

CASE REPORT

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# Unicentric castlemans disease located in the left popliteal fossa: a case report

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## Abstract

**Background:** Castleman disease (CD) is a lymphoproliferative disease of unknown etiology, it can affect any lymph nodes of the body but rarely affects the popliteal fossa.

**Case presentation:** We present a 67-year-old woman with touching solitary painless mass in the left popliteal fossa for one week. Imaging showed multiple soft-tissue masses of different sizes in the left popliteal muscle space, the T1 weighted image showed hypointense to isointense, the fat-suppressed T2 weighted images showed subtle hypersignal intermingled with linear of hypointense, and displayed homogeneous contrast enhancement after administration of gadolinium. Complete surgical resection was performed. Pathologically demonstrated plasma cell type CD.

**Conclusion:** We described a rare case plasma cell type of UCD located in the popliteal fossa which might help to enrich the clinical spectrum of this rare site and unique subtype of UCD. This case illustrates that CD should be considered in the differential diagnosis of every hypervascularity soft tissue tumor in any anatomic location, especially when they occur in the region of lymph node distribution.

**Keywords:** Castleman disease, Lower extremity, Case report

## Background

Castleman disease (CD), also known as giant lymph node hyperplasia, was first described by Benjamin Castleman in 1954. It is a lymphoproliferative disease of unknown etiology, which can occur in any area where lymphoid tissue is normally found, but rarely affects the popliteal fossa. Pathologically, CD is divided into hyaline-vascular, plasma cell variants or mixed type, hyaline-vascular is more common. We report an unusual case of a 67-year-old female with a plasma cell type of an unicentric CD in the left popliteal fossa.

## Case presentation

A 67-year-old female patient was admitted to the hospital for one week after touching solitary painless mass in the left popliteal fossa. Her past medical history had

been uneventful, and her family history exhibited no malignancies.

**Physical examination:** A round soft tissue nodule was touched behind the left knee joint, approximately 3.0 cm in size, with mild tenderness and mobility. Physical examinations of the chest and abdomen were unremarkable.

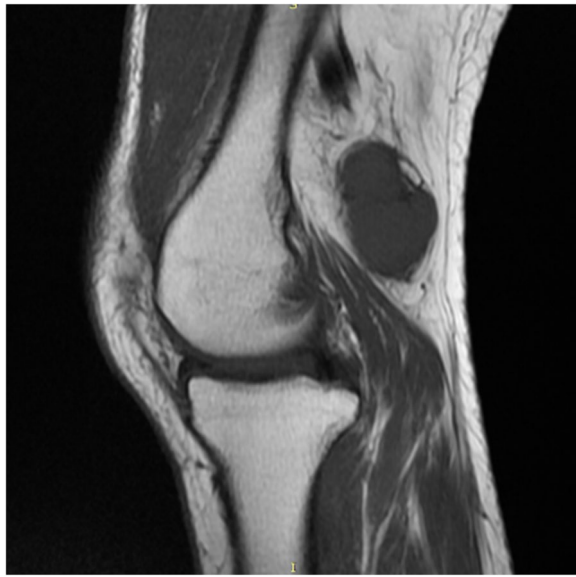
**Laboratory examination:** uric acid 488.0  $\mu\text{mol/L}$  (normal reference value 255~357  $\mu\text{mol/L}$ ), other laboratory examinations showed no abnormalities.

**Imaging examination:** Ultrasound showed a solid hypoechoic mass of 3.2 cm  $\times$  2.4 cm in the left popliteal fossa, with blood flow visible, it was hard to determine whether the lesions are benign or malignant. An unenhanced MRI scan showed multiple soft-tissue masses of different sizes in the left popliteal muscle space. The T1 weighted image showed hypointense to isointense, the fat-suppressed T2 weighted images showed subtle hypersignal intermingled with linear of hypointense, and the larger lesion was an oval shape, lesion size was 3.2 cm  $\times$  1.9 cm  $\times$  2.1 cm, smooth edges, and displayed homogeneous contrast

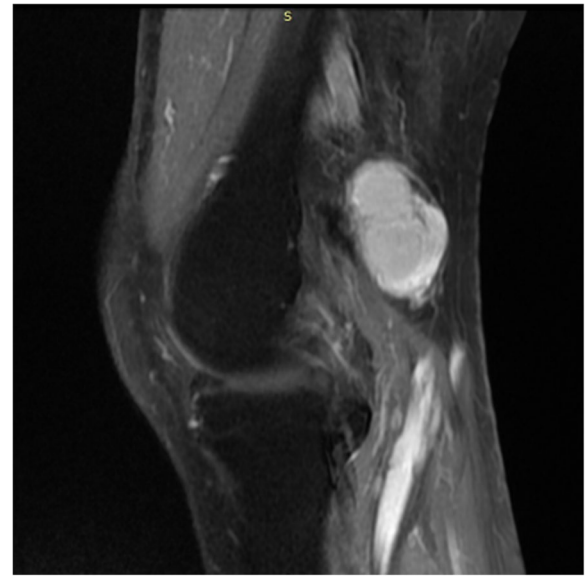
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**Fig. 1** MRI T1 weighted image (sagittal view) showed a hypointense to isointense mass in the left popliteal fossa



**Fig. 3** MRI fat-suppressed gadolinium-enhanced T1-weighted image (sagittal and Coronal view) show a sharply demarcated mass with avidly homogeneous enhancement

enhancement after administration of gadolinium; two small lesions with similar signal and enhancement degree can be seen in the muscle space above the lesion (Fig. 1, 2, 3 and 4).

Preoperative biopsy revealed a lymphoid tumor of unknown dignity, routine histopathological examination should be performed.

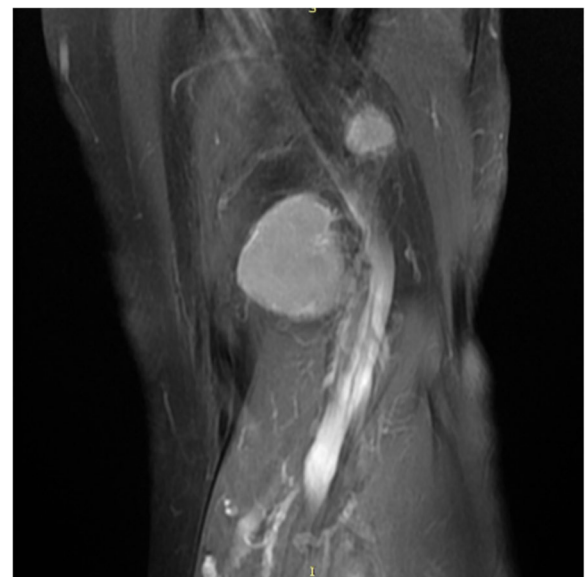
Surgical and pathological findings: Complete surgical resection was performed and three soft nodules having a maximum diameter of 3.2 cm were removed, presented

as a sharply demarcated mass lesion. The popliteal arteries and veins were intact.

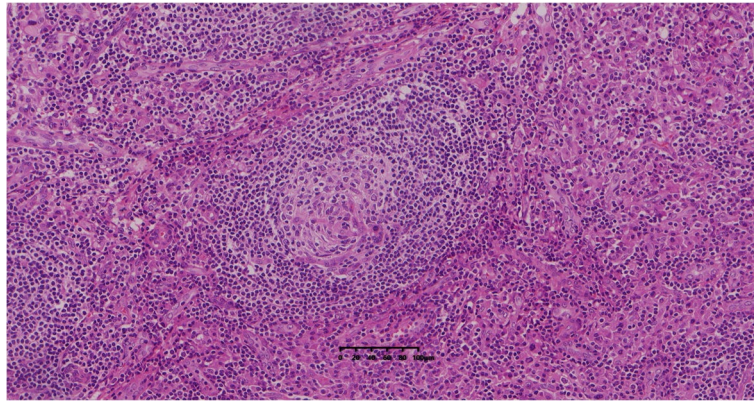
Postoperative pathology: Grossly, the resected tumor specimen displayed a sharply demarcated and medium hardness mass lesion with a gray-red cut surface. Microscopically, the lymph node structure was basically present, the envelope was intact, the lymphatic follicles in the lymph nodes were hyperplastic, the



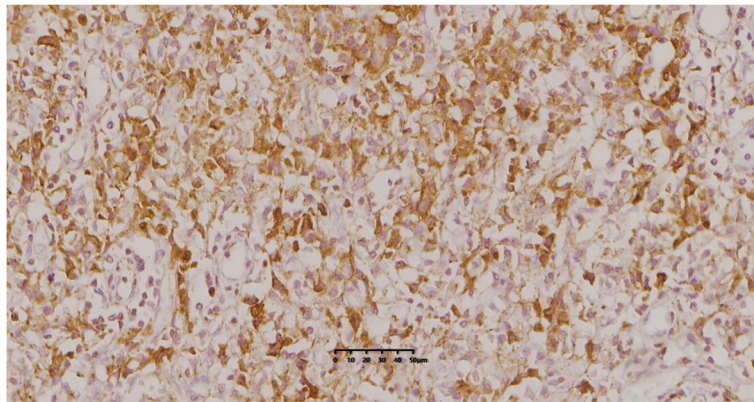
**Fig. 2** Fat-suppressed T2 weighted image (sagittal view) showed a hyperintense lesion, the size was 3.2 cm × 1.9 cm × 2.1 cm



**Fig. 4** Coronal view shows multiple enhancing soft-tissue mass in the left popliteal fossa



**Fig. 5** Microscopically, a large number of plasma cell infiltrates were seen in the interfollicular area (H&E, 200 ×)



**Fig. 6** Immunohistochemistry with a primary monoclonal antibody to CD138 shows lymph node infiltration by plasma cells (syndecan-1, 400 ×)

small blood vessels showed varying degrees of hyperplasia, and a large number of patchy, dense plasma cell infiltrates were seen between the follicles (Fig. 5). Immunohistochemical staining: CD3 interfollicular areas (+), CD43 interfollicular areas (+), Pax-5 follicular areas (+), germinal center (Bcl-2-, Ki-67 >70%), extra-germinal center (Bcl-2 +, Ki-67 about 5%), CD10 germinal center (+), CD34 vascular (+), CD21 FDC (+), plasma cells. CD138 (+) (Fig. 6),  $\lambda$  (+) >  $\kappa$  (+). The morphological examination was performed using Carl Zeiss Axio Lab.A1. The slides were scanned by using the KF-PRO-005 digital pathology scanner (KFBIO85 company, Ningbo City, China).

Pathological diagnosis: (Multiple left popliteal fossa) Castleman's disease, plasmacytic type.

Postoperative recovery was good and discharged. The patient documented in this report was doing well on 12-month follow-up.

## Discussion and conclusions

Castleman disease (CD) is generally regarded as a benign condition, it is more frequent in women with a median age at diagnosis in the third or fourth decade. Clinically, CD is classified as unicentric CD (UCD) and multicentric CD (MCD) based on anatomical distribution. Unicentric CD tends to be asymptomatic or present with mild symptoms. Multicentric CD can be severely or life-threatening. Recently, a novel clinical classification was mentioned, patients who have more limited lymph node involvement and are referred to as having "regional" or "oligocentric" CD [1].

According to histopathological characteristics, CD can be classified as a hyaline vascular, plasma cell, or mixed type, and the incidence rate is 72%, 18%, and 10%, respectively [2]. UCD affects multiple lymph nodes throughout the body, over 70% of patients with UCD present with the disease in the thorax, with the majority of the cases seen in the mediastinum [3]. Here, we reported an unusual



case of UCD located in the lower extremity, an extremely rare site of the disease.

To the best of our knowledge, only a few cases of lower extremity CD have been reported in English literature [2, 4, 5]. Pathological findings for previously reported cases have included hyaline vascular CD and mixed cellularity CD. In the present case, pathologically demonstrated plasmacytic cell type CD, a finding which is rarely reported in the popliteal fossa.

Since the clinical signs and symptoms of UCD are often nonspecific, making them easy to miss or misdiagnose. Lesions in the popliteal fossa require careful evaluation because a number of non-neoplastic and neoplastic lesions can mimic this entity, diagnosing CD without pathological findings is difficult. Generally, the unenhanced CT/MRI scan of CD shows a nonspecific lobulated soft tissue mass, a well-defined border and clearly delineated from adjacent structures, associated with an intact envelope of the lesion. The characteristic image shows that CD is homogeneous enhancement of the lesion, usually without necrosis liquefaction or hemorrhage, and the hypervascularity soft tissue tumor are associated with the proliferation of small and medium-sized blood vessels in the tissue of the lesion [5]. Although the relatively rare plasmacytic cell type UCD should demonstrate less intense enhancement, however, given the intense lymph node enhancement seen in plasmacytic cell type UCD, it is intuitive that plasmacytic cell type UCD would also avidly enhance [3]. Radiological differential diagnosis of solitary hypervascularity soft tissue tumor includes vascular tumors, extrapleural solitary fibrous tumors, lymphoma, soft tissue sarcoma and metastatic tumor [2].

The guidelines suggest that UCD should be managed in the first-line setting with surgery in both children and adults. Complete surgical excision will usually eliminate any systemic symptomatology and laboratory abnormalities, if present [6]. In cases of unresectable disease, aggressive local therapy with radiation should be considered for patients with symptoms or as consolidation after systemic therapy. Asymptomatic patients may be suitable for observation [7]. Oligocentric CD should be managed more like UCD [1].

This report also has some limitations. First, there was no systemic imaging was performed to look for multicentricity, but notably there were no physical findings suggestive of multicentricity. Second, UCD is virtually always HHV-8-, but rare positive cases have been reported [6], unfortunately, no relevant viral testing was performed in this case.

In conclusion, we have described a rare case plasmacytic cell type of UCD located in the popliteal fossa which might help to enrich the clinical spectrum of this rare site and unique subtype of UCD. This case illustrates that CD should be considered in the differential diagnosis of every hypervascularity soft tissue tumor in

any anatomic location, especially when they occur in the region of lymph node distribution.

#### Abbreviations

CD: Castleman disease; UCD: Unicentric castleman disease.

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#### Authors' contributions

Analysis and interpretation of patient data and literature review were done by HJL, HWZ guided and corrected the final manuscript. All authors read and approved the final manuscript.

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#### Availability of data and materials

All data generated or analysed during this study are included in this published article.

#### Declarations

##### Ethics approval and consent to participate

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the principles of the 1964 Declaration Helsinki and its later amendments or comparable ethical standards. Voluntary written informed consent was obtained prior to the study from the patients.

##### Consent for publication

Written informed consent for publication of this case report and any accompanying images was obtained from the patient.

##### Competing interests

All authors declare that they have no competing interests.

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