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Unicentric Castleman's Disease: Laparoscopic Approach of a Para-Duodenal Retroperitoneal Mass

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Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Patient: Female, 34-year-old
Final Diagnosis: Unicentric Castleman's disease
Symptoms: Hematuria
Medication: —
Clinical Procedure: Laparoscopic approach of a para-duodenal retroperitoneal mass
Specialty: Surgery

Objective: Rare disease

Background: Castleman's disease is a benign, lymphoproliferative disorder that is extremely uncommon. Multiple classifications have been described; however, the exact etiology remains unknown. Preoperative diagnosis is not common, as imaging cannot distinguish the disease from other processes, and biopsy is insufficient to provide the architecture of the mass, which is necessary for diagnosis. Unicentric retroperitoneal disease has been described, and management includes complete resection of the mass, which is usually curative.

Case Report: A 34-year-old previously healthy woman presented with hematuria. Evaluation revealed a retroperitoneal mass that was abutting the duodenum and head of the pancreas. Biopsy failed to provide a diagnosis, so laparoscopic resection was performed. Postoperative diagnosis was consistent with unicentric Castleman's disease.

Conclusions: Castleman's disease is an uncommon process, and one that is difficult to diagnose. Unicentric Castleman's disease should always be a differential diagnosis of solitary retroperitoneal masses that are well-demarcated, as treatment can be curative with surgical resection.

MeSH Keywords: Laparoscopes • Medical Oncology • Retroperitoneal Neoplasms

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/918444

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Background

Castleman's disease (CD) is a rare, benign, lymphoproliferative disorder first described in 1954 as a "peculiar form of lymph-node hyperplasia" at Massachusetts General Hospital [1]. Benjamin Castleman further described the disease as unicentric versus multicentric [2]. The unicentric and multicentric variants are classified based on the number of lymph nodes that are involved in the disease process. The most common location for these enlarged lymph nodes is the mediastinum (in 70% of patients); other locations include the neck (15%), abdomen and pelvis including the retroperitoneum (12%), and axilla (3%) [3]. The presentation of unicentric CD (UCD) varies, with some patients being asymptomatic. The literature differs on the rate of symptomatic presentation of UCD, with one study showing 28.6% of patients being symptomatic [4], and other findings of symptoms in 69% of patients [5,6]. In this report, we present an unusual case of a para-duodenal, peripancreatic, retroperitoneal UCD.

Case Report

A 34-year-old previously healthy woman presented with persistent hematuria following an uncomplicated vaginal delivery, and she denied any other symptoms. Abdominal and pelvic exams did not show any abnormalities. Basic laboratory work-up was done and results were within normal range. An abdominal ultrasound was done, which showed a 6.5×4.7 cm hypoechoic mass adjacent to the upper pole of the right kidney. To further evaluate this mass, an abdominal MRI was performed, which showed a 6.2×9×4.5 cm solid mass located between the liver, right kidney, and duodenum. Based on these findings, it was concluded that the mass was retroperitoneal. Furthermore, the mass had a hypointense signal on T1 and hyperintense signal on T2, with enhancement after IV contrast administration.

Radiologic diagnosis as to the nature of this mass was inconclusive, so a CT scan was recommended to view any calcification in the mass and to see whether there was extension into the duodenum. An abdominal CT showed a 6×8.5×4.2 cm solid, well-delineated mass with marked enhancement with IV contrast. The mass was shown to be retroperitoneal, adjacent to the right kidney and in close proximity to the duodenum. It was also shown that the mass was well-vascularized, with a large vein arising from it. A chest CT scan was normal.

The differential diagnosis at this point was neuroendocrine tumor versus gastrointestinal stromal tumor (GIST), considering the close proximity of the mass to the duodenum, which could be the origin of this mass. A work-up for a secretory neuroendocrine tumor, including blood and urine metanephrine and normetanephrine, urine epinephrine, norepinephrine, and dopamine, was negative.

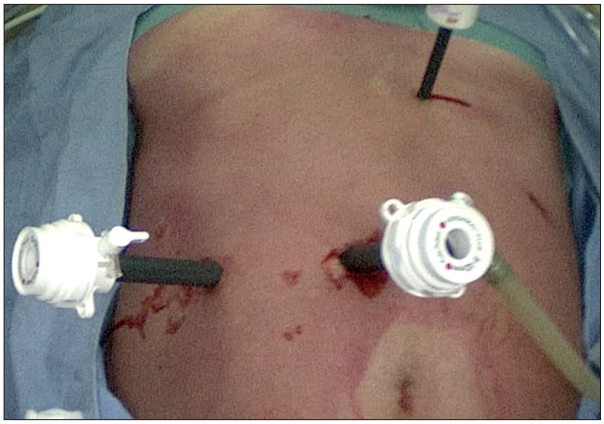

Figure 1. 2×12 mm ports in the right paramedian area and right subcostal area, with a 2×5 mm port in the epigastric area mid-distance between the xyphoid and umbilicus.

An endoscopic ultrasound (EUS)-guided biopsy was planned, and endoscopic evaluation showed an 8×5 cm, well-demarcated, hypoechoic mass with few hyperechoic spots and small vasculature impinging on the wall of the 3rd portion of the duodenum. Doppler evaluation showed multiple vessels traversing the mass. A histologic evaluation of the biopsy did not show any evidence of GIST or any other carcinoma; it showed multiple lymphoid cells with no characteristics of lymphoma, but that diagnosis could not be ruled out. A decision was made to excise the mass laparoscopically for both diagnostic and therapeutic advantages.

In preparation for the surgery, and considering the vascularity of the mass, a triphasic abdominal CT was done, which showed the para-duodenal enhancing mass with a small focus of hypoattenuation within the mass that appeared to be abutting the posterior aspect of the duodenum, with no clear evidence of invasion. It also showed the mass compressing the underlying inferior vena cava (IVC), with a clear fat plane between them and no invasion of the IVC.

The procedure was conducted with the patient in the lithotomy position, using 2×12 mm ports in the right paramedian area and right subcostal area, and 2×5 mm ports in the epigastric area mid-distance between the xyphoid and umbilicus (Figure 1). Exploration of the abdomen showed the retroperitoneal mass anterior to the right kidney and IVC, pushing the duodenum and the head of the pancreas anteriorly and medially (Figure 2). The mass was very well-vascularized and supplied by the medial branches of the renal artery and venous drainage into the medial branches of the renal vein (Figure 3). The medial branches of the renal vessels were carefully dissected, clipped, and divided (Figure 4). The mass was dissected and freed from all its attachments. Large lymph nodes were

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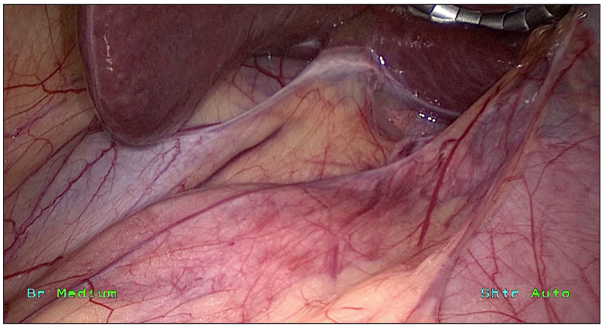


Figure 2. Retroperitoneal mass anterior to the right kidney and IVC, pushing the duodenum and the head of the pancreas anteriorly and medially.

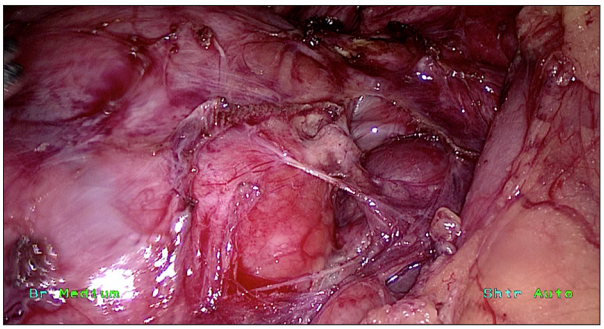


Figure 3. Well-vascularized mass supplied by the medial branches of the renal artery with venous drainage into the medial branches of the renal vein.

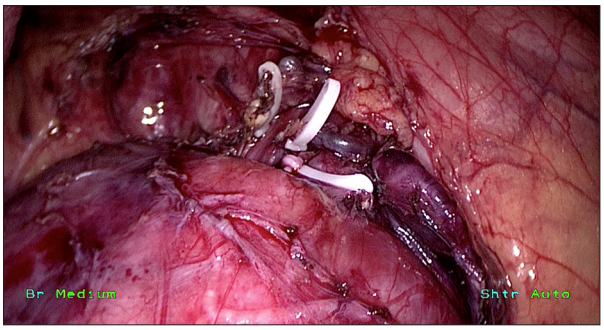
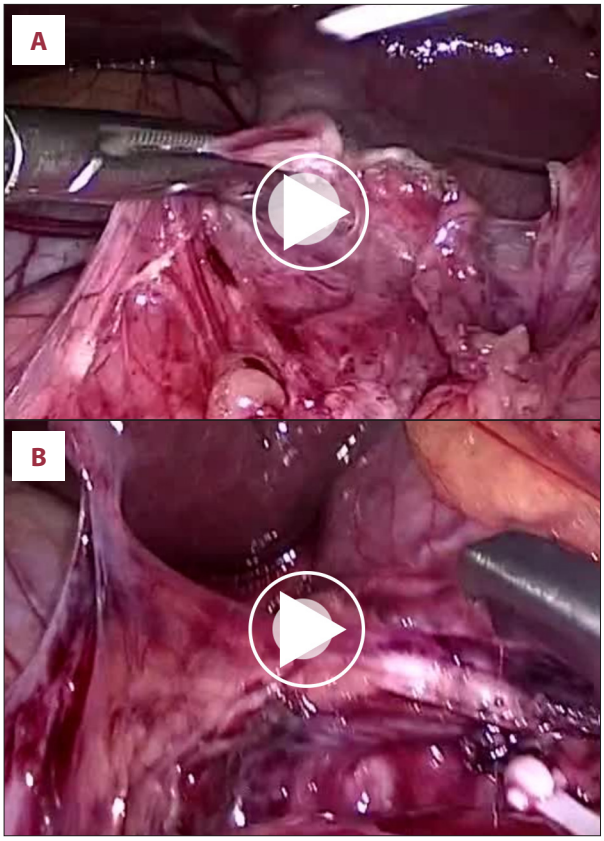


Figure 4. Medial branches of the renal vessels dissected, clipped, and divided.

found medial to the mass and were also resected. The procedure was conducted in 120 minutes (as seen in the summarized version of Video 1A, 1B), with no intraoperative complications and minimal blood loss.

Discussion

Castleman's disease is a rare entity with a variety of subtypes described and an unclear etiology. Clinical classification of the disease is based on the number of regions of enlarged lymph nodes [7] and can be described as either unicentric (one region)



Video 1. (**A, B**) A summarized version of the procedure that was conducted in 120 minutes with no intraoperative complications and minimal blood loss, showing the proper resection of the retroperitoneal mass anterior to the right kidney and IVC, which was pushing the duodenum and the head of the pancreas anteriorly and medially.

or multicentric (multiple regions). The multicentric variant (MCD) can be further subclassified based on Human Herpesvirus-8 (HHV-8) status of the patient, as either HHV-8-positive MCD or HHV-8-negative MCD, which itself has 2 variants. POEMS-associated HHV-8-negative MCD presents with polyneuropathy, organomegaly, endocrinopathy, monoclonal plasma cell disorder, and skin changes. Idiopathic HHV-8-negative MCD can either be with or without TAFRO syndrome (thrombocytopenia, anasarca, myelofibrosis, renal dysfunction, and organomegaly). Histologically, CD can be divided into 3 subtypes, the most common being hyaline-vascular (HV-CD) type (76% to 91% of cases), plasma cell (PC-CD) variant (9% to 24% of cases), and a rare mixed type [8]. Our patient had a unicentric disease with a hyaline-vascular histologic subtype. The etiology of UCD is not currently known, but there are 3 hypotheses as to the origin of this disease [7]: viral, neoplastic, and reactive inflammatory mechanisms. The literature has more evidence pointing towards a neoplastic cause, where one study found monoclonality in 76% of UCD cases [9].

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The incidence and prevalence of the disease are unclear; but one study showed the incidence of all forms of CD to be 21 in one million [10], with estimated cases in the United States ranging from 30 000 to 100 000 [3]. UCD mainly occurs in the third and fourth decades of life, with an average age of 35 years, and a slight predilection to females [3]. The location of CD as retroperitoneal is found in 11–12% of cases [3,4], but the literature review we conducted showed only 3 cases of peripancreatic UCD [11,12] that were below the pancreas, and 1 report of duodenal CD [13]. Based on our literature review, the location of the disease in our patient appears to be unique, as the combination of the mass being retroperitoneal, peripancreatic, and para-duodenal has not yet been described. Concerning the presentation of our patient with isolated hematuria, only 2 other case reports were found with similar symptoms [1,14].

Diagnosis of UCD can be challenging, as patients are either asymptomatic or have symptoms related to the mass effect [3], and many cases have been found incidentally on physical exam or imaging [3,14]. Imaging can be useful to delineate the location of the mass and to determine whether there is invasion of any structures, but the diagnosis of retroperitoneal UCD cannot be made based on imaging alone, as it cannot be distinguished from neuroendocrine tumors, lymphomas, or paragangliomas [11]. As in our case, biopsy is usually not diagnostic and generally is not recommended due to the risk of seeding and spread of the tumor, and risk of bleeding in the hyper-vascular mass [3]. However, biopsy is useful when radiologic diagnosis is inconclusive and when a preoperative diagnosis could change the choice of management [8]. The diagnosis of lymphoma had to be ruled out in our patient, as further work-up and management would have changed the decision to remove the mass. Furthermore, EUS-guided biopsy has been proven effective and relatively safe for retroperitoneal masses that are in close proximity to the bowel [15]. Ultrasound, CT, and MRI have all been described for evaluation of retroperitoneal masses and the diagnosis of UCD [11,14]. Ultrasound is useful for assessing the vascularity of the lesion by use of Doppler technique. CT imaging of retroperitoneal UCDs shows a well-defined mass, with a variable morphology, which is usually heterogeneously enhancing with IV contrast [11] and is useful to view calcifications, which are rare in UCD [14]. MRI evaluation typically shows hypointense signaling on T1-weighted images and an increased signal on T2-weighted images [11,14]. Even with these radiologic findings being described in retroperitoneal UCD, diagnosis is still dependent on postoperative histology [7,8,11].

There is a consensus in the literature that surgical resection of UCD is curative, with an extremely low recurrence rate [1,3,4,8,11,14]. The 5-year survival rate ranges between 90% and 100% [4,16], and the 5-year disease-free rate has been reported to exceed 80% [16]. For unresectable UCD, partial resection followed by adjuvant radiotherapy was shown to be effective in a small case series [17]. Other proposed therapies include anti-interleukin 6 (IL-6), which has been implicated in the pathogenesis of CD [1, 7]. Generally, the prognosis of UCD is excellent, with a 10-year mortality rate of only 5% [16]. Laparoscopic approach to retroperitoneal UCD offers a safe and effective method for resection, and it also benefits the patient with a lower rate of postoperative pain and complications [3], which drove our decision to pursue a laparoscopic approach.

Conclusions

Castleman's disease is an uncommon disease with multiple subtypes, an unknown etiology, and a variable presentation. The case presented in this report is unusual in the presentation with isolated hematuria and the unique location of peripancreatic and para-duodenal UCD. Resection of the mass was uncomplicated, and if this case follows the course of UCD described in the literature, then resection was curative. CD is an unusual diagnosis; however, it should always be a differential diagnosis of well-demarcated masses that are either asymptomatic or with symptoms related to mass effect. Further studies are needed to clearly define the etiology of the disease and the best course of treatment for CD patients.

Department and Institution where work was done

Clemenceau Medical Center, Beirut, Lebanon.

Conflicts of interest

None.

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