

# Mesenteric Castleman's disease- a case report

M Hasan, F Siddiqui, K Gaur, M Naim

## Citation

M Hasan, F Siddiqui, K Gaur, M Naim. *Mesenteric Castleman's disease- a case report*. The Internet Journal of Pathology. 2008 Volume 8 Number 1.

## Abstract

Out of the varying conditions affecting the mesentery, Castleman's disease is a rarely thought of entity. The authors take this opportunity of reporting a case of mesenteric Castleman's disease occurring in a 75 year old female (mother of the main author). The diagnosis was reached after thorough histopathological and immunohistochemical evaluation.

## INTRODUCTION

Castleman's disease is a rare lymphoproliferative disease of unknown origin.<sup>1</sup> In the majority of the reported cases, the disorder is located in the mediastinum, whereas the mesenteric involvement is very unusual.<sup>2</sup> Although it is most commonly seen in adults, it can also occur during childhood.<sup>3</sup> We present a case of mesenteric Castleman's disease which presented as an abdominal mass.

## CASE REPORT

A 75 year old female (mother of the main author) presented to Jawaharlal Nehru Medical College with complaints of acute abdominal pain for the past one day. She also had long history of fever and fatigue, off and on for the past ten years. The patient had consulted many doctors and was prescribed antipyretics but she did not get any permanent relief. The last practitioner made a clinical diagnosis of tuberculosis and the patient was put on antitubercular treatment (ATT). However, the patient did not respond to ATT, rather developed drug intolerance and hence, it was discontinued. The patient developed acute abdominal pain since 1 day and was referred to our hospital.

The patient was of average built and her vitals were normal. Her systemic examination was unremarkable. Haemogram showed mild anaemia (10.2 gm%) with raised ESR (82 mm in 1<sup>st</sup> hour with Westergreen's method). Blood sugar and renal function test were normal.

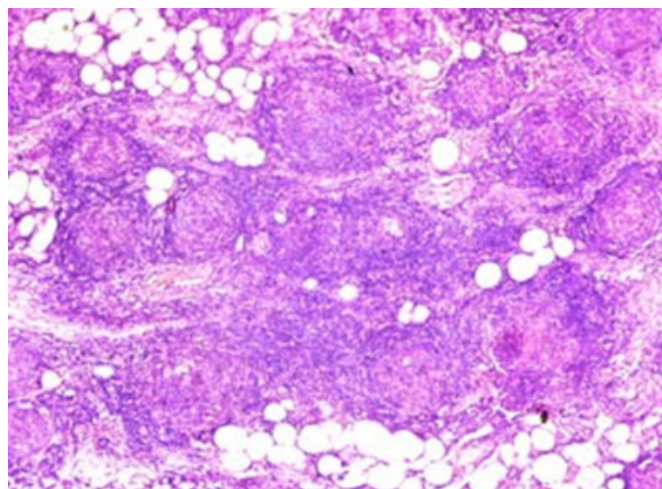
Ultrasonography (USG) showed a well defined solid mass apparently arising from the mesentery with increased mesenteric thickness and mesenteric lymphadenopathy. Fine needle aspiration cytology (FNAC) was performed which was unremarkable. An exploratory laparotomy was done

which revealed an oval mass in the epigastric region (near to mesenteric artery) measuring 1.5 cms, pink in colour and hard in consistency.

Biopsy from the mass was performed and sent for histopathological examination. On histopathological examination, many hyperplastic follicles were seen with mantle-zone expansion poorly formed germinal centres, endothelial hyperplasia and hyaline changes (Fig.1). The characteristic "onion-skin" rimming of B-cell lymphocytes was also seen (Fig.2) with the surrounding cells showing positivity to T-cell (Fig.3). Positivity for both  $\kappa$  and  $\lambda$  chains confirmed the polyclonal nature of the immunoglobulin (Fig.4,5).

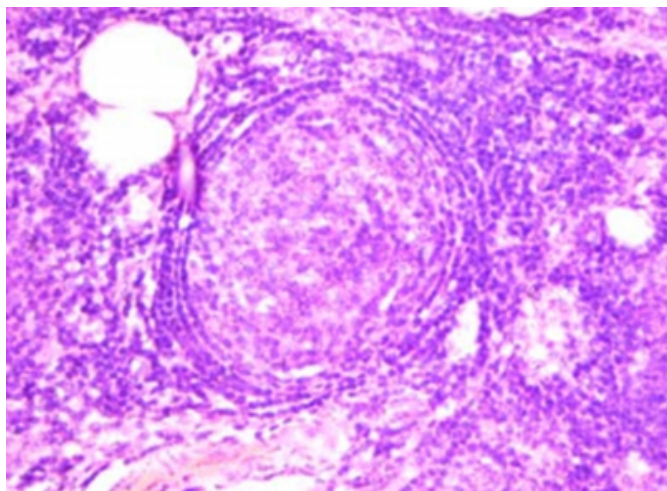
## Figure 1

Figure 1: section showing numerous different sized follicles with germinal centre surrounded by concentric rim of lymphocytes (H & E x 100).



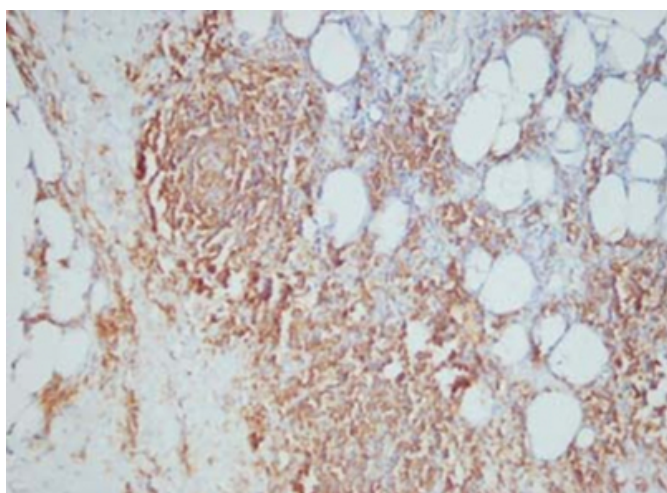
**Figure 2**

Figure 2: section showing germinal centre surrounded by tight concentric rim of lymphocytes, resulting in an onion skin appearance (H & E x 400).



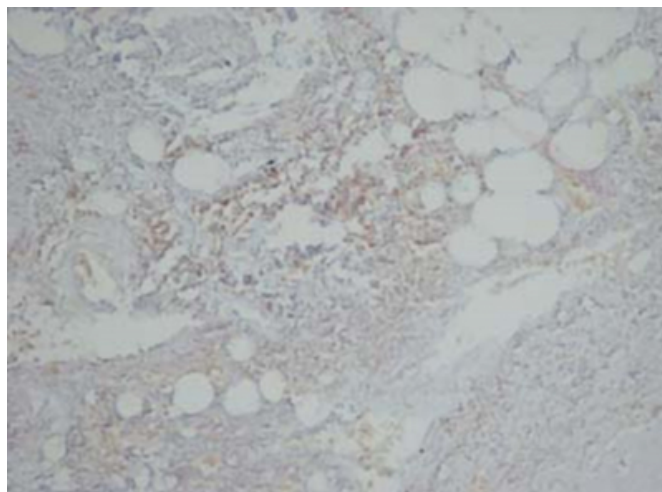
**Figure 3**

Figure 3: section showing cells with positivity to B-cells (x 200).



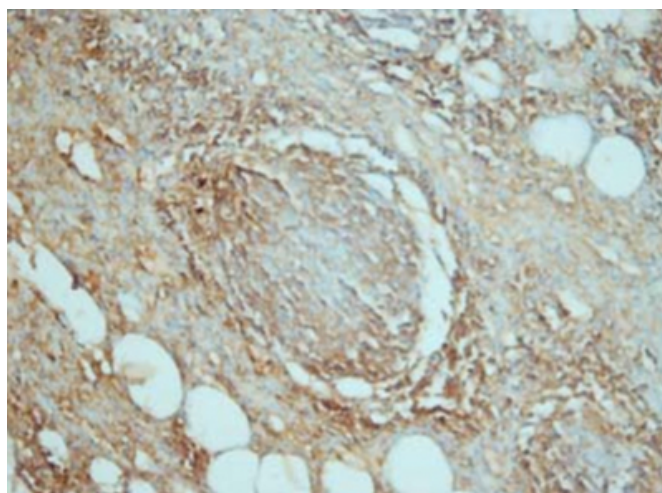
**Figure 4**

Figure 4: section showing cells with weak positivity to T-cells (x 200).



**Figure 5**

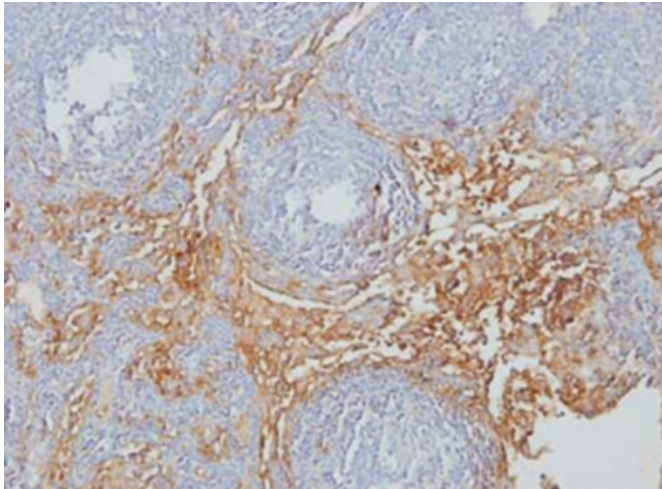
Figure 5: section showing cells with positivity to kappa light chains (x 200).





**Figure 6**

Figure 6: section showing cells with positivity to lambda light chains (x 200)



Since the mass was very close to the mesenteric artery, it was regarded as inoperable and a conservative management was advised. The patient was put on high dose steroids and for the past many months, she is leading a near normal life without any major complications except for occasional fever and bodyache.

**DISCUSSION**

Castleman's disease is a rare lymphoproliferative disease with a varied clinical presentation. It is also called as "angiofollicular lymph node hyperplasia", "giant lymph node hyperplasia", "lymphoid hamartoma" and "follicular reticuloma".<sup>4</sup> First described by pathologist Benjamin Castleman in 1956 while observing anterior mediastinal masses, it was subsequently described in the lung, neck, pelvis, axilla with the mesenteric localization being an uncommon variant.<sup>5</sup>

The disease has been classified as unicentric or multicentric.<sup>4</sup> The unicentric type usually has a benign course presenting as a solitary mass without much constitutional symptoms except for abdominal or chest mass. The graver multicentric type presents with widespread lymphadenopathy, raised IL-6 levels and is usually associated with human-herpes virus-9.<sup>6</sup>

The exact mechanisms causing this condition are poorly understood. Immune dysregulation through immunosuppression, autoimmune processes or continuous low grade inflammation is a key factor.<sup>578</sup> In the quest for a definite etiology, theories like hamartomatous origin, an abnormal host reaction, infections with *Mycobacterium tuberculosis*, *Toxoplasma*, Epstein-Barr virus, HIV, possibly

HCV and proliferation in a neoplastic setting have been proposed.<sup>910111213</sup> IL-6 elevation is associated with both unicentric and multicentric forms.<sup>14</sup> It stimulates B-cell proliferation leading to hyperplastic follicles and hence, enlarged lymph nodes promotes vascular endothelial growth factor (VEGF) secretion activating angiogenesis, promoting hypervascularity and activates acute phase reactants leading to raised levels of erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), immunoglobulins, fibrinogen and serum associated amyloid (SAA).<sup>14</sup>

Histologically 2 major types have been described - hyaline vascular type and plasma cell type. About 90% are of the hyaline vascular variety which is more often associated with benign presentation.<sup>4</sup> This type is frequently found in younger patients and is by large self-limiting. It is believed to be of hamartomatous origin.<sup>9</sup> The hyaline-vascular form has a poorly formed germinal centre surrounded by an expanded mantle zone having rims of CD20+ mature lymphocytes arranged concentrically in an "onion-skin"/whorled appearance.<sup>2</sup> Endothelial hyperplasia, interfollicular stroma with proliferating capillaries, plasma cells and eosinophils are other features.<sup>4</sup> In spite of the bright prognosis, long term follow up is mandatory as very rarely, conversion to malignant lymphoma and Kaposi sarcoma may occur.<sup>4</sup> Plasma cell variant shows larger follicles, a prominent germinal centre and mature plasma cells in interfollicular areas.<sup>10</sup> This is usually associated with the clinically graver multicentric presentation. The patient has a plethora of symptoms like widespread lymphadenopathy, severe anaemia, peripheral oedema, sometimes peripheral neuropathy, raised acute phase reactants and in general a progressively worsening course. It has a known association with POEMS syndrome, Kaposi's sarcoma, osteosclerotic myeloma and AIDS.<sup>41516</sup>

The diagnosis of Castleman's disease is highly evasive with a combination of modalities being used. CT, angiography, histopathological examination and recently immunohistochemistry for  $\kappa$  and  $\lambda$  establishes the polyclonality of the immunoglobulin, as was seen in our case. The hyaline vascular type has a rim of CD20+ lymphocytes and therefore B-cell and T-cell markers are employed while differentiating from lymphoma.

To conclude, we would like to say that in a country like ours where tuberculosis is so endemic, a clinical picture involving lymphadenopathy, abdominal mass and weight loss should not always be diagnosed as abdominal Kochs, rather a far

fetching diagnosis like Castleman's disease should also be kept in mind.

## **CORRESPONDENCE TO**

Dr. Mahboob Hasan C-11 Medical Colony Aligarh Muslim University, Aligarh-202002 U.P., India E-mail: farhan414@rediffmail.com Telephone: 0091-9415460231 (mobile) 0091-5712720666 (residence)

## **References**

1. Castleman B, Towne VW. Case records of the Massachusetts General Hospital. Case 40011. *N Engl J Med* 1954; 250: 26-30.
2. Kimura T, Inoue T, Katayama K, Hirose K, Imamura Y, Yamaguchi A. Mesenteric Castleman's disease: report of a case. *Surg Today* 2002; 32: 651-654.
3. Wei BP, Taylor R, Chan YF, Waters K, Alex G. Mesenteric Castleman's disease in childhood. *ANZ J Surg* 2004; 74: 502-504.
4. Rosai J. Rosai and Ackerman's surgical pathology 9th ed. New Delhi: Elsevier; 2004: p 1905-1908.
5. Francis ND, Hollowood K, Gabriel R. Angiofollicular lymph node hyperplasia. *J Clin Pathol* 1988; 41: 353-354.
6. Cesarman E, Knowles DM. Kaposi's sarcoma-associated herpesvirus: a lymphotropic human herpes virus associated with Kaposi's sarcoma, primary effusion lymphoma and multicentric Castleman's disease. *Semin Diagn Pathol* 1997; 14: 54-66.
7. Frizzera G. Castleman's disease. More questions than answers. *Hum Pathol* 1985; 156: 202-205.
8. Castleman B, Iverson L, Menendez VP. Localized mediastinal lymph node hyperplasia resembling thymoma. *Cancer* 1956; 9: 822-830.
9. Abell MR. Lymph node hamartoma VS. Thymic choristoma of pulmonary hilum. *Arch Pathol* 1957; 64: 584-588.
10. Keller AR, Hochholzer L, Castleman B: Hyaline-vascular and plasma cell types of giant cell hyperplasia of mediastinum and other locations. *Cancer* 1972; 29: 670-683.
11. Hongbei W, Rosemary L W, Michael EZ et al. Castleman's disease in the head of the pancreas: report of a rare clinical entity and current perspective on diagnosis, treatment and outcome. *World J Surg Oncol* 2007; 5: 133.
12. Hzarir NL. Hypervascular follicle hyperplasia and Kaposi's sarcoma in patients at risk for AIDS. *N Engl J Med* 1984; 310: 462-463.
13. Zettergreen L. Probable neoplastic proliferation of lymphoid tissue. *Acta Pathologica et Microbiologica Scandinavica* 1961; 51: 113-126.
14. Yoshizaki K, Matsuda T Nishimoto N et al. Pathogenic significance of interleukin-6 in Castleman's disease. *Blood* 1989; 74: 1360-1369.
15. Sahota P, Das CP, Prabhakar S, Rajwanshi A, Singh P, Khurana D. Polyneuropathy in osteosclerotic myeloma coexisting with hyaline vascular Castleman's disease. *Journal of Clinical Neuromuscular Disease* 2005; 7: 29-35.
16. Saif MW. Castleman disease in an HIV infected patient with Kaposi sarcoma. *AIDS Read* 2001; 11: 572-576.

**Author Information**

**Mahboob Hasan, MD**

Lecturer, Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University

**Farhan Asif Siddiqui, MD**

Senior Resident, Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University

**Kavita Gaur, MD**

Junior Resident, Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University

**Mohamed Naim, MD**

Professor, Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University