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Castleman's disease in the pelvic retroperitoneum: A case report and review of the Japanese literature

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ABSTRACT

INTRODUCTION: Castleman's disease is a fairly rare benign tumor of lymphoid origin. It can develop anywhere lymphoid tissue is found, but the expected origin is mediastinum and rarely pelvic retroperitoneum.

PRESENTATION OF CASE: A 22-year-old woman was admitted to our hospital for a mass in the pelvic retroperitoneum that was detected incidentally on an ultrasonography during a routine medical checkup with no signs of symptoms. After laboratory examination, ultrasonography, and magnetic resonance imaging (MRI), surgical resection was performed successfully through a lower midline incision. But the patient was needed transfusion because of massive bleeding. Postoperative histopathological diagnosis was hyaline-vascular type of Castleman's disease. The patient is leading an active social life without any signs of sequelae or recurrence.

DISCUSSION: Through the review of Japanese literature on Castleman's disease in the retroperitoneum, the characteristics of preoperative imaging findings are studied. Castleman's disease is easily misdiagnosed clinically because of its scarcity and no specific imaging findings. And the embolization via angiography should be considered in the hypervasular tumors such as in this case to prevent massive bleeding and transfusion.

CONCLUSION: Although Castleman's disease is uncommon, it should always be included in the differential diagnosis of pelvic retroperitoneal tumors. A better knowledge of this disease would help surgeon to avoid unnecessarily extensive resection and massive bleeding for transfusion when dealing with retroperitoneal tumors.

1. Introduction

Castleman's disease is a fairly rare benign tumor of lymphoid origin with unknown etiology.[1] It is also known as an angiofollicular lymph node hyperplasia or giant lymph node hyperplasia. It was first described in the mediastinum, although it may also be found in cervical, axillary, and other regions. Seven percent of cases may be found in the retroperitoneal space.[2] Here a case of Castleman's disease localized in the pelvic retroperitoneum is presented.

2. Presentation of case

A 22-year-old woman who had paraplegia due to congenital spina bifida was incidentally found to have a pelvic mass near the uterus on ultrasonography during the routine medical checkup. Complete blood count, erythrocyte sedimentation rate and routine blood biochemistry were within normal limits. The chest and abdominal X-ray films showed normal appearance. Ultrasonography revealed a large round soft tissue mass showing relatively homogeneous and hypoechogenic in front of the uterus (Fig. 1). MRI revealed that the mass was further delineated neighboring structures. It was homogeneous and nearly isointense to uterus in signal intensity on T1-weighted images (Fig. 2a), and heterogeneously hyperintense signal characteristics within the mass on T2-weighted images (Fig. 2b). It was heterogeneously hyperenhanced and increased intensity in the periphery of it on dynamic T1-weighted fat images (Fig. 3a), and heterogeneously enhanced on dynamic T2-weighted images (Fig. 3b). No abdominal lymphadenopathy or surrounding invasion was noted, and the lesion was found to be hormonally inactive after an endoclinological survey.

Although a definitive preoperative diagnosis could not be made, the operation was performed. By means of lower midline abdominal incision, an elliptical mass with diffuse and rich vascularity in the pelvic retroperitoneum was found that was adherent surrounding tissues. The tumor appeared to be mainly perfused by the branch of the median sacral artery. The tumor was completely resected after cut of these vessels, but she was needed blood transfusion because of massive hemorrhage due to oozing from the tumor capsule. The surface of the tumor was smooth,

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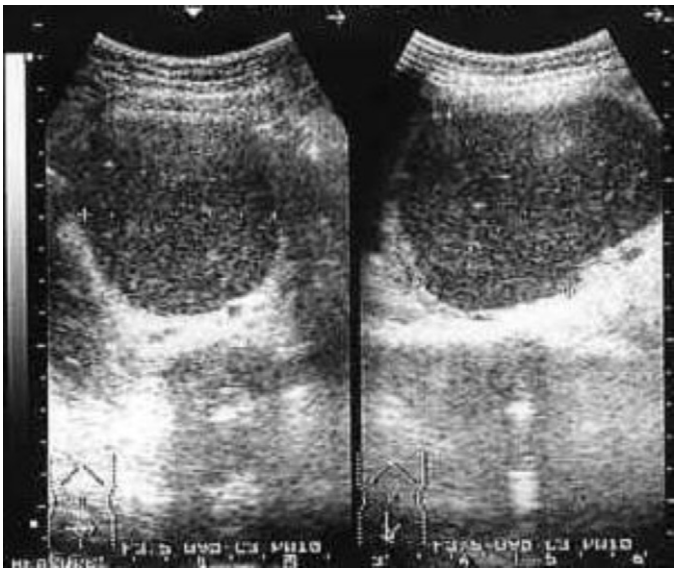

Fig. 1. US shows a large round shaped soft tissue mass showing relatively homogeneous and hypoechogenic.

well-delineated, and encapsulated. There were no findings of tumor invasion to the adjacent organs and structures. The tumor was 95 mm × 70 mm × 70 mm in size (Fig. 4a). The cut surface of the resected tumor showed a well-defined capsule and a homogeneous, grayish-yellow and fleshy texture with some scattered small bleeding spots (Fig. 4b).

Further pathological studies showed hyaline-vascular follicles with highly vascularized interfollicular areas and numerous mature

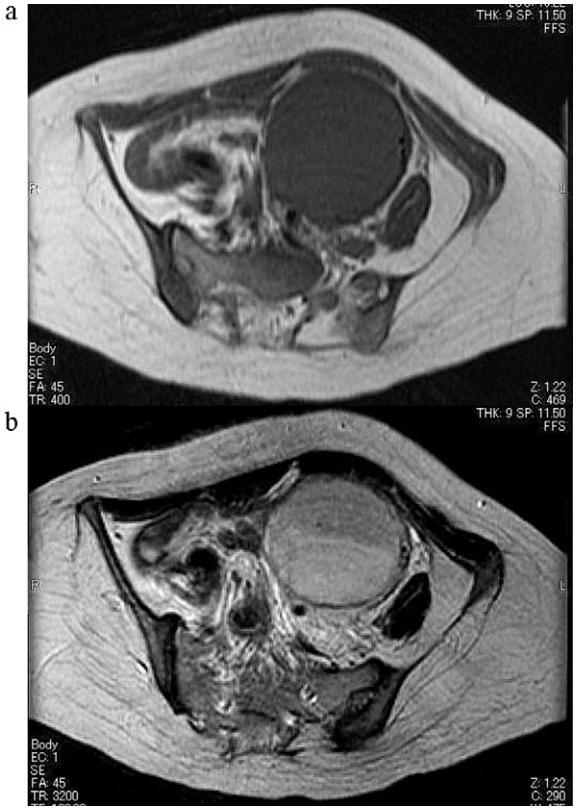

Fig. 2. (a) MRI on T1-weighted images shows a homogeneous and nearly isointense mass. (b) MRI on T2-weighted images shows a heterogeneous and hyperintense mass.

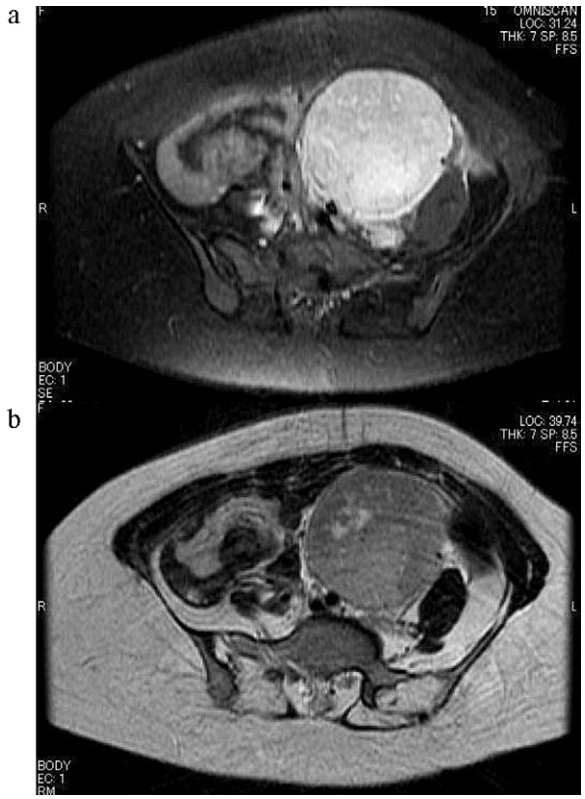

Fig. 3. (a) MRI on dynamic T1-weighted fat images shows a heterogeneously hyperenhanced mass. (b) MRI on dynamic T2-weighted images shows a heterogeneously enhanced mass.

lymphocytes. No malignant cells were seen. These histological findings were consistent with the hyaline-vascular type of Castleman's disease (Fig. 5). There were no postoperative complications, and the patient was discharged 8 days later. The patient was leading an active social life without any signs of sequelae or recurrence until the present for nine years.

3. Discussion

Castleman's disease was first reported in 1956 by Castleman et al. as a different mediastinal mass, easily confused with the thymoma.[1] Castleman's disease is located in the mediastinum in 70% of cases. While the superficial nodal groups constitute 20%, retroperitoneal localization that account for 7% of the cases, are very rare.[2,3] 105 cases of localized retroperitoneal Castleman's disease have been reported in Japanese literature since 1976–2011 out of which 49 cases (46.7%) were in peri-renal region, 17 cases (16.2%) were in peri-pancreatic region, and 22 cases (21.0%) were in pelvic retroperitoneal region. Furthermore, in these 105 cases, 63 were female cases, and 38 were male cases. Their average age was 43.4 ± 15.8 (standard deviation), and tumor size was 61.7 ± 25.0 (same as above) mm. It is easily misdiagnosed clinically because of its scarcity and no specific imaging findings. In these 105 cases, there were only 8 cases that suspected Castleman's disease preoperatively. Ultrasonography generally showed a focal hypoechoic mass with small hyperechoic regions. Computed tomographic findings were described in 69 cases of them, and generally showed a homogeneous or partially heterogeneous soft tissue density mass with early contrast enhancement, often accompanied by calcification typically called arborizing calcification. MRI findings were described in 44 cases of them, and generally showed an isointense mass to muscle on T1-weight images and diffuse or partially hyperintense mass on T2-weight images as is described in present case.

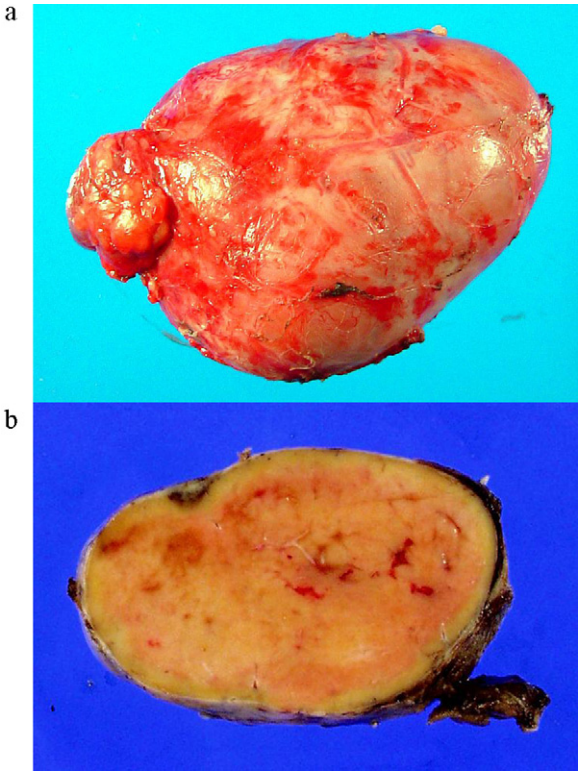

Fig. 4. (a) Surgical specimen. (b) Cut surface of the specimen.

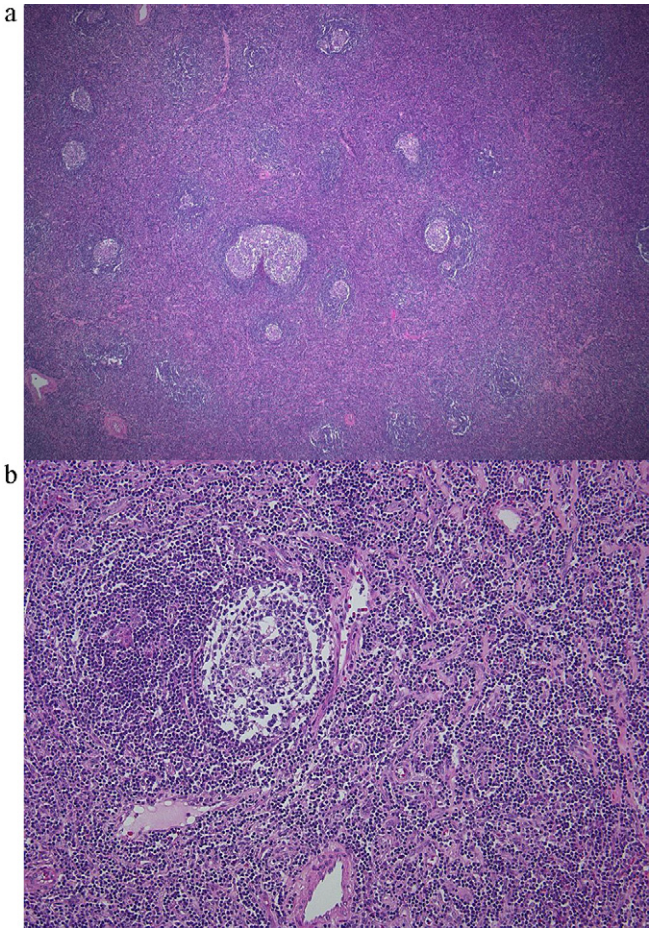

Fig. 5. Histopathological findings. (a) HE, 20× and (b) HE, 100×.

Two histopathological types of the disease have been described as hyaline-vascular and plasma cell types.[4] The hyaline-vascular type accounts for 90% of cases and the plasma cell type is found in the remaining one tenth. Recently, a mixed type of hyaline-vascular and plasma cell types has also been reported.[5] Both types are found at times in the same patient in separate places. These two types are thought to be associated with chronic low grade inflammatory process by infection with Human herpes virus 8.[6]

As is the case presented here, the hyaline-vascular type is usually solitary and asymptomatic except for possible compression of adjacent structures while plasma cell type frequently occurs to systemic signs and symptoms in about half of patients, and is the predominant form in multifocal disease.[4] The most common signs and symptoms are fever, anemia, night sweat, malaise, weight loss, elevated erythrocyte sedimentation rate, and polyclonal hypergammaglobulinemia. 19 (86.4%) of 22 pelvic retroperitoneal Castleman's disease in the Japanese literature were hyaline-vascular type, solitary, and asymptomatic. Another 3 cases were plasma cell type, solitary, and 2 of 3 cases had the clinical abnormalities such as fever, polyclonal hypergammaglobulinemia, high level of serum C reactive protein and interleukin 6, and so on.

Castleman's disease has been divided into two types as localized and multicentric types except the histopathological classification.[5] Localized type is predominantly of the hyaline-vascular type and solitary mass is detected. The treatment is surgical resection, which gives good results. On the other hand, multicentric type is predominant form of the plasma cell type, and has a poor prognosis despite treatment with corticosteroids and chemotherapy.[7]

Preoperative diagnosis of Castleman's disease is difficult because of its rare frequency and nonspecific radiographic sign. The imaging characteristics and the solitary nature without paraneoplastic activity in an otherwise healthy young patient favor a benign tumor. But benign retroperitoneal tumors are relatively uncommon, comprising only about 20% of all primary retroperitoneal neoplasms.[3] Actually, in 105 cases of retroperitoneal Castleman's tumors in Japan, only 8 cases were suspected them preoperatively. However, it is important to recall Castleman's disease as a differential diagnosis.

Although Castleman's tumors show benign histology, surgical excision is not always easy. The hypervascularity is frequently associated with massive hemorrhage at excision as in our case. And pelvic retroperitoneal Castleman's tumor is often accompanied by remarkable fibrous adhesion to the surrounding tissues.[8] Therefore, the embolization of the feeding artery of the tumor before surgery should be considered to prevent operative bleeding. Complete surgical resection is as yet the only treatment option and is all that is required for complete recovery in the majority of cases.

4. Conclusion

This is a rare case report of pelvic retroperitoneal Castleman's disease. Although it is easily misdiagnosed clinically because of its scarcity and no specific imaging findings, it is important to recall Castleman's disease as a differential diagnosis. The embolization of the feeding artery of the tumor before surgery should be considered to prevent operative massive bleeding because of its hypervascularity.

Conflict of interest statement

None.

Funding

None.

Ethical approval

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.

Author contributions

A. Sato – study design, data collections, data analysis, writing, final corrections.

The author approves paper for submission.

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