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Case report

Complete excision and management of large hyaline-vascular pelvic Castleman's disease with coexisting cholecystolithiasis: A rare case report

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ABSTRACT

Introduction: Castleman's disease (CD) refers to scarce lymphovascular hyperplasia categorized as unicentric (UCD) or multicentric (MCD). Retroperitoneal UCD is uncommon (6.7 %) and frequently grows undetected, making diagnosis challenging. The hypervascular nature of hyaline-vascular variant and the average larger size of UCD may result in complete resection difficulty. We present a case of large pelvic UCD completely resected without previous prophylactic procedures or massive intraoperative bleeding with coexisting cholecystolithiasis, which follows the SCARE 2023 guidelines.

Case presentation: A 45-year-old male was accidentally observed with pelvic mass suppressing ureter and right internal iliac artery in preoperative MRI for cholecystolithiasis and cholecystitis. Epigastric pain, vomiting, and jaundice due to the inflamed gallbladder had resolved before mass removal. Biopsy and immunohistochemistry (IHC) tests confirmed the hyaline-vascular CD. Complete resection per laparotomy was done due to lesion adherence and identified a 7x5x2.5 cm solid mass with cystic degeneration suppressing the ureter. After five months, the patient fully recovered with no symptoms.

Discussion: Asymptomatic mass presented as lymphadenopathy on imaging should suggest CD. A prompt lymph node biopsy followed by a pre- or postoperative IHC test is important in diagnosing CD. Total pelvic mass excision with rectum preservation is feasible for larger (>5 cm) hypervascularized masses without remarkable bleeding and promotes recovery without adjunctive treatments.

Conclusion: Complete excision per laparotomy is accessible for the uncommon large pelvic retroperitoneal UCD-suppressing ureter.

1. Introduction

Castleman's disease (CD) represents a rare enlargement of angio-follicular lymphatic tissue, classified as multicentric (MCD) or unicentric (UCD). MCD involves widespread lymph node hyperplasia with more hostile clinical results. UCD appears as a single lymphadenopathy often larger than MCD, commonly observed in the chest and neck [1–4]. Only 6.7 % are retroperitoneal and most UCD was hypervascular, thus the correct approach is important as UCD is considered rare [2–4]. Complete resection is the standard therapy [3,4]. This study described a large hypervascular retroperitoneal UCD of pelvic origin, aligned the SCARE Criteria 2023 [5].

2. Case presentation

A 45-year-old male hospitalized in March 2024 due to intermittent epigastric pain, nausea, vomiting, jaundice, and dark yellow urine. Family, social, medication, and surgical history were unremarkable alongside normal vital signs, except for the icteric sclera. Contrast-enhanced MRI revealed biliary dilatation and multiple gallstones with the largest size of 0.8 × 1 cm. However, a cystic solid mass sized 7.7 × 6.4 × 6.5 cm was also discovered in the right pelvic cavity suppressing

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the ureter and right internal iliac artery suggesting malignancy. The pelvic mass was planned for removal following laparoscopic cholecystectomy. However, the mass was intraoperatively found hypervascularized and adhered tightly to the ureter and right lateral pelvic wall, making removal unobtainable. Therefore, a biopsy was conducted through converted exploratory laparotomy.

Microscopically, numerous atretic follicular centers were distributed across lymphoid tissue surrounded by a broadened mantle zone of lymphocytes in layered, onion-skin patterns [Fig. 1]. Penetrating vessels traversed the germinal centers and hyalinized creating a lollipop appearance. These findings align with the hyaline-vascular (HV) subtype of CD, excluding malignancy. Analysis on immunohistochemistry (IHC) showed CD20 (+), CD3 (+), CD10 (+), CD23 (+), CD34 (+), CD138 (+), MUM-1 (+) Ki-67 (+), IgD (+), bcl-6 (+), bcl-2 (−), Cyclin D1 (−), CD30 (−), AE1/AE3 (−), and kappa/lambda portraying a polyclonal pattern, confirming the HV subtype.

Mass removal was scheduled two months later on May 2024. Prior to surgery, a laboratory workup revealed normal blood count, liver and kidney function, coagulation profile, and electrolytes. Exploratory laparotomy through midline vertical incision was decided after considering the lesion adherence to the ureter and major vessels. Pelvic retractors were used to expose the tumor. Intraoperatively, cystic tissue was discovered in the right lateral pelvic wall apart from the rectum and other intestinal organs, allowing rectum preservation. Bilateral ureteral access sheaths were placed during ureteroscopy to protect ureter patency. Margin determination was challenging, however, no injury to right internal iliac artery and ureter was observed. A passive drain was installed at Cavum Douglas to monitor fluid drainage. The 6-h surgery completely excised the hypervascular mass with gross histopathology confirmed a pelvic mass sized 7x5x2.5 cm with cystic degeneration and firm white surfaces on sectioning [Fig. 2].

Postoperatively, the patient was monitored by a multidisciplinary team involving digestive, oncologic, and urologic surgeons in general inpatient ward. The patient was stable and discharged after five days with an abdominal drain installed and got prophylactic antibiotics, pain relief, and antiemetics as take-home medications, with unremarkable complications. The drain was removed after one week. After five months, no signs of recurrence were reported following routine blood tests and imaging at the hospital [Fig. 3].

3. Discussion

Castleman's disease (CD) was first discovered by Benjamin Castleman in the 1950s [3,4]. Etiology remains unknown, yet studies indicated relations to immune alteration and infection of HHV-8 or HIV [3]. CD can be found in the mediastinum (70 %), abdomen (11 %), retroperitoneum (7 %), or axilla (4 %). Specifically, UCD occurs in 16–20 cases per million annually, emphasizing the rarity of pelvic retroperitoneal UCD, as presented in our study [2,3].

UCD is usually larger than MCD (5.5 versus 3.8 cm), generally manifests in 30s to 40s, and has no preference for either gender [2–4,8]. It is often discovered by chance, while MCD might present with symptoms, including weight loss >10 % per six months, fever, night sweats, and discomfort due to mass compression [4]. There is no proof of either HHV-8 or HIV infection and signs of immune alteration in our case, except for jaundice, icteric sclera, and dark yellow urine which are pathologically related to cholecystolithiasis. Several studies have reported CD coexisted with hepatobiliary diseases. Jiang et al. discovered CD adhered densely to common bile duct during operation for choledocholithiasis [6]. Dhakre et al. reported CD in the porta hepatica of a female with gallbladder cancer. A prevailing hypothesis suggests a low-level inflammation induced by chronic cholecystitis triggered a localized increase of IL-6, resulting in CD to nearby lymphatic tissue [7]. Although our case presented with symptoms related to inflamed gallbladder, CD-related symptoms were not discovered and could be considered asymptomatic [4,8].

Early diagnosis is challenging as UCD frequently grows undetected and presents with normal laboratory tests, while abnormalities indicate MCD. Systemic inflammation of MCD causes anemia through hepcidin-mediated disruption of iron metabolism, hypoalbuminemia by impairing liver albumin synthesis, and hypergammaglobulinemia through immunoglobulin overproduction [4,8–11]. Our case initially showed abnormalities related to inflamed gallbladder. However, after cholecystectomy, normal laboratory tests before mass removal revealed a suggestive UCD.

Definitive diagnosis typically follows lymph node biopsy [8]. Histopathologically, CD is classified as plasma cellular (PC), hyaline-vascular (HV), and mixed variants. Most UCDs present as HV (70–80 %) characterized by follicular enlargement with dysplastic follicular dendritic cells (FDC) and atretic germinal centers portraying twinning or

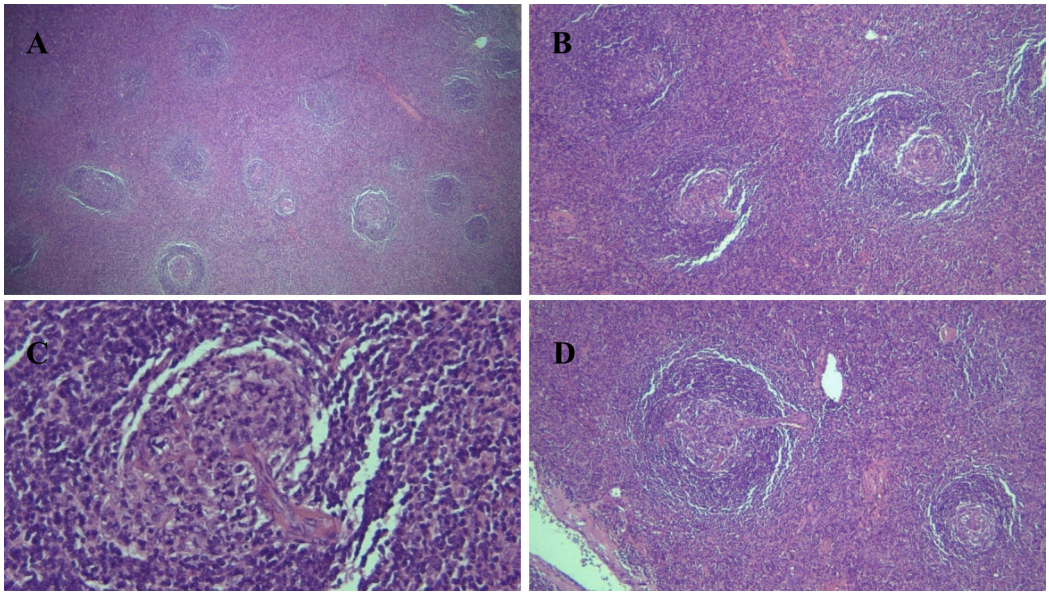

Fig. 1. Histopathological examination of biopsied mass.
(A: lymph node with atretic follicles and thickened mantle zones HE 40×; B: thickened mantle zones HE 100×; C: atretic germinal center with sclerotic and hyalinization of vascular HE 400×; D: lollipop follicles HE 400×).

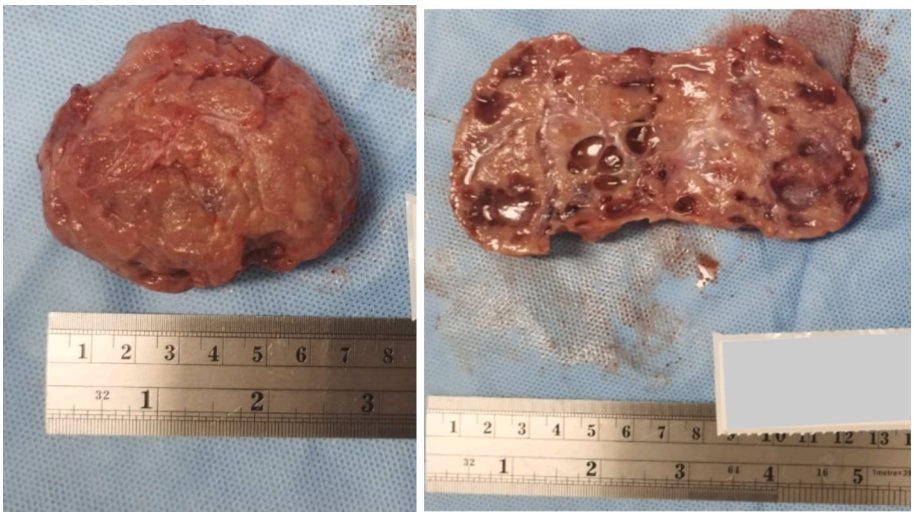

Fig. 2. Complete excision of pelvic mass.
(A: tissue sized 7x5x2.5 cm completely excised; B: firm cystic degeneration and white surface on sectioning).

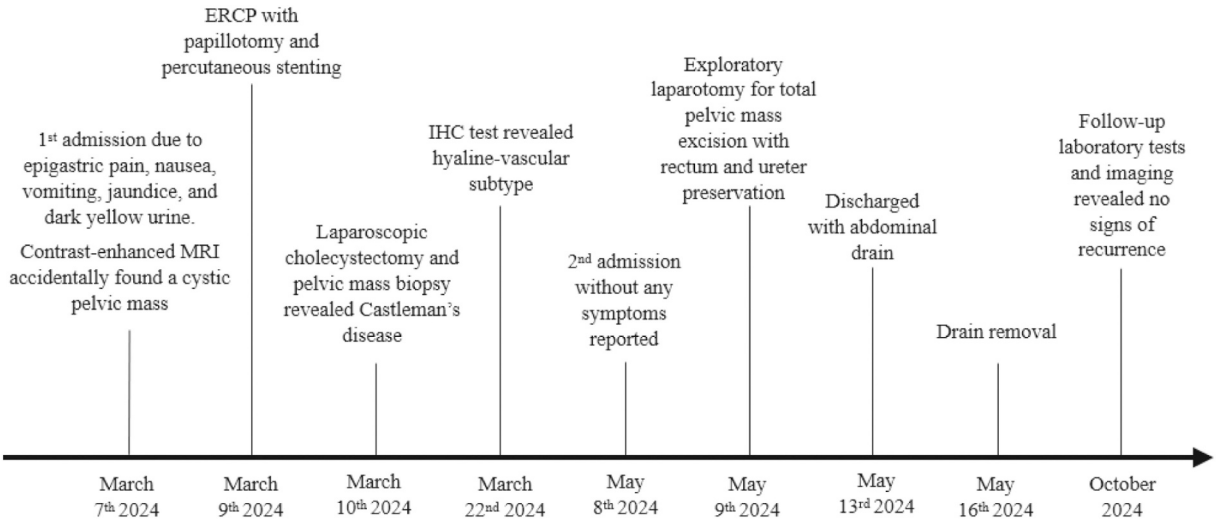

Fig. 3. Timeline of case presentation.

budding patterns. The expanded mantle zones exhibit onion-skin layers, while radial hyalinized blood vessels create lollipop appearances, as discovered in our patient [4,8,12,13]. PC subtype is characterized by Russell bodies, plasmacytosis, and enlarged lymphoreticular nodules, whereas mixed subtypes exhibit both HV and PC features [8,12,13]. However, numerous diseases share characteristics similar to CD. Thymomas or lymphomas may overlap with HV. Syphilis or EBV infection, autoimmune, immunodeficiencies, and lymphomas also resemble PC [8,13]. Therefore, we conducted an IHC test to provide detailed information.

HV on IHC shows depleted follicular centers with B cells within enlarged mantle zones, confirmed by B-cell markers (CD20 and MUM1) and IgD expression. While some dispersed polyclonal plasma cells may be found throughout the lymph node, this is not a notable feature of HV [13]. Our case showed expression of CD20, MUM1, IgD, and polyclonal pattern aligned with HV. Radial blood vessel distribution was marked by CD34, while dysplastic dendritic cells were highlighted by FDC antigens (CD21 and CD23), as presented in our case [13,14]. High Ki-67 in central follicle and low in interfollicular section indicates high germinal cell proliferation confirming follicular hyperplasia of HV. The absence of cyclin D1 and bcl-2 further supports this conclusion, as these are associated with lymphoma [14].

In contrast-enhanced MRI, CD appears as well-demarcated enhancing mass persisting during the venous phase owing to vessel hyalinization but decreases in the later phase as contrast escapes into the extravascular space. This reduction aids in distinguishing CD from neuroendocrine tumors. CT-positron emission tomography (PET) shows low uptake while high uptake may suggest lymphoma [4,8]. We discovered a well-defined pelvic mass suggestive malignancy. The definitive diagnosis was confirmed through biopsy and IHC.

The optimal therapy for UCD involves total mass excision as previous studies described R0 resection margin is associated with 5-year and 10-year survival rates of 100 % and 95 %, respectively [1,4,8,15,16]. No consensus exists regarding better choices between minimally invasive and open surgery in achieving complete resection [4,17]. Either laparoscopy or laparotomy has been associated with no recurrence after 6 and 27 months, respectively [18,19]. Total mass excision per laparotomy was performed after considering lesion's hypervascular nature and adherence to ureter and major vessels.

Treatment strategies should involve a multidisciplinary team [4,8]. Consider the possibility of perioperative bleeding in mass $>$ 5 cm, particularly UCD with an average larger size [8,18]. UCD is also linked

to hypervascularity and pelvic mass commonly exhibits fibrous adhesion to surrounding tissues, therefore, some studies recommended preoperative embolization to enhance surgical safety [3,8,18,19]. Kitakaze et al. reported an uneventful clinical course and no recurrence in 21 months following preoperative embolization of pelvic mass with middle sacral artery as feeding vessel [18]. However, our case was previously measured 7.7 × 6.4 × 6.5 cm on MRI, sized 7x5x2.5 cm upon excision, and was successfully evacuated without prophylactic procedure and massive bleeding intraoperatively. Any incidental damage to vessels and ureters was minimized through vascular loops and ureteral sheaths, whereas rectum preservation was done through careful dissection and gentle traction. Postoperative complications were unremarkable, yet the patient got routine medications aligned with grade I Clavien-Dindo Classification [20]. During follow-up, the patient mentioned that the surgery resulted in psychological relief and improvement in daily activity as less fatigue was reported, highlighting a safe yet successful surgery for a large UCD.

Our strength focused on large hypervascular pelvic UCD completely excised without prophylactic procedure, guiding future surgical approaches for similar cases. However, since findings are based on a single patient, it limits generalizability to broader populations with no direct comparison to other managements. Therefore, further investigation involving multiple characteristics could be undertaken.

4. Conclusion

Complete excision per laparotomy is accessible for large (> 5 cm) pelvic UCD-suppressing ureters without any prophylactic procedure. It is also crucial to involve CD as potential diagnosis of pelvic mass. Further research is essential to elucidate varied clinical manifestations and best treatment approaches for CD.

Ethical approval

Ethics approval is not required for case reports or case series as these are not categorized as research involving human subjects according to our institutional guidelines. It generally focuses on individual patient cases and is viewed as part of clinical practice.

Registration of research studies

1.Name of the registry: N/A.
2.Unique identifying number or registration ID: N/A.
3.Hyperlink to your specific registration (must be publicly accessible and will be checked): N/A.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Authorship contribution

Wifanto Saditya Jeo and Dismas Adiasa Chaspuri performed the surgery.

Wifanto Saditya Jeo, Samuel Haryono, Fajar Lamhot Gultom, and Rachmat Christian Nikijuluw performed conceptualization, methodology, resources, and supervision.

Wifanto Saditya Jeo, Fajar Lamhot Gultom, and Safira Farah Yoladifa performed data curation, visualization, and investigation.

Wifanto Saditya Jeo and Safira Farah Yoladifa performed writing – original draft and writing – review & editing.

All authors read and approved the final manuscript.

Guarantor

Wifanto Saditya Jeo is the guarantor of this study.

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Declaration of competing interest

All authors declared no conflicts of interest associated with this publication.

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