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CASTLEMAN'S DISEASE PRESENTING AS A RARE ABDOMINAL WALL MASS: A CASE REPORT AND SURGICAL APPROACH

General Surgery

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ABSTRACT

Castleman's disease (CD) is a rare, nonclonal lymphoproliferative disorder of uncertain etiology that affects lymph nodes. It is classified as either unicentric (localized) or multicentric (systemic) based on clinical and radiological findings. Pathologically, CD is further categorized into hyaline vascular, plasmacytic, or mixed subtypes. Unicentric CD typically presents in the 3rd to 4th decade, while multicentric CD is more common in the 5th to 6th decade, often associated with immunocompromised individuals, especially those with HIV and HHV-8. We present the case of a 45-year-old male with a long-standing abdominal mass, diagnosed as unicentric Castleman's disease. Surgical excision led to complete resolution with no recurrence on follow-up. This report highlights the diagnostic challenges and emphasizes the importance of histopathological confirmation in managing CD.

KEYWORDS

Castleman's disease, Unicentric Castleman's disease, Abdominal lump, Abdominal wall mass, Lymphoproliferative disorder, Hyaline vascular type.

INTRODUCTION:

Castleman's disease (CD) is a rare lymphoproliferative disorder that can mimic both benign and malignant diseases, particularly involving the lymph nodes. The disease can affect any lymph node region, including the neck, chest, abdomen, and pelvis. Due to the lack of specific clinical features, diagnosis often requires histopathological examination after exclusion of other lymphadenopathies. CD is classified into unicentric and multicentric forms, with the unicentric type generally associated with a more favorable prognosis following surgical excision. The disease can also be classified histopathologically into hyaline vascular, plasmacytic, or mixed types, with the hyaline vascular form being the most common. The etiology remains unclear, though there is a strong association with immunodeficiency, especially in multicentric CD, linked to HHV-8 and HIV infections.

CASE REPORT:

A 45 year old male presented to the Outpatient department with chief complaints of a lump in abdomen since 20 years and associated on and off pain since 6 months. This lump gradually increased in size. Patient gave history of decreased appetite and loss of weight (not documented by him). He also gave history of malena since last 10 days. No history of fever, vomiting, altered bowel/bladder habits, haematemesis, Jaundice, cough. No known co-morbidities. No previous operative history.

On Per abdomen examination, Abdomen was soft, non tender, no distension, 6x4 cm lump palpated in right iliac fossa. No organomegaly present, Hernial sites and genitals were normal. PR examination was unremarkable.

Outside MRI reports suggested a 4.7 x 4.1 x 5 cm well defined homogenously enhancing lesion in Right iliac fossa, abutting adjacent bowel loops with mild free fluid in pelvis and splneomegaly. The Bowel loops and rest of the abdomen was normal.

Patient was further advised USG guided biopsy which on histopathological examination was suggestive of a lymphoid neoplasm. On further consultation with Oncologist Excision biopsy was planned. Haematological investigations revealed Hb of 11.3, thrombocytosis (489000/cumm) and increased Plasma fibrinogen levels of more than 700. Rest of the laboratory investigations were within normal limit.

Laparascopy was performed which showed a large solid tumor of size 8x5x4 cm arising from right lateral pelvic wall (Fig 1.). Dark straw colored fluid was present in peritoneal cavity (Fig 2.). Whole peritoneal surface, diaphragmatic surface, liver, spleen, omentum and bowel loops were studded with tubercles (Fig 3.). The mass was excised laparoscopicaly and specimen was excised through a samll infraumblical midline incison. Omental biopsy was aslo taken. Specimen was sent for Histopathology, Fluid was sent for Cytology, Gene-expert, AFB stain and culture sensitivity.

The procedure was uneventful, and the patient was transferred to the ward. Soft diet was introduced on postoperative day 2, and the patient was discharged on day 4.

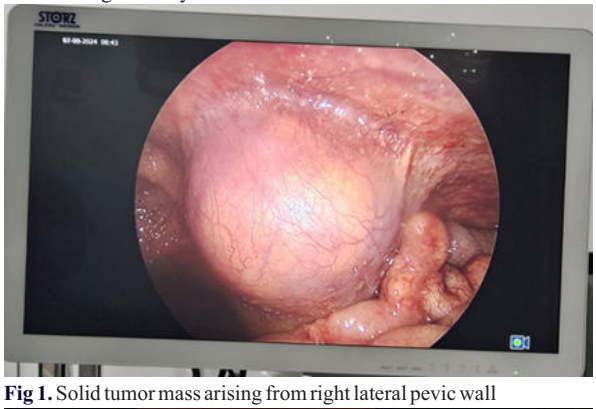

Fig 1. Solid tumor mass arising from right lateral pevic wall

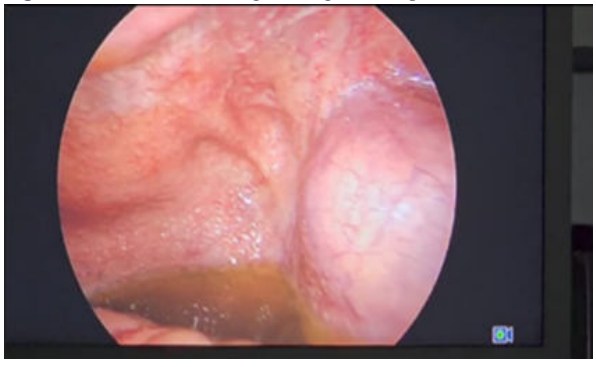

Fig 2. Dark straw colored peritoneal fluid

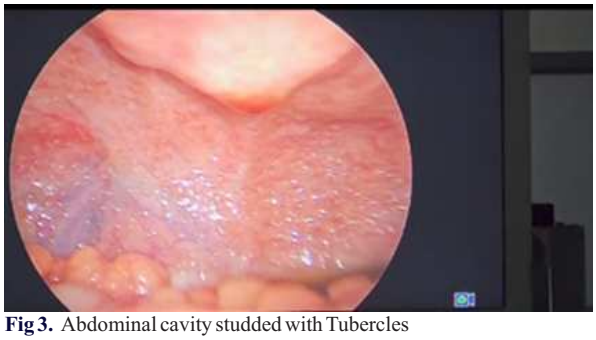

Fig 3. Abdominal cavity studded with Tubercles

Histopathological analysis confirmed the diagnosis of unicentric Castleman's disease (hyaline vascular type), with typical features such as atretic germinal centers, endothelial venules, and the characteristic "onion skin" appearance of lymphocytes around hyalinized capillaries. Immunohistochemical staining showed positivity for CD20 (follicular cells), CD3 (paracortical cells), PAX5, and CD138 (plasma cells), confirming the diagnosis.

The excised omental specimen showed granulomatous inflammation with foamy macrophages, lymphocytes, foreign body and Langhans giant cells, occasional epithelioid cells, fibroblasts, and few histiocytes. Cytology of the peritoneal fluid was negative for malignancy and AFB culture, with sputum and GeneXpert negative for M. tuberculosis. Following consultation with the surgeon and pulmonologist, empirical anti-tubercular therapy (ATT) was initiated.

Fig 4. Cut open excised mass was fleshy, grey white with areas of Haemorrahge.

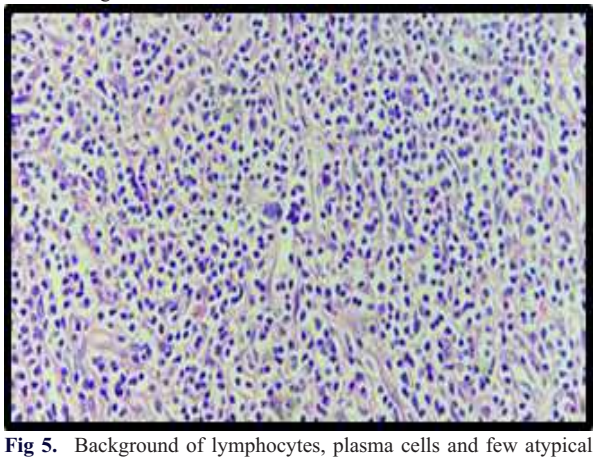

Fig 5. Background of lymphocytes, plasma cells and few atypical cells which are large, polygonal with vesicular nuclei, 1-2 prominent nucleoli and moderate cytoplasm

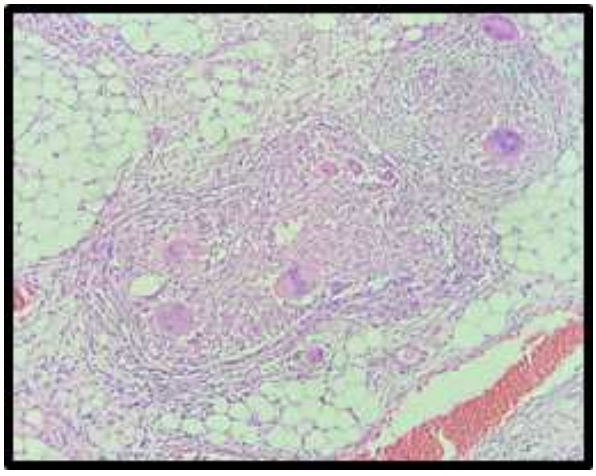

Fig 6. Omental Biopsy showing Giant cell, foamy macrophages, lymphocytes and epitheloid cells

On follow-up, the patient showed good clinical progress, with tolerance to anti-tubercular therapy and notable improvement in overall health.

DISCUSSION:

Castleman's disease (CD) is a rare, non-neoplastic lymphoproliferative disorder that presents significant diagnostic challenges due to its variable clinical, radiological, and pathological features. Although it can affect lymph nodes in various regions of the body, its presentation as a mass arising from the abdominal wall is extremely uncommon, further complicating diagnosis. In our case, the patient presented with a long-standing abdominal mass, which, based on imaging and clinical findings, initially raised suspicions of a neoplastic process. The eventual diagnosis of unicentric Castleman's disease (UCD) was made through histopathological evaluation, underscoring the importance of biopsy in such cases.

UCD is typically localized and is usually curable by complete surgical excision, as was demonstrated in this case. However, distinguishing it from malignant lymphoid neoplasms, as well as other causes of granulomatous inflammation, such as tuberculosis or sarcoidosis, is essential for appropriate management. In our patient, histopathological examination revealed characteristic features of the hyaline vascular subtype, which is the most common form of UCD. The presence of atretic germinal centers, hyalinized capillaries, and the typical “onion skin” appearance of lymphocytes surrounding follicles were key histological clues that led to the diagnosis.

The granulomatous inflammation observed in the excised omental biopsy, combined with the patient's clinical presentation, raised the possibility of tuberculosis as a differential diagnosis. However, cytology of the peritoneal fluid, along with negative results for AFB culture and GeneXpert, ruled out active tuberculosis. Despite this, based on the clinical suspicion and consultation with the pulmonologist, empirical anti-tubercular therapy (ATT) was initiated. This decision highlights a common challenge in clinical practice, where empirical treatment is sometimes necessary due to the overlapping presentations of granulomatous diseases.

The management of UCD primarily involves surgical excision, which is typically curative, as seen in our case. The prognosis for patients with UCD is excellent, with minimal risk of recurrence following complete resection. In contrast, multicentric Castleman's disease (MCD), which involves multiple lymph node regions and systemic symptoms, has a more variable prognosis and often requires systemic therapy, including corticosteroids, immunotherapy targeting IL-6, or antiviral therapy in cases associated with HHV-8. While our patient's disease was localized, the presence of systemic symptoms, such as weight loss and mild splenomegaly, necessitated thorough investigation to rule out MCD and other causes of systemic inflammation.

This case emphasizes the diagnostic complexities associated with Castleman's disease, particularly when it presents in atypical locations like the abdominal wall. It also illustrates the importance of a

multidisciplinary approach, involving both surgical and medical specialties, in managing such rare conditions. The successful surgical outcome and the patient's recovery highlight the efficacy of early diagnosis and intervention in UCD. Additionally, the decision to initiate empirical ATT reflects the need to balance clinical suspicion with laboratory findings, especially in regions with a high prevalence of tuberculosis.

This case contributes to the limited literature on Castleman's disease presenting as an abdominal wall mass and reinforces the role of histopathological examination in establishing a definitive diagnosis. While surgical excision remains the cornerstone of treatment for unicentric disease, the management of associated conditions, such as granulomatous inflammation, requires careful clinical judgment and close interdisciplinary collaboration.

CONCLUSION:

In conclusion, this case highlights the diagnostic and therapeutic challenges posed by Castleman's disease, particularly when presenting as an abdominal wall mass, a rare occurrence. Histopathological confirmation remains critical in distinguishing it from other lymphoproliferative disorders and granulomatous diseases. Surgical excision was curative in this unicentric case, with an excellent post-operative outcome. The overlap with tuberculosis necessitated empirical ATT, reflecting the complexity of managing such cases in high-prevalence regions. Early diagnosis and a multidisciplinary approach are key to optimal outcomes in rare conditions like Castleman's disease.

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Conflicts Of Interest:
The authors have no conflicts of interest to declare.

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