometimes, long-term cures. In the United States, 5-year survival ranged from 84% to 94% (lower in blacks than whites) (Emmanuel, 2008). The risk of second malignancies has been observed in patients with HCL due to the disease itself or secondary to the immunosuppressive effects of the therapy. Skin cancers (melanoma and non-melanoma) are the most common, representing 33-36% of all secondary malignancies. Other malignancies include prostate cancers, gastrointestinal malignancies, non-Hodgkin lymphomas, and ovarian, cervical, and breast cancer (Emmanuel, 2008).

Conclusion

The findings were consistent with a diagnosis of Hairy cell Leukaemia

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