

## Localized Castleman's Disease in Retroperitoneum Mimicking Broad Ligament Leiomyoma – a Diagnostic Challenge

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**Received:** February 23, 2015 ; **Accepted:** May 5, 2015

Primary retroperitoneal tumours pose a diagnostic dilemma and therapeutic challenges because of their location and late detection. Localized Castleman Disease (CD) in retroperitoneal location is a rare occurrence, with incidence of only 4% cases occurring in diverse locations like retroperitoneum. A rare case of unicentric localized Castleman Disease localized in broad ligament is hereby reported which was clinically & radiologically suspected as broad ligament leiomyoma but finally diagnosed as Castleman's Disease, hyaline vascular type in a young female posing a diagnostic challenge both for the pathologist and clinician. Definite diagnosis is essential as complete surgical excision in case of unicentric Castleman's Disease has good prognosis after resection.

**Keywords:** castleman's disease; leiomyoma; retroperitoneum.

### INTRODUCTION

The retroperitoneum can host a wide spectrum of pathologies, including a variety of rare benign tumors and malignant neoplasms that can be either primary or metastatic lesions. Retroperitoneal tumors can cause a diagnostic dilemma and present several therapeutic challenges because of their rarity, relative late presentation, and anatomical location, often in close relationship with several vital structures in the retroperitoneal space.<sup>1</sup>

Localized Castleman Disease (CD) as a differential diagnosis of retroperitoneal tumours is a rare occurrence, with incidence of only 4% cases occurring in diverse locations like retroperitoneum.<sup>2</sup> Castleman Disease, is a distinct clinicopathological entity and a rare lymphoproliferative disorder, first described by Dr Benjamin Castleman and colleagues in 1956, in patients presenting with mediastinal masses. They have been histologically categorized into hyaline-vascular type, plasma cell type and mixed type, and clinically into localized and multicentric type.<sup>1,3</sup>

It is of pivotal importance to identify unicentric as opposed to multicentric disease at a clinical level as the therapeutic intervention is different. A rare case

of unicentric localized Castleman Disease localized in broad ligament is hereby reported which was clinically & radiologically misdiagnosed as broad ligament leiomyoma in a young female posing a diagnostic challenge both for the pathologist and clinician.

### CASE

Thirty seven years old female attended the outpatient Department of Obstetrics & Gynaecology with chief complaint of pain abdomen. The pain was continuous, of low intensity and mainly limited to the right side of the abdomen. There was no history of fever, weight loss, and diarrhea.

On per abdominal examination, a supra-pubic fixed firm mass was palpated. Per vaginal examination also revealed an irregular mass which was not freely moving, palpated through right fornix. The other fornix and uterus was unremarkable. Her routine hematological and biochemical examination were within normal limits apart from mild anaemia and raised fasting blood glucose levels.

Patient underwent transvaginal sonography, which showed a hypoechoic mass measuring 15.5 cm in greatest axis, attached to right broad ligament and separate from the right ovary and tube. The uterus and other side of the adnexae were within normal limits. Keeping in mind, the clinical and radiological findings, a provisional diagnosis of broad ligament fibroid was made. After stabilizing her blood

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glucose levels, en bloc dissection of broad ligament fibroid along with hysterectomy was done and the specimen was sent to the department of pathology for histopathological examination.

On gross, the specimen consisted of body of uterus and cervix along with a large round well circumscribed mass measuring 17 cm in its greatest dimension. The cut surface of the mass was completely solid and gray white (Figure 1). Representative sections from mass was taken and processed.



**Figure 1. Specimen comprising body of uterus and cervix along with a large round well circumscribed mass measuring 17 cm in its greatest dimension.**

Interestingly, on microscopic examination, the tumour comprised of variable sized lymphoid follicles along with marked vascular proliferation and hyalinization of abnormal germinal centre.

The periphery of the follicles showed concentric layering resulting in onion-skin appearance. Numerous post capillary venules and conspicuous absence of sinuses were also seen and a histopathological diagnosis of Castleman Disease, hyaline vascular type of broad ligament was offered.

## COMMENT

Castleman's disease (CD) is a rare and poorly understood disease entity that may resemble more common conditions and represents a clinical challenge to the treating surgeon. The hyaline vascular type represents 76% -91% of localized disease while plasma cell type represents 9%- 24% cases with generalized involvement and constitutional symptoms.<sup>4</sup>

The localized forms are asymptomatic in 51% of the cases and are often discovered at the time of routine

physical examination. The clinical presentation of localized CD can be due to symptoms related to compression effect on surrounding organs such as nausea, vomiting, weight loss, post-prandial discomfort, abdominal and or lumbar pain.<sup>5</sup> The present case also presented with the chief complaint of pain abdomen and had a huge broad ligament mass and clinical presentation was related to its location and size.

Ultrasonography, computed tomography (CT) scan, and magnetic resonance imaging (MRI) have proven to be useful in the diagnosis of masses located in the retroperitoneum. Ultrasonography usually reveals a hypoechogenic and homogenous mass. CT scan shows a solid homogenous hypervascular mass when the tumor diameter is less than 5 centimeters, whereas larger tumors (>5 centimeters), because of necrosis or fibrosis, tend to have more heterogeneous enhancement with a central low-attenuation area.<sup>6</sup> However, the images of CD resemble other masses including lymphoma, tuberculosis, sarcoidosis, and retroperitoneal sarcomas, the present case being misdiagnosed as broad ligament leiomyoma on ultrasonography. Contrast-enhanced CT scan in localized CD may show dotted and stripy or rim like enhancement along with star-shaped microcalcifications, while MRI show hypodense mass or hyperdense lesion, sometimes with star-shaped calcifications.<sup>7,8</sup> Increased vascularity on radiological investigations in case of CD in rare locations may confound the diagnosis and cause suspicion for primary malignancy with associated angiogenesis.<sup>9</sup> Radiologically, the present case was misinterpreted as leiomyoma because of its unusual location and non specific radiological findings.

Many different therapeutic approaches have been used namely, curative resection, radiotherapy, steroids, immunotherapy such as interferon-alpha or anti-IL-6 antibodies and combination chemotherapy such as cyclophosphamide, vincristine, and doxorubicin to manage the disease.<sup>6</sup> Complete surgical excision is the treatment of choice for localized, unicentric lesions in any organ domain. But, there is no curative indication for surgery in cases of multicentric type and various forms of immunochemotherapy have to be given for treatment. At present, the role of the surgeon in cases of multicentric CD is limited to gaining tissue for an appropriate biopsy and to debulking of dominant foci of multicentric disease in presence of specific organ-

related indications.<sup>10</sup>

Ultimately, biopsy of an enlarged lymph node or mass is mandatory to reach at a definitive diagnosis and other ancillary investigations are required to differentiate localized from multicentric variety.<sup>7</sup> This is also required in lieu of therapeutic option which is complete surgical excision in case of unicentric CD with the prognosis being close to 100% after resection.<sup>5</sup> Complete surgical resection was done in the present case also with no post operative complications. Patient was kept under follow up and has not shown any evidence of the disease till date.

## CONCLUSIONS

In conclusion, pathologists as well as clinicians should be aware of the possibility of pelvic involvement of this disease in spite of its extreme rarity. As CD can mimic number of different lesions clinico-radiologically, it should be included in the differential diagnosis of any hypervascular and heterogeneous tumor mass in the retroperitoneum detected radiologically.

## DISCLOSURE

The authors report no conflicts of interest in this work.

No violation of human rights and safety.

Funding: Nil

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