



Oncology

## Surgical challenges of Castleman's disease of the pelvis

Sat Prasad Nepal<sup>\*</sup>, Takeshi Shichijo, Yoshio Ogawa, Michio Naoe, Kazuhiko Oshinomi, Jun Morita

Department of Urology, Showa University, School of Medicine, Japan

### ARTICLE INFO

#### Keywords:

Castleman's disease  
Pelvis  
Adhesion  
Vascularity

### ABSTRACT

Castleman's disease (CD) is a unique lymphoproliferative disorder. It commonly occurs in the mediastinum, neck, axilla, and abdomen, and retroperitoneal involvement is rare. Here we report a unique case of CD in the pelvis. Laparotomy was performed and surgery was complicated by adhesions and vascularity. Total surgical duration was Five hours and 45 min with 4.5 L of blood loss. Ten pints of blood was transfused. The mass was histopathologically diagnosed as hyaline-vascular CD. The patient was free of recurrence after 10 years of follow-up.

### Introduction

Castleman's disease (CD) is a unique benign lymphoproliferative disorder originally discovered by Castleman in 1954.<sup>1</sup> It can be classified into unicentric and multicentric types based on morphology, or hyaline-vascular (HV) and plasma-cell (PC) types based on histology.<sup>2</sup> There is also a plasmablastic variant of CD, which is observed in human herpesvirus 8 (HHV-8)-positive cases.<sup>1,2</sup> CD commonly occurs in the chest,<sup>1-4</sup> and CD in the pelvic region is rare, with an incidence of 2%.<sup>2</sup> Here we report on the difficulties faced during surgery and outcome of a unique case of unicentric retroperitoneal pelvic CD.

### Case presentation

A 41-year-old male was admitted to the surgical ward for abdominal pain and vomiting. He underwent emergency operation for incarcerated hernia with intestinal obstruction. Preoperative CT scans revealed a retroperitoneal tumor on the left side of the bladder measuring 90 × 70 mm, with a contrast effect. MRI revealed a T1 hypointense, T2 hyperintense lesion (Fig. 1 A and B).

The mass was initially considered a pelvic retroperitoneal tumor. CT needle biopsy showed marked lymphocyte infiltration with T cell markers (CD3<sup>+</sup>, CD5<sup>+</sup>). Open biopsy was suggested by the pathology department, and revealed CD10 (+), TIA-1 (+), CD56 (+), and BCL2 (+).

Elective surgery was performed to remove the retroperitoneal pelvic

tumor (Fig. 2). During laparotomy, the tumor was found to resemble lymph node enlargement near the internal iliac artery compressing the bladder. The tumor was strongly adhered to the pelvic cavity, and an attempt was made to dissect the tumor off the bladder and internal iliac artery. During dissection, the feeding artery of the tumor was torn and bleeding occurred, requiring consultation with the vascular surgery department. The torn vessel was ligated and angioembolization was not required. No bladder damage was noted after tumor removal. The ureter was difficult to locate and was confirmed by intravenous indigo calamine infusion, which flowed out of the ureteral ostium.

Total surgery duration was 5 hours 45 minutes with blood loss of 4.5 L. Eight pints of Red cell concentrates (RCC) was transfused during surgery and Two pints in Surgical Intensive care unit (SICU). Patient had no post-operative problems and was discharged on the seventh post-operative day.

On histopathology, lymphoid follicle proliferation around the germinal center was observed, with blood vessel growth in the perifollicular region and the germinal center (Fig. 3).

The resected specimen was positive for T cell markers (CD3 CD5), a B cell marker (CD20), B cell and plasma cell markers (CD79a, CD138), and a plasma cell marker (VS38c), as assessed by immunohistochemistry. K lambda positive cells were also observed. The patient was HHV-8-negative.

After discharge patient had CT investigation every six months with no abnormalities. The patient remained recurrence-free after 10 years of follow-up.

<sup>\*</sup> Corresponding author. Department of Urology, Showa University, School of Medicine, 1-5-8 Hatanodai, Shinagawa-Ku, Tokyo, 142-8555, Japan.

E-mail addresses: [satprasad1@med.showa-u.ac.jp](mailto:satprasad1@med.showa-u.ac.jp) (S.P. Nepal), [shichijo@med.showa-u.ac.jp](mailto:shichijo@med.showa-u.ac.jp) (T. Shichijo), [ogawayos@med.showa-u.ac.jp](mailto:ogawayos@med.showa-u.ac.jp) (Y. Ogawa), [oshikazu@med.showa-u.ac.jp](mailto:oshikazu@med.showa-u.ac.jp) (K. Oshinomi), [moritajun@med.showa-u.ac.jp](mailto:moritajun@med.showa-u.ac.jp) (J. Morita).

<https://doi.org/10.1016/j.eucr.2020.101518>

Received 2 November 2020; Accepted 30 November 2020

Available online 1 December 2020

2214-4420/© 2020 Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

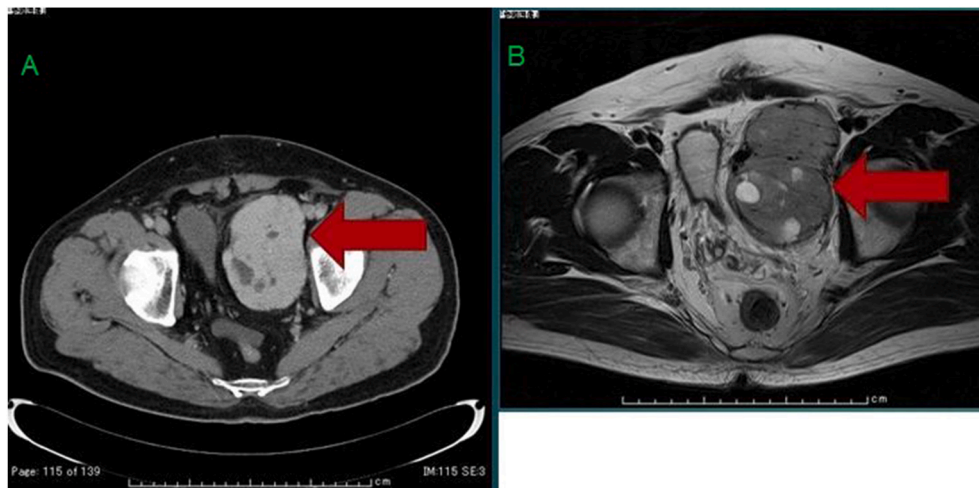


Fig. 1. A and B: CT of Pelvic mass (A) and MRI of Pelvic mass (B).



Fig. 2. Gross specimen of Pelvic tumor.

## Discussion

HVCD accounts for 90% of all CD cases. It is generally asymptomatic, with a median age of diagnosis of 40 years.<sup>1</sup> The PC variant is less common, occurring in  $\leq 10\%$  of cases. HVCD is mostly associated with unicentric disease.<sup>3</sup> According to a systematic review, histopathological classification is unlikely to be important for long-term survival outcome.<sup>3</sup>

Multicentric disease is characterized by systemic symptoms and is associated with Human Immunodeficiency Virus (HIV) infection, Kaposi's sarcoma, and POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal proteins, and skin changes).<sup>2</sup> Moreover, multicentric disease increases non-Hodgkin's lymphoma risk in patients with HIV infection.<sup>2</sup> As for the etiology of CD, interleukin-6-mediated inflammation in response to HHV-8 infection or unknown factors is believed to play a role.<sup>2</sup>

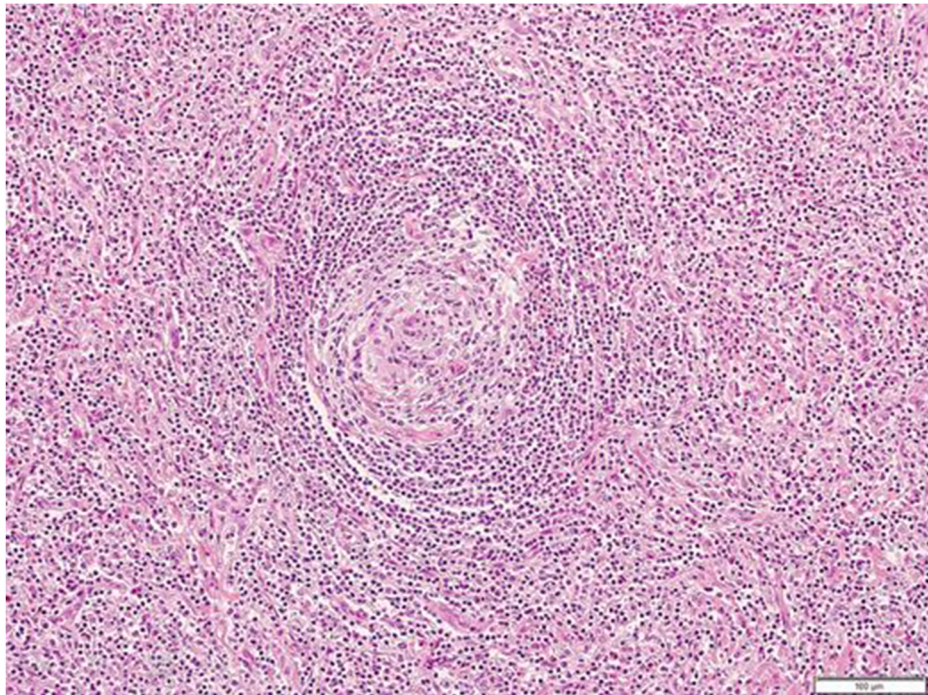
Radiography is not completely reliable in diagnosing CD. Castleman's tumor presents as hypoechoic lesions in ultrasonography. CT scans show homogenous enhancement of the lesions if they are smaller

than 5 cm and heterogenous enhancement if they are larger than 5 cm. Calcification (arborizing, punctate, coarse, peripheral) is also reported in 31% of cases.<sup>4</sup> On MRI, T1 lesions are hypodense, whereas T2 lesions are hyperdense.<sup>4</sup>

Histopathology is necessary for the confirmatory diagnosis of CD. HVCD shows damaged medullary sinuses, lymphoid follicle proliferation around the germinal center in the mantle region (onion-skinning), and blood vessel growth in the perifollicular region (lollipop sign) and the germinal center. On the other hand, PCCD is characterized by patent medullary sinuses, less blood vessel growth, and interfollicular plasma cells.<sup>2,3</sup> Tests for IL-6 and HHV-8 levels, although helpful, are not routinely carried out.

Multicentric CD requires systemic treatment and surgery is reserved for cases in which adverse effects on organs are observed due to enlarged lymph nodes.<sup>1</sup> Multicentric CD is associated with poor outcomes, with frequent recurrences.<sup>1,2</sup> On the other hand, in cases of unicentric CD, complete excision of the tumor results in excellent outcomes.<sup>1,2</sup> Further if surgery cannot be performed, radiotherapy is another option.<sup>2</sup>

Our case presented challenges during surgery due to adhesions and



**Fig. 3.** Histopathology of the Pelvic mass.

vascularity, with an operative duration of more than 5 h. There was 4.5 L of blood loss requiring ten pints of blood transfusion. Thus, we have to anticipate the complications during surgical removal of the tumor.

#### Conclusion

In this report, we described a rare case of retroperitoneal pelvic CD. It was complicated by adhesions and vascularity.

#### Consent

The authors certify that they have obtained all appropriate patients consent forms. In the form, the patients have given their consent for the images and other clinical information to be reported in the journal. The patients understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

#### Funding information

No funding was received for this article.

#### Declaration of competing interest

No conflict of interest exists.

#### Abbreviations:

|       |                                    |
|-------|------------------------------------|
| CD    | Castleman's disease                |
| HV    | Hyaline-vascular                   |
| PC    | Plasma-cell                        |
| HHV-8 | Human herpesvirus 8                |
| USG   | Ultrasonography (USG)              |
| HIV   | Human Immunodeficiency Virus (HIV) |
| IL-6  | Interleukin-6                      |

#### References

1. Castleman B, Towne VW. CASE records of the Massachusetts general hospital weekly clinicopathological exercises: case 40011. *N Engl J Med.* 1954;250:26–30. <https://doi.org/10.1056/NEJM195401072500107>.
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. [https://doi.org/10.1002/1097-0142\(197203\)29:3<670::AID-CNCR2820290321>3.0.CO;2-#](https://doi.org/10.1002/1097-0142(197203)29:3<670::AID-CNCR2820290321>3.0.CO;2-#), 29: 670-683.
3. Talat N, Belgaumkar AP, Schulte KM. Surgery in Castleman's disease: a systematic review of 404 published cases. *Ann Surg.* 2012;255:677–684. <https://doi.org/10.1097/SLA.0b013e318249dcdc>.
4. Meador TL, McLarney JK. CT features of Castleman's disease of the abdomen and pelvis. *AJR Am J Roentgenol.* 2000;175:115–118. <https://doi.org/10.2214/ajr.175.1.1750115>.