

Castleman's Disease- A Case Report

Md. Ismail Patwary¹, Md. Abed Hossain², Md. Matiur Rahman³, Md. Ashfaul Islam Chowdhury⁴, Md. Khalilur Rahman⁵, Md. Ruhul Kabir⁶

¹Professor & Head, Medicine Department; Sylhet MAG Osmani Medical College Hospital, ²Professor & Head, Pathology Department; Jalalabad Rgib-Rabeya Medical College, ³Associate Professor of Neurology, ⁴Assistant Professor of Medicine, S.O.M.C.H, ⁵Student of M.D.(Internal Medicine)-final part; S.O.M.C.H, I.M.O. Medicine Unit-1; S.O.M.C.H.

Abstract:

Castleman's disease (CD) is a heterogeneous group of lymphoproliferative disorders of uncertain cause presenting with lymphadenopathy. It is histologically and prognostically distinct from malignant lymph-node hyperplasia. It was first described in a group of patients with benign localised hyperplastic lymph-nodes in 1956 by Castleman et al. We report a case of a 70 year old gentleman who was clinically suspected to have lymphoma, but later histologically confirmed to have Castleman's disease.

Key words: Lymphadenopathy, Lymphoma, Castleman's disease.

[BSMMU J 2012; 5(1):79-80]

Introduction:

Castleman's disease (CD) is a lymphoproliferative disorder which is histologically characterised by angiofollicular lymph-node hypertrophy.¹ It may be borne in mind in the differential diagnoses of localised/diffuse lymphadenopathy with or without systemic manifestations. This is rather rare and relatively benign disorder which, though mimicking lymphoma clinically, varies from the latter histologically, prognostically and in its treatment options.

Case Report:

A 70-year-old, smoker, normo-tensive, non-diabetic male got admitted into Medicine ward, Sylhet M.A.G Osmani Medical College with the complaints of generalized, multiple, slowly enlarging, painless, non-matted, discrete lymphadenopathy involving entire groups of both cervical, axillary and inguinal regions and significant weight loss of at least 10 kg for last five months. He also complained of non specific constitutional symptoms like anorexia, weakness, apathy and loss of well being. On query patient gave history of occasional low grade fever but did not give any history of hoarseness of voice, dysphagia, dyspnoea or severe abdominal pain. There were no history of cough, productive sputum, breathlessness, hemoptysis, loose motion, drenching night sweats and pruritus. He did not give any prior history of arthralgia and arthritis, dry mouth, erythema nodosum, skin rash and any ocular complaints. There was no significant past medical and sexual history. On systemic examination, there was mild



Fig-1: 70 year old gentleman

splenomegaly. Hemogram showed normocytic normochromic anaemia and raised ESR (80 min in 1st hour). Chest x-ray revealed bilateral hilar adenopathy. Screening for HIV 1&2 and syphilis were negative. The biopsy of cervical lymph node revealed large follicle with vascular proliferation and hyalinization of their germinal centres. Some of the follicle show tight concentric layering of lymphocytes at the periphery of the follicle resulting in a characteristic onion-skin appearance which comply with multicentric Castleman's disease. The CD3 & CD20 were found positive whereas CD30 reported as negative on Immuno histochemistry which was compatible with Castleman's disease. The patient was finally diagnosed as a case of Castleman's disease-multicentric type (considering clinical and laboratory features); and

Address for Correspondence: Dr. Md. Ismail Patwary, Professor & Head, Medicine Department; Sylhet MAG Osmani Medical College Hospital, Sylhet

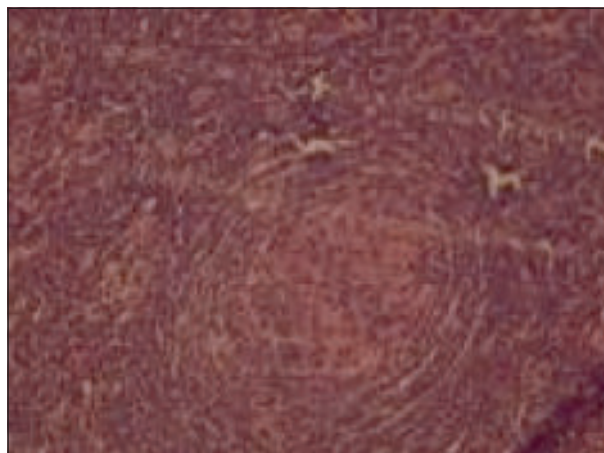


Fig.-2: *Onion Skin appearance in multicentric Castleman's disease on histopathology*

discharged with Tab. Prednisolone 1mg per kg body weight and advised to come in regular follow up. In his first follow up there was significant regression of lymphadenopathy after one month treatment.

Discussion:

The elderly gentleman presented in our hospital with generalized lymphadenopathy and splenomegaly with no other significant systemic features. Bilateral hilar lymphadenopathy and high E.S.R. were found on routine tests. The biopsy report was compatible with Castleman's disease. The diagnosis was further strengthened as the evidence of immuno histochemistry clearly excluded the presence of Lymphoma. Our patient presented to us in 70 years of age in contrast to a case report in Iran where a 5 year old girl only presented with severe abdominal pain and later laparotomy confirmed the mass in the hilum of the liver as hyaline vascular type Castleman's disease.⁵

Localised CD is, by definition, localised to one site. There are features of lymphoid hyperplasia associated with excessive angiogenesis². It is asymptomatic in over 50% of patients³ and is often discovered incidentally. Histological diagnosis requires lymph-node biopsy.

Multicentric CD is characterized by a predominantly lymphadenopathic presentation consistently involving peripheral lymph-nodes and manifestations of multisystem

involvement. It is considered as a systemic B cell lympho proliferation, probably arising in immunoregulatory deficit, and resulting in the outgrowth of clonal B-cell populations.² It is always symptomatic. Symptoms, primarily a consequence of elevated Interleukin-6(IL-6) production, are asthenia(65%), weight loss(67%) and fever(69%).¹ Polyadenopathy is common(84%) with a mean of four sites involved and is often associated with hepatosplenomegaly¹ and this particular feature is consistent with our case. Histological diagnosis is made upon biopsy of an excised peripheral lymph node. A POEMS(Peripheral polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal gammopathy-M protein & Skin signs) syndrome is observed in 24% of patients.¹

The etiology of Castleman's disease is poorly understood and no genetic or toxic factor has so far been identified.

Conclusion:

This case report brings to light the importance of obtaining definitive histological diagnosis in patients presenting with lymphadenopathy and systemic symptoms. Multicentric Castleman's disease is a relatively uncommon cause for such a presentation. Though clinically synonymous with lymphoma, it is an entity that is distinct from malignant lymphoproliferative disorders histologically and prognostically. It may be borne in mind as a differential diagnosis in lymphadenopathic presentations with symptoms of systemic involvement.

References:

1. Sarrot-Reynauld F: Castleman's Disease; Orphanet encyclopedia. August 2001
2. Frizzera G: Castleman's disease and related disorders. *Semin Diagn Pathol.* 1988; 5(4): 346-364.
3. Herrada J, Cabanillas F, Rice L, Manning J, Pugh W. The clinical behaviour of localized and multicentric Castleman disease. *Ann Intern Med.* 1998; 128(8): 657-62.
4. Rose C, Mahieu M, Hachulla E, Facon T, Hatron PY, Bauters F, et al. Le POEMS syndrome. *Rev Med Interne.* 1997; 18(7): 553-62.
5. Hossein Karami, Farzad Masiha. Hyaline vascular type Castleman's disease in the hilum of the liver-Report of a case. *Cases J.* 2010;3:74