# MIXED HYALINE VASCULAR AND PLASMA CELL TYPE CASTLEMAN'S DISEASE: REPORT OF A CASE

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**Abstract-** Castleman's disease (angiofollicular lymphoid hyperplasia) includes a heterogeneous group of lymphoproliferative disorders. The cause of this disease remains uncertain. There are two types of localized Castleman's disease: the more common hyaline vascular and the plasma cell types. Mixed variant is an uncommon localized lesion in general population. The lesions can occur in any part of the body that contains lymphoid tissue, although seventy percent are found in the anterior mediastinum. We report a thirty years old boy with Castleman's disease who presented with fever, anorexia, weight loss, sweating, anemia and abdominal mass. The histologic examination of the biopsy specimens revealed a mixed hyaline vascular and plasma cell type of Castleman's disease.

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**Keywords**: Castleman's disease, angiofollicular, lymphoid, mixed Castleman's disease

## INTRODUCTION

Castleman's disease (angiofollicular lymphoid hyperplasia) includes a heterogeneous group of lymphoproliferative disorders. The most common variety is hyaline vascular. Plasma cell variety is rare and mixed type is the rarest form. The disease is not responsive to usual chemotherapy manipulation but is treatable by surgery even though the cause is incorporation of a viral genome into the cell of lymphoid organ.

Here we report a 13-years-old boy with Castleman's disease of mixed hyaline vascular and plasma cell type.

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## CASE REPORT

A 13-year-old boy was referred to our center with fever sweating, weight loss (7 kg), watery diarrhea and anorexia starting 8 months back. He had experienced vomiting and colicky pain in abdomen 2 weeks before admission. There was no history of previous disease. There is no contributory disease in her family and drug history was negative. Physical examination indexes are shown in table 1 and the laboratory values obtained during initial examination are mentioned in table 2. HIV and EBV virus serologic test were negative. HHV-8 antibody and Interleukin 6 antibody test were not done. Peripheral blood smear for malaria and borrelia were negative. Tuberculin test was 5 mm. Liver enzymes, thyroid function tests and urine analysis were normal and blood culture was negative. Bone marrow exam revealed erythroid hyperplasia combined with micro and macroblastic reaction (mixed anemia) and mild eosinophilia.

**Table 1.** Findings in physical examination of the patient

| Index | Measure or condition |
|---|---|
| Body Temperature (oral) | 38.5 ° C |
| Blood pressure | 100/70 mm Hg |
| Pulse rate | 100 beat/min |
| Respiratory rate | 15 /min |
| General observation | Cachectic |
| Lung and heart auscultation | Normal |
| Spleen palpation | Just palpable |
| Liver span in palpation | Normal |
| Abdominal Examination | Normal |

Multiple hypo-echoic masses in left paraaortic region and mild splenomegaly were evident in abdominal sonography. Upper GI endoscopy and colonoscopy were normal. Contrast enhanced computed tomogram, showed a lobular mass (34× 40 mm) near abdominal aorta and $L_3$ vertebrae. This mass compressed the adjacent colon (Fig. 1).

Enlarged lymph node, 4×5 mm in size and moderately enlarged spleen were found on exploratory laparotomy and the entire pathologic mass was removed.

**Table 2.** Findings in laboratory evaluation of the patient

| Index | Measure |
|---|---|
| WBC | 7900 cell/mm$^3$ (70% granulocyte) |
| Hemoglobin (Hb) | 8.9 gr/dl |
| Hematocrit (HCT) | 29.8% |
| MCV | 74.3 |
| MCH | 22.3 |
| MCHC | 29.2 |
| Platelet | 467000 |
| Retic count | 1% |
| ESR | 135 |
| CRP (Qualitative) | Positive |
| Iron (Free serum) | 25 mg/dl |
| Ferritin | 283 mg/dl |
| TIBC | 330 |

Abbreviations: MCV, mean cellular volume; MCH, mean cellular hemoglobin; ESR, erythrocyte sedimentation rate.

Pathologic report was compatible with Castleman's disease with mixed hyaline vascular and plasma cell type (Fig. 2).

After surgery patient recovered quickly, sweating reduced and anemia improved. After 2 month follow up, all clinical symptoms improved.

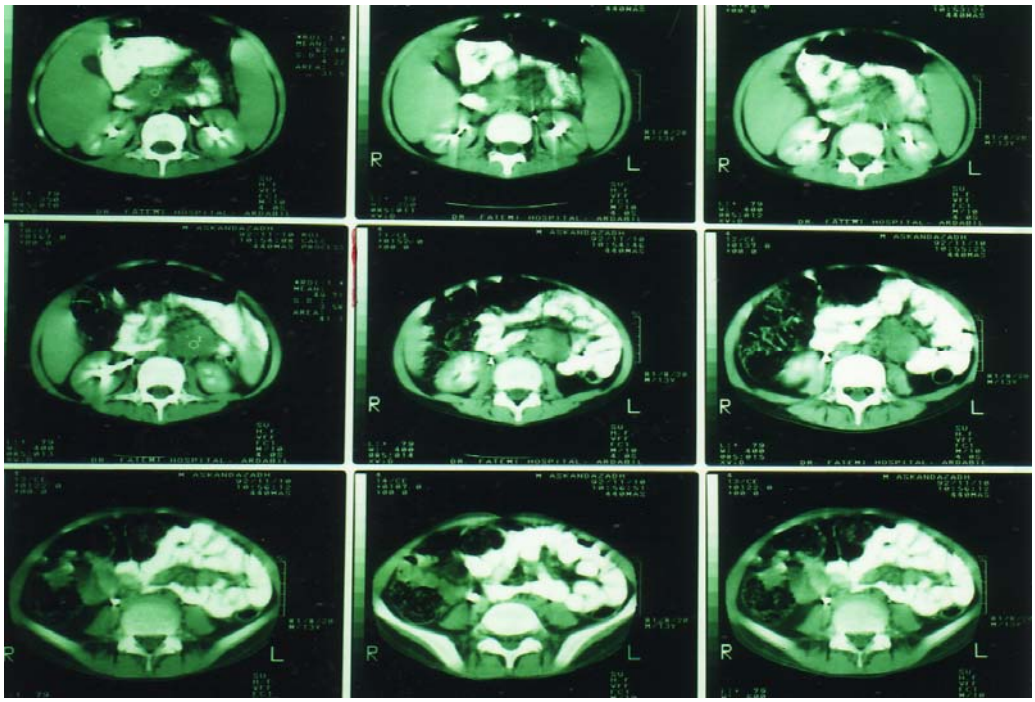

**Fig. 1.** Contrast enhanced computed tomogram of patient, showing a lobular mass near abdominal aorta and $L_3$ vertebrae.

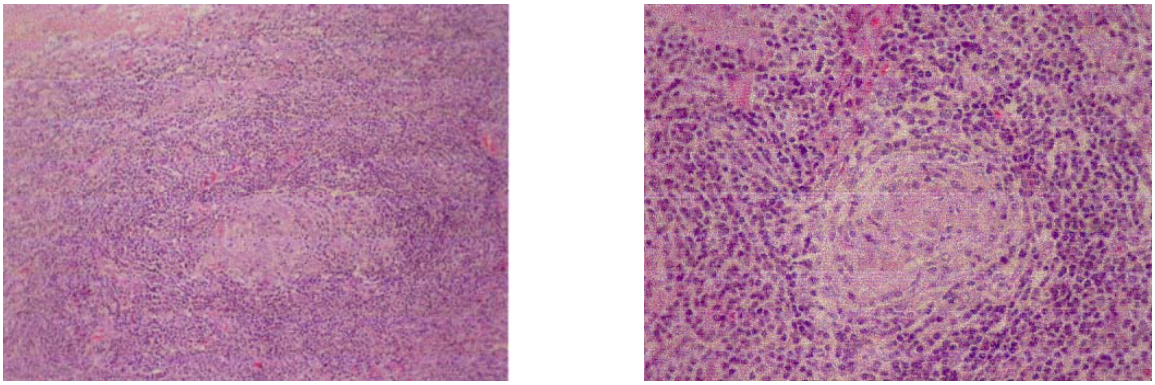

**Fig. 2.** Pathologic report of enlarged lymph node was compatible with Castleman's disease with mixed hyaline vascular and plasma cell type.

## DISCUSSION

In 1956, Castleman *et al.* described a group of patients with benign mediastinal lymphoid masses which they entitled mediastinal lymph node hyperplasia and named Castleman's disease. The pathophysiology of Castleman's disease was previously related to the deregulation of the immune system associated with Epstein-Barr virus infection (3). Later investigations showed that the etiology of the disease could be variable. Localized Castleman's disease, usually the plasma cell type, may be associated with HIV infection. Castleman's disease is now known to be nearly always associated with the presence of human herpes virus 8 (HHV-8)/Kaposi sarcoma-associated herpes virus (KSHV) (4). Also, the association of preceding or concurrent Castleman's disease with Kaposi sarcoma has been well documented (5). Therefore, the relationship among Castleman's disease, Kaposi sarcoma, and KSHV/HHV-8 brings new insights into the pathophysiology of this disease. Multicentric Castleman's disease is usually variable in etiology and generally not of plasma cell type. The most common type is the hyaline vascular type accounting for 90 percent of all lesions. Most patients are young (70% less than 30 years) (1). Another study mentioned no sex dependency and showed an age ranging between 8 to 69 years (median 33 years) in case studies (2). Pathologic processes represent a paracortical lymphoid architecture affected by a proliferation of post–capillary venule associated with lymphocyte immunoblasts and a variable number of plasma cells. The germinal centers are considerably affected. Vascular structures replace the core of the germinal center. Hyaline vascular type is benign and usually cured after extirpation.

The second form, or plasma cell variety, is associated with enormous germinal center hyperplasia and infiltration of the tissue with sheet of plasma cell in 50 percent of cases. The plasma cell variant of Castleman's disease may be localized or multicentric. Multicentric disease is a systemic lymphoproliferative disorder characterized by lymphadenopathy, hepatosplenomegaly and constitutional symptoms. Anemia, hypoalbuminemia, and hypergammaglobulinemia are also common in this type.

Interleukin-6, a cytokine with pleiotropic effects on the immune system, hematopoiesis, and acute-phase reactions, is a putative growth factor in multiple myeloma and may also be central to pathophysiology of Castleman's disease (3).

The transitional or mixed variant is an uncommon localized lesion. Patients are usually asymptomatic, and pathologic lesion resembles the hyaline vascular type except that there are foci of plasmacytosis and possibly some hyperplastic germinal centers (4-6). Arced interfollicular plasmacytosis with hyaline vascular germinal centers is even less common. In our case, surgical removal of abdominal mass, improved all clinical symptomatology. This is uncommon in mixed type, but usual in hyaline vascular type (6).

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