# Mesenteric Castleman's Disease Presenting As Sub-Acute Intestinal Obstruction- ACase Report & Review of Literature.

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Abstract: Castleman's disease is a rare benign lymphoproliferative disease. The most common sites of the disease are mediastinum, neck and axilla. Considering its lymphatic origin, the disease can occur theoretically anywhere in the human body. The location of the disease in mesentery is rare and usually associated with generalized form and is multifocal. We report a rare presentation of isolated mesenteric Castleman's disease in a young 28 year old male presenting as sub-acute intestinal obstruction. The final diagnosis was made after resection of the diseased intestine on histopathological examination as unicentric mesenteric Castleman's disease of the hyaline vascular type.

Keywords: Castleman's disease, mesenteric lymphadenopathy, intestinal obstruction, hyaline vascular type.

\_\_\_\_\_ Date of Submission: 29 -07-2017

Date of acceptance: 23-08-2017

### I. Introduction

Castleman's disease was first described by Dr. Benjamin Castleman in 1954[1]. It is a rare benign disease caused by proliferation of lymphoid tissue which can be in localized or generalized form. The localized form of the disease is usually asymptomatic whereas the multicentric form is associated with generalized symptoms and commonly seen in association with syndromes like AIDS, POEMS, Kaposi's sarcoma, lymphomas, paraneoplastic pemphigus and plasma cell dyscrasias. There is no sex predilection. Castleman's disease is a pathological diagnosis and can be of three types- hyaline vascular type(70%), plasma cell type(20%) or mixed type(10%)[2]. The location of the disease involving primary lymphatic tissues is approximately 70% in the chest, 15% in the neck and 15% in the abdomen and pelvis.[3,4]. We report a case of isolated mesenteric Castleman's disease of the hyaline vascular type in a young male who presented as sub-acute intestinal obstruction.

### **Case report** II.

A 28 yr old male patient presented with severe constant lower abdominal pain over 2 days associated with several episodes of bilious vomiting. Patient was hospitalized frequently in the past 6 months at different hospitals for recurrent pain abdomen for which he was treated conservatively. Clinical examination revealed diffuse tenderness and distension in lower abdominal quadrants with normal bowel sounds and impacted stools on per-rectal examination. Laboratory investigations showed normal counts and levels. X-ray erect abdomen showed dilated small bowel loops with multiple air- fluid levels. Ultrasound abdomen revealed mesenteric lymphadenopathy with dilated bowel loops. Mantoux test, widal test, serology testing and TB quantiferon gold test were done and was negative. CECT abdomen study showed dilated small bowel loops with multiple airfluid levels with coiling and segmental luminal narrowing of distal ileum. No evidence of free fluid or pneumoperitoneum was noted. Features were suggestive of sub-acute intestinal obstruction likely distal ileal stricture/knotting.

Exploratory laparotomy was done which revealed dense small bowel adhesions with segmental intertwinning of ileum over its own loop causing gangrene of 20 cm of ileum, with distal dilated segments of small bowel(fig.1). Multiple enlarged mesenteric lymph nodes were noted. On surgery, adhesions were released, 25 cm of gangrenous ileum was resected and end to end ilealanastamosis done. The resected specimen with mesentry involving lymph nodes were sent for HPE. Postoperative period was otherwise uneventful. Patient recovered well. Histopathological examination of the specimen revealed gangrenous small intestine with the mesenteric lymph nodes showing large scattered follicles which were arranged in tight concentric layers of lymphocytes giving an onion skin appearance. The follicles had a characteristic expansion of their mantle zone with small germinal centres. At the centre of the follicle, hyaline material and proliferating blood vessels were seen which is characteristic of unicentricCastleman's disease- hyaline vascular type(fig.2). The patient was reviewed at regular intervals for a period of two years and there is no evidence of recurrence.

# III. Discussion

Castleman's disease is a rare pathological process with undefined incidence and etiology characterized by lymphoid hyperplasia. It accounts as one of the causes for non-neoplastic lymphadenopathy[5]. The disease is divided histologically into two major sub types- the hyaline vascular type and the plasma cell type. Patients with the hyaline vascular type are usually asymptomatic and the disease is discovered incidentally, in contrast to patients with plasma cell type, which are often symptomatic at the time of diagnosis [6]. Our case is an exeption in the hyaline vascular type becoming symptomatic. As discussed above, the occurrence of the disease can be a solitary mass form or the less common multicentric form. Irrespective of the histopathological type, the localized form always shows a benign behaviour and complete surgical excision provides cure of the disease with no reported cases of recurrence. On the contrary, the multicentric form follows a more aggressive course and is associated with poor prognosis. The multicentric disease is characterized by disseminated lymphadenopathy, and is always associated with systemic symptoms[7]. The treatment remains controversial which includes surgery, chemotherapy, cortico-therapy or combination of these. Castleman's disease is a great mimic of both benign and malignant findings in abdomen and pelvis. It has to be differentiated from the mesenchymatous lesions of mesentry such as stromal tumors, leiomyomas, sarcomas, lipomas, desmoids tumors, lymphomas and metastatic lymphadenopathy and inflammatory/infectious causes such as terminal ileitis and tuberculosis. The preoperative diagnosis of the disease is still very difficult even with latest medical imaging techniques[8]. In most cases, the diagnosis of mesenteric Castleman's disease is confirmed only after resection and histopathological examination of the specimen.

## IV. Conclusion

Castleman's disease should be considered as a differential diagnosis of every mass located in the mesentery including lymphomas, metastatic lymphadenopathy, and infectious / inflammatory causes of mesenteric adenopathy especially when clinical and imaging features are unclear.

## References

- [1]. Castleman B, Iverson L, Menendez VP. Localized mediastinal lymph node hyperplasia resembling thymoma. Cancer 1956; 9(4) : 822-830.
- [2]. Keller AR, Hochholzer L, Castleman B. Hyaline vascular and plasma-cell types of giant lymph node hyperplasia of the mediastinum and other locations. Cancer 1972;29(3):670-683.[Medline].
- [3]. Lee JP, NgaiRL : Mesenteric giant lymph node hyperplasia report of a case and review of the literature. AustralasRadiol. 1990, 34:162-164. 10.1111/j. 1440-1673.
- [4]. Talat N, Belgaumkar AP, Schulte KM. Surgery in Castleman'sdisease : a systematic review of 404 published cases. ANN surg. 2012 Apr. 255 (4): 677-84.[Medline].
- [5]. Wei BP, Taylor R, Chan YF, waters K, Alex G. Mesenteric Castleman's disease in childhood. ANZ J Surg 2004; 74: 502 504.
  [6]. Kimura T, Inoue T, Katayama K, Hirose K, Imamura Y, Yamaguchi A. Mesenteric Castleman's disease : report of a case. Surg
- [6]. Kimura T, Inoue T, Katayama K, Hirose K, Imamura Y, Yamaguchi A. Mesenteric Castleman'sdisease : report of a case. Surg Today 2002; 32: 651-654.
- [7]. Libson E, Fields S, Strauss S et al. Widespread Castleman disease: CT and US findings. Radiology 1988; 166 : 753.
- [8]. Kunishima S, Taniguchi H, Koh T, Yamaguchi A, Yamagishi H. F-18 fluorodeoxyglucose positron emission tomography in mesenterialCastleman's lymphoma. ClinNucl Med 2001; 26:789-790.



Fig 1- Exploratory laparotomy showing severe adhesions with segmental inter-twinning of ileum over its own loop with gross mesenteric lymphadenopathy.

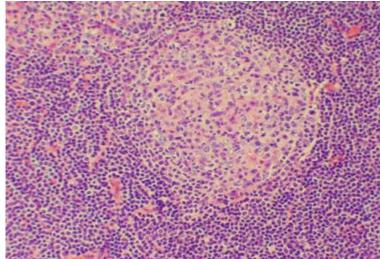


Fig 2- Onion skin appearance- UnicentricCastleman's disease - hyaline vascular type.

\*Dr.Tharun Ganapathy. " Mesenteric Castleman's Disease Presenting As Sub-Acute Intestinal Obstruction- ACase Report & Review of Literature." IOSR Journal of Dental and Medical Sciences (IOSR-JDMS) 16.8 (2017): 10-12

DOI: 10.9790/0853-1608081012