Case Report

Unicentric Castleman’s disease in the female pelvis

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\textbf{Abstract}

Castleman’s disease (CD) or angiofollicular lymphoid hyperplasia is a rare disorder with unknown aetiology that can easily be misdiagnosed as lymphoma, neoplasm, or infection. Diagnosis is challenging due to its non-specific symptoms and radiologic signs as well as its rarity. We report a case of a middle-aged woman with a mass adjacent to the uterus that was accidentally detected by ultrasound; it was believed to be of ovarian origin. Subsequently, the patient underwent a successful tumorectomy. Pathological examination confirmed the hyaline–vascular type of CD in a pelvic location. This was a typical case of an asymptomatic unicentric and hyaline–vascular type of CD.

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Introduction

Castleman’s disease (CD), also known as giant lymph-node hyperplasia and angiofollicular lymphoid hyperplasia, is a rare benign disorder of uncertain aetiology characterised by the proliferation of lymphoid tissue. It can occur in any lymph node, although it is usually found in the chest, neck, and abdomen; its occurrence in the pelvis is infrequent [1]. However, establishing a preoperative diagnosis can be very challenging due to its nonspecific imaging signs and its rarity; most cases have been diagnosed based on postoperative pathological examination [2]. Here, we present a case of unicentric CD localised in the pelvic retroperitoneum of a 44-year-old female patient who underwent curative surgical resection.

Case report

A 44-year-old woman underwent an abdominopelvic ultrasound examination as part of a routine health check-up, and several uterine fibroids and a mass located at the right side of the uterus, suspected to be an ovarian tumor, were detected. She did not show symptoms such as pelvic pain, gynecologic problems, fever, fatigue, or weight loss, and she had no

Abbreviations: CD, castleman’s disease; MRI, magnetic resonance imaging; PET, positron emission tomography; 18F-FDG, 18F-fluorodeoxyglucose.

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family history of the condition. She underwent transvaginal ultrasonography, which showed an echogenic heterogeneous and well-defined solid mass measuring $42 \times 39 \times 25$ mm within the right pelvic fascia between the uterus and iliac vessels; there was hypervascularity, especially at its periphery, on color Doppler (Fig. 1). Note the confirmed multiple uterine fibroids with no free flow of fluid into the recto-uterine pouch.

Physical examination did not include the palpation of the swollen lymph nodes in the neck, axillaries, and inguinal area. Blood tests, including CA-125 and CA-19.9, showed no evidence of inflammation, infection, or malignancy. Chest X-ray also revealed no abnormal findings. For more information, magnetic resonance imaging (MRI) 3.0 Tesla showed a mass within the right extraperitoneal space similar to that shown on ultrasound, which was an enhanced solid tissue (Fig. 2). In addition, the uterus had several parietal uterine fibroids of different sizes. No other mass or enlarged lymph nodes were observed along the iliac vessels.

The patient successfully underwent partial hysterectomy through a lower midline incision to remove the multiple fibroids and the tumor, which was considered benign. The mass was located on the right parietal fascia, easily dissected without bleeding, and there were no findings of tumor invasion of the adjacent organs and structures. Both ovaries were conserved. No other enlarged lymph nodes were found at the pelvic retroperitoneum. Microscopic examination of the mass confirmed the diagnosis of a hyaline–vascular variant of CD (Fig. 3). There was no evidence of local recurrence or systemic disease within 5 years after diagnosis.

Discussion

CD, also known as giant lymph-node hyperplasia or angiofollicular lymph-node hyperplasia, is a rare benign disorder with unknown aetiology [3]. Based on the number of sites with enlarged lymph nodes, CD is categorised into unicentric and multicentric, which are important for clinical presentation and determining therapy [4]. Two principal histopathological types of CD have been described as the hyaline–vascular and plasma-cell. The hyaline–vascular type is the most common, accounting for 76%-90% of unicentric CD cases [5]. According to Talat, it accounts for 78% of unicentric and 18% of multicentric CDs [4].

They can occur in any lymph nodes, but unicentric CDs usually occur in the chest (29%), neck (23.5%), abdomen (21%), and retroperitoneum (16.5%) and rarely in the axilla (5.5%) and groin (2.5%); its occurrence in the pelvis is the rarest (2%) [4]. Recently, Nakata et al. reported that only 15 cases of unicentric CD in the female pelvic retroperitoneum were reported in the English literature from 1962 to 2019; the paraaortic vessel was the most common disease site (53%) [6].

Unicentric CD most commonly presents in people aged 30-40 years (mean, 34 years; range, 2-84 years), with slight female predominance (1.4:1) [4]. Most patients with unicentric CD are asymptomatic, and it is usually accidentally discovered during an ordered imaging study to investigate a different medical condition. Their presentation of systemic symptoms, including Type B symptoms (fevers, night sweats, and weight loss), and abnormal laboratory findings (anemia, hyperggammaglobulinaemia, hypoalbuminemia, and elevated sedimentation rate) are rare, but such symptoms and findings can be more frequently observed in multiple CD and plasma-cell types during the fifth to sixth decade of life, with a modest higher prevalence in males (1.6:1) [4]. Furthermore, a combination of these hyaline–vascular types, solitary and asymptomatic, are more commonly diagnosed in patients with pelvic CD, which is prevalent in 86.4% [6].

Ultrasoundography generally shows a focal hypoechoic mass with a small hyperechoic centre [6,7], with peripheral hypervascularity on colour Doppler [5]. Classic computed tomography findings include localised nodal masses that are well-defined, homogeneous, or partially heterogeneous, with marked or moderate contrast enhancement and an imaging appearance mimicking retroperitoneal adenopathy [5-8]. Additionally, in the central mass, low attenuation due to central fibrosis or necrosis can be unusual, appearing to be of a diameter longer than 5 cm; it is accompanied by punctate calcifications in 31% of cases [5,8,9]. Peripheral rim-like enhancement at the early phase of an enhanced CT scan or a local peritoneal thickening around the lesion may also be found [10].
Fig. 2 – Pelvic MRI shows the mass (arrow) that is nearly isointense to the normal uterine wall with relatively heterogeneous enhancement. Multiple fibroids were observed in the large uterus. T2W axial (a), T1W fats at axial (b), with gadolinium axial (c) and coronal (d).

Fig. 3 – Hematoxylin-eosin stain at magnifications of ×50 (A) and ×200 (B) show a hyaline–vascular variant of Castleman’s disease. The lymph follicles (A, arrows) show small lymphocytes concentrically forming an “onion-skin” pattern, surrounding the germinal centre with hyalinised blood capillaries spreading towards the centre (B, arrows).

MRI findings generally show a mass that is relatively isointense to the skeletal muscle as hypointense on T1W and hyperintense on T2W and contrast enhancement after gadolinium; flow voids showed feeding vessels, but these characteristics are non-specific [5,6,8,11]. MRI is suitable for evaluating the extent of the disease and its relationship with adjacent structures, although with limited evaluation of calcifications. In the present case, we concluded that the mass was located within the retroperitoneal pelvic region and not the ovary based on the MRI findings.

Whole-body positron emission tomography is used to assess the metabolic status of the enlarged lymph nodes suspected for CD, but that can be misdiagnosed as lymphoma or metastatic adenopathy [8]. 18F-fluorodeoxyglucose uptake is usually lower than what is observed in active lymphomas [8]. The 18F-fluorodeoxyglucose-positron emission tomography facilitates high metabolic uptakes even in nonenlarged lymph nodes, thus playing a role in the staging and monitoring of the disease [12].

Pelvic unicentric CD is not easily diagnosed because of its low prevalence and non-specific imaging features; hence, most cases are usually confirmed by histopathologic examination after surgery or biopsy [4,13]. The differential diagnosis of a retroperitoneal mass includes lymphoma, sarcoma, metas-
tasis, neural tumor, desmoid tumor, and granulomatous disease [5,8,12]. Among these, lymphoma is the most difficult to distinguish from CD because of its similar homogeneity on radiologic findings [13]. CD may even resemble tubo-ovarian infections, an endometriotic cyst, or a dermoid ovary cyst [7,13]. Sato reviewed a retrospective study in the Japanese literature (1976–2011), and found that of the 105 cases of localized retroperitoneal CD, only 22 cases (21%) were located within the pelvic retroperitoneal region; only 8 (36.4%) of them were suspected before surgery [6]. However, unicentric CD is an important differential diagnosis of pelvic masses in women.

Unicentric CD responded well to resection, which is considered as the standard treatment. The removal of unicentric CD is feasible in the majority of cases, and it is almost always curative [4], even for cases arising from the pelvic lymph nodes; nearly 100% with 5 years survival [11]. However, recurrences have been reported, especially for incomplete resection [4]. The risks of surgical complications were severe haemorrhage following hypervascularity and marked fibrous adhesion to the surrounding tissues of the pelvic CD [6,13].

Conclusion

Unicentric CD is very rare, and it is characterised by non-systemic symptoms and nonspecific radiologic features; however, it is important to consider it as a differential diagnosis for hypervascular and heterogeneous lymphadenopathy favouring solitary benign tumor located within the female pelvis. Complete surgical resection can be curative for unicentric CD, and it can be used for histopathologic examination.

Patient consent

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

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