Case Report

Castlemans Disease in HIV Infected Patient from Eastern India

D Modak*, RR Ganguly**, SN Haldar**, PS Samanta***, N Pramanik****, SK Guha+

Abstract

Castlemans disease is a rare lymphoproliferative disorder. We report a patient of Castlemans disease with advanced HIV infection who responded well to conventional HAART. This case is interesting because of the rarity of the disease in the eastern part of India and its good outcome with antiretroviral therapy.

INTRODUCTION

Castlemans disease is a non-cancerous neoplastic proliferation of lymph node. It remains a clinicopathologic diagnosis. In 1956, Benjamin Castleman & his colleagues first described it. The synonyms of Castlemans disease are (a) Hamartoma of lymphatic, (b) Giant lymph node hyperplasia, (c) Angiofollicular lymph node hyperplasia, (d) Angiomatous lymph node, (e) Giant benign lymphoma.

The disease is classified histologically into three varieties and clinically into two types.

1) Hyaline vascular type- it is most common variety (90 %.). There is localized single or regional lymph node hyperplasia & symptoms are mainly due to pressure effect of enlarged lymph node. Commonly affected sites are abdomen, mediastinum and rarely peripheral lymph nodes. They are completely curable either by surgical excision or by localized radiotherapy.

2) Plasma cell type – it is less common & systemic symptoms are present. Common sites of involvement are mediastinum, abdomen, neck, axilla and other areas. Plasma cell type usually associated with fever, significant weight loss, anemia, skin rash, organomegaly and hypergammaglobulinemia. Some times mixed variety may be present. Plasma cell type frequently changes into multicentric type.

3) Multicentric disease (MCD) is a special type of castlemans disease, mainly associated with Human herpes virus-8 (HHV-8) or Kaposi sarcoma herpes virus (KSHV). Multicentric disease is associated with HIV infection. HHV-8 is a gamma virus that produces Kaposi sarcoma & primary effusion lymphoma. There is hyper- proliferation of B cells that releases cytokines, mainly IL-6 which interact with other interleukins and TNF and is responsible for B symptoms like weight loss, fever and night sweat. This variety frequently turns into B-cell lymphomas; sometimes it is associated with polyadenopathy, organomegaly, endocrinopathy, monoclonal antibody and rash (POEMS). Multicentric type has a poor prognosis. This type is commonly seen in HIV infected one.

CASE REPORT

A thirty-two years young male patient attended viral diseases OPD with chief complaints of fever, upper abdominal pain, nausea, extreme weakness of three and half month's duration. He also lost 6 Kg in last two months; He had past history of jaundice six months back. On examination, apart from pallor and tender hepatomegaly of 2 cm below costal margin, no other abnormality was detected. On routine examination Hemoglobin was 7.2 gm%, WBC-5500/cmm, Neutrophil-62%, Lymphocyte-29%, Eosinophil-9%, Monocyte-0 Basophil-0, Platelet-1.2 lacs / cmm, serum total bilirubin-0.8mg%, (conjugated-0.3mg%) serum albumin-2.4g/dl,serum globulin-5.7g/dl, SGPT-56 U/L, SGOT-32 U/L, Alkaline phosphatase-756 U/L. Blood urea-23mg%, creatinine-0.9mg%. HBsAg and anti-HCV- non reactive. On routine stool and urine examination no abnormality was detected. He was diagnosed as HIV positive by VCCTC. Chest x-ray was normal. Ultrasonography of abdomen revealed hepatomegaly with homogeneous echotexture, proximal part of common bile duct was distended, 0.8-1cm solitary lymph node at porta hepatis, and there was also enlarged mesenteric group of lymph nodes. MRCP (magnetic resonance cholangio-pancreatography) also revealed one large node at porta hepatitis obstructing common bile duct.

Exploratory laparotomy was done and biopsy was taken from different areas of mesenteric group of lymph node, node at porta hepatitis, and from liver.

Histopathological study revealed scattered lymphoid follicles some of which showed deposition of an amorphous...
materials, expanded interfollicular areas with large number of plasma cells, a few histiocytes and lymphocytes, scattered immunoblasts with marked vascular proliferations. Immunohistochemistry revealed that the plasma cell expressed CD-138 and were polytypic admixture of kappa and lambda expressing cells suggestive of plasma cell variant of castlemans disease.

As his CD4 count was 45/cmm, he was put on antiretroviral therapy (HAART) with Lamivudine, Stavudine and Nevirapine (3TC + d4T + NVP) in standard doses with co-trimoxazole prophylaxis. After two weeks of treatment fever subsided and he improved symptomatically. Six months later his CD4 became 212/cmm. Follow-up sonography did not show any lymph node. Even after one and half year of follow-up, he remains symptom free and his CD4 count has increased up to 453/cmm.

**Discussion**

In HIV infected patients, the plasma cell variety of Castlemans disease frequently turns into multicentric variety, which has a poor prognosis, as most of them changes into B cell type of non-Hodgkin’s lymphomas. They may be treated either with chemotherapy or radiotherapy or combined chemo and radiotherapy or splenectomy. Various clinical trials with intravenous/oral gancyclovir, anti-CD20 antibodies, thalidomide, anti interleukin -6 antibody, viral ablation have been done in different parts of the world. Splenectomy may be another option. Antiviral drugs like cidofovir, foscarnet do not have any response.

The number of HIV infected patient attending viral diseases OPD during 2003 – 2006 and the number of indoor admissions were 6024 and 1678 respectively. In more than seven hundred cases, we have done lymph node aspiration cytology and biopsy, if needed. However, this is the only case where Castlemans disease was diagnosed.

As HIV associated Castlemans disease has poorer prognosis and seen only in advanced HIV infection (CD4 <200 cells /cmm) we started HAART with two NRTI and one NNRTI in standard doses. We observed that only HAART could also improve life expectancy in this patient. Inhibition of HHV-8 replication and virokine release through the restoration of immunity by HAART was the basis of disappearance of clinical symptoms. Mean survival of MCD in HIV infected people are 46 (18-57) months. Most of the literatures are of the opinion that no specific type of treatment can prolong the life of the patient with Castlemans disease (MCD type). Our study subject is doing well even after 20 months of diagnosis and his CD4 count increasing significantly. In other parts of the world the reported MCD in HIV positive patient were mostly associated with Kaposi sarcoma. In eastern part of India Kaposi sarcoma is very rare in HIV and the study case did not have any evidence of Kaposi sarcoma. Further study is necessary whether early initiation of HAART can delay or prevent development of lymphomas in a HIV positive patient having Castlemans disease or improve quality of life.

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**References**