

Case Report-II

Father and Son with Hodgkin's Disease – A Case Report

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INTRODUCTION

Hodgkin's Disease is a common neoplasm among young adults. However, familial Hodgkin's disease is rare and only few cases have been reported in the literature.^{1,2} Here we report a case of familial Hodgkin's disease, both father and son were affected.

CASE

DG, a 9 year old male child presented with cervical lymphadenopathy in April 2003. There was no history of 'B' symptoms. Examination revealed - bilateral cervical lymphadenopathy with right-sided adenopathy being more compared to the left. There was no axillary or inguinal lymphadenopathy. Per abdomen- no hepatosplenomegaly. Investigations : Blood – Hb - 13.1 Gm%, WBC – 8,250/mm³, DLC- N 54%, L46%. Platelet count – 3,22,000/mm³. Renal and liver functions were within normal limits. LDH - 549 U/L. Chest X-ray - anterior mediastinal widening. CT scan Chest revealed: anterior mediastinal adenopathy and spleen enlarged with multiple focal lesions of varying sizes. Supraclavicular lymph node biopsy revealed : Hodgkin's disease (mixed cellularity). Bone marrow aspirate and biopsy did not reveal any involvement by disease. In view of Stage IIIA, S, he was advised ABVD chemotherapy.

His father was diagnosed to have Hodgkin's disease, stage III A in 1997. He was 28 years old then. He had pelvic and retroperitoneal lymphadenopathy, and

splenomegaly at diagnosis. Histopathology-revealed Hodgkin's disease(Mixed cellularity). Bone marrow was not involved. He received 6 cycles of ABVD and continues to be disease-free.

DISCUSSION

Familial Hodgkin's lymphoma (HL) is rare. Further, parent and child pair is even rarer. We are not aware of similar cases in Indian literature. Shugart et al³ found thirty parent-child pairs in literature with a well-documented diagnosis of HL. In all pairs with the exception of one, HL children were diagnosed at a younger age than their HL-affected parent. For example, the mean age at onset was 46 years in parents and 22 years in children. This significant difference between the age at diagnosis of parents and that of children was detected by use of the Mann-Whitney test ($N=30$, $U=40.5$, $P<.0001$). The Swiss Lymphoma Registry has reported 13 cases of parent child Hodgkin's disease.⁴

In the present parent and child pair the father presented at an age of 28 years and the child presented at an age of 9 years. Since Epstein-Barr virus (EBV) has been implicated in the etiology of familial HL, it is therefore possible that the occurrence of HL could be related to simultaneous parent-child exposure to this viral infection. In the present case EBV was negative in the child and we could not ascertain the EBV status of the parent at the time of original presentation.

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The resistance and susceptibility to HL has been linked to HLA class II alleles. Bodmer et al⁵ reported observation of an association of DPB1*0201 with resistance to HL, and DPB1*0301 with susceptibility to HL. This has been further corroborated in the study by Taylor et al.⁶ One potential explanation for the association of DPB1 alleles with susceptibility and resistance to HL is that they are in linkage disequilibrium (LD) with more strongly associated DRB1 alleles. Taylor et al⁶ found that DRB1*1501 was associated with susceptibility and 0101 with resistance. Unfortunately we could not look for DPB1 alleles in the present case pair.

It is clear that knowledge of anticipation in HL and other disorders could lead to important insights into their etiology and pathogenesis.⁷ For example, if the EBV theory regarding HL is correct, then exceedingly early treatment/vaccine in the progeny of an exposed parent, if and when such treatment becomes available, could conceivably lead to prevention of HL in that individual's progeny. Just as early age of onset of cancer is one of the cardinal features of most forms of hereditary solid tumours, where anticipation may or may not be present, it would be important in familial cases of HL to focus attention on members of the succeeding generations of those affected by HL (HL-affecteds). If therapy and/or other possible means to prevent HL become available in the future, the target population in familial HL would be the progeny of HL-affecteds with

particular attention being given to the earlier age of onset of HL in this high-risk group. Such HL patients/families showing anticipation will be ideal candidates for researching the etiologic and pathogenetic significance of this putative biologic phenomenon.⁸

In conclusion we would like to emphasize that Hodgkin disease occurring in parent-child pairs is rare. The phenomenon of anticipation in such pairs is known and the present case also demonstrates the same.

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