Multicentric Castleman disease of hyaline vascular variant presenting with unusual systemic manifestations

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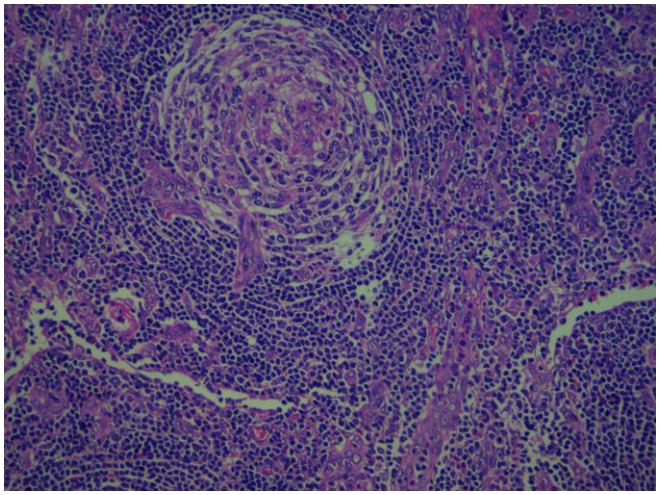
I. Abstract

Castleman disease is a rare lymphoproliferative disorder presenting with localized or disseminated lymphadenopathy and systemic manifestations. It can be categorized in numerous ways, such as unicentric versus multicentric, histopathological variants (hyaline-vascular, plasma cell and mixed), subtypes based on causative viral infections (HIV, HHV-8, Kaposi sarcoma herpesvirus). Presentation ranges from asymptomatic to symptoms involving multiple organs. Even though the exact mechanism of pathogenesis is unknown, treatment is directed towards possible etiologies such as interleukin-6, cluster of differentiation 20 and viral agents.

A 36 year old Sri Lankan lady presented with generalized body swelling and frothyuria for two weeks duration. Examination revealed pallor, generalized edema, axillary, cervical & inguinal lymphadenopathy, hypertension and hepatomegaly. Investigations showed bicytopenia, nephrotic range proteinuria with hypoalbuminaemia, hypogammaglobulinaemia and features of hyaline vascular type Castleman disease in lymph node biopsy. She was managed with Rituximab and made a good clinical improvement.

Castleman disease has a broad spectrum of clinical manifestations, disease pathogeneses and associations and/ or complications. Medical professionals need to be thorough with this spectrum as timely diagnosis and aggressive targeted therapy are the cornerstones in managing the patient.

II. Image:



III. Reference:

1. Bonekamp D, Horton KM, Hruban RH, Fishman EK. Castleman disease: the great mimic. *Radiographics*. 2011 Oct 4; 31(6):1793-1807

2. Casper C. The aetiology and management of Castleman disease at 50 years: translating pathophysiology to patient care. *British journal of haematology*. 2005 Apr 1; 129(1):3-17

3. Abdul-Rahman IS, Al-Amri AM, Ghallab KQ. Castleman's disease: a study of a rare lymphoproliferative disorder in a university hospital. *Clinical Medicine Insights. Blood Disorders.* 2009 Jan 1; 2: 5-19

4. Mah NA, Peretsman SJ, Teigland CM, Banks PM. Castleman disease of the hyaline-vascular type confined to the kidney. *American journal of clinical pathology*. 2007 Mar 1; 127(3):465-468

IV. Biography:

Dr BMDB Basnayake has completed his MBBS at the age of 26 years from University of Peradeniya and Postgraduate Studies from University of Colombo, Sri Lanka. He is a Senior Registrar in Nephrology works at Teaching Hospital Kandy, Sri Lanka. He has published more than 10 articles in reputed journals and has been serving as an editorial board member of repute.

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