

The Stroma-Rich Variant Subtype of Castleman's Disease in the Retroperitoneum: A Case Report¹

Jung-Ho Kang, M.D., Myeong-Jin Kim, M.D., Young Chul Kim, M.D.,
Yong Eun Chung, M.D., Jin-Young Choi, M.D.

The stroma-rich variant of the hyaline-vascular type of Castleman's disease is a rare disease entity that is characterized by the overgrowth of a variety of stromal cells in the widened interfollicular area. We report a case of a 21-year-old woman who was admitted with a retroperitoneal mass that was initially suspected to be a liposarcoma, malignant fibrous histiocytoma or neurogenic tumor, but later confirmed as the stroma-rich variant subtype of Castleman's disease. Preoperative computed tomography (CT) and magnetic resonance imaging (MRI) showed an arborizing pattern of central calcifications and centrifugal contrast enhancement pattern of the mass. Herein, we discuss the MRI findings of this stroma-rich variant subtype.

Index words : Giant lymph node hyperplasia
Stroma-rich variant subtype
Magnetic resonance (MR)
Retroperitoneal space

Castleman's disease is a benign condition of unknown etiology characterized by angiofollicular lymph node hyperplasia (1). It is classified into two major histological subtypes: the hyaline-vascular and the plasma cell variants. The hyaline-vascular type is more common (90%) and has greater vascularity; it is characterized by abnormal lymphoid follicles, numerous vessels and wide fibrous septa, and is generally seen as a localized disease. Meanwhile, the plasma cell type, which accounts for less than 10% of all cases of Castleman's disease, is characterized by polyclonal mature plasma cell proliferation and fewer vessels, and is almost always associated with

systemic manifestations. The disease is encountered in the mediastinum in the vast majority of cases, but it may occur anywhere along the lymphatic chains such as in the neck, axilla, mesentery, thorax, pancreas, spleen, adrenal glands and retroperitoneum (2). We report a rare stroma-rich variant of Castleman's disease of the hyaline-vascular type that was encountered in the retroperitoneum. To the best of our knowledge, the radiologic findings of this type of Castleman's disease have not been previously described.

Case Report

A 21-year-old woman with a medical history of Graves' disease was admitted because of a three-year period of weight loss and a recently palpated abdominal mass. The physical examination revealed a large palpable mobile mass in the left lower quadrant of the abdomen. The results of routine laboratory tests were nor-

¹Department of Diagnostic Radiology, Research Institute of Radiological Science, Yonsei University Health System

Received November 11, 2008 ; Accepted July 9, 2009

Address reprint requests to : Jung-Ho Kang, M.D., Department of Diagnostic Radiology, Research Institute of Radiological Science, Yonsei University Health System, 250 Seongsanno, Seodaemun-gu, Seoul 120-752, Korea.

Tel. 82-2-2228-7400 Fax 82-2-393-3035 E-mail : jhkang@yuhs.ac

mal.

Ultrasonography (US) revealed a large solid hypoechoic mass with a central vascular core and the presence of vascularity within the mass was noted on color Doppler (Figs. 1A, B). Computed tomography (CT) demonstrated an approximately 11.5 cm, well-defined, lobulated, soft-tissue, retroperitoneal mass with an arborizing pattern of central calcifications (Fig. 2A). On the portal phase, the mass showed strong enhancement with a distinct radial or fissured non-enhanced area in the center and no peripheral enhancement (Fig. 2B). Percutaneous biopsy of the mass was avoided because of the abundance of vascularity demonstrated on US and CT, and further investigation was done via magnet-

ic resonance imaging (MRI).

MRI also showed a well-defined lobulated mass abutting the psoas muscle in the left infrarenal paraaortic region. The mass showed strong enhancement with gradual centrifugal and peripheral 'rim-like' enhancement on the delayed phase. There was visible vessel proliferation around the tumor, and any invasion of the adjacent organs or vessels was not depicted (Fig. 3).

The patient underwent resection of the well-encapsulated retroperitoneal mass. The gross specimen exhibited solid, homogeneous and yellowish nature, which was consistent with the diagnosis of Castleman's disease of the hyaline-vascular type (Fig. 4). In addition, there was strong alpha-SMA immunoexpression in the inter-

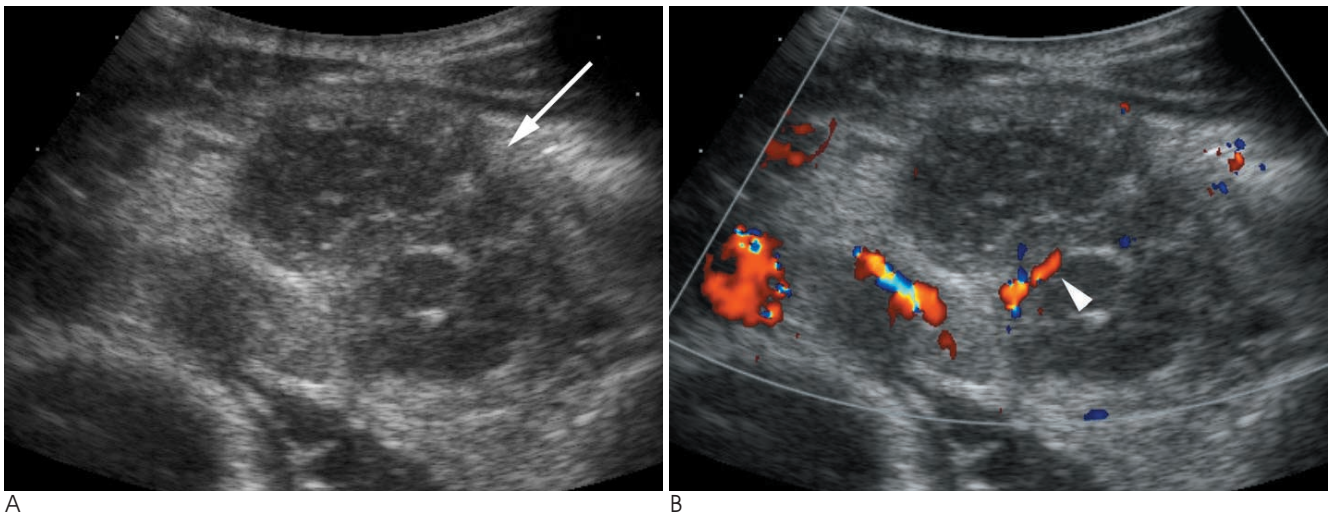


Fig. 1. US images of the retroperitoneal mass.

A. Gray-scale US reveals a large solid hypoechoic mass (arrow) in the left lower quadrant of the abdomen.

B. Color Doppler US shows a central vascular core (arrow head) and the presence of vascularity within the mass.

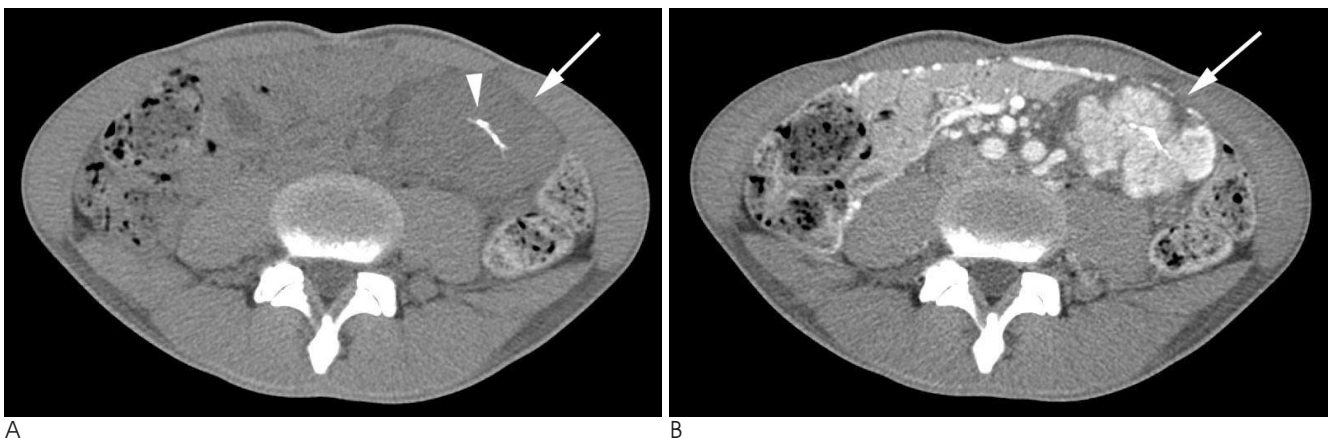


Fig. 2. CT images of the retroperitoneal mass.

A. The pre-contrast axial CT image shows an arborizing pattern of central calcifications (arrow head) within the homogeneous mass (arrow).

B. The contrast-enhanced axial CT image shows a well-defined enhancing retroperitoneal mass (arrow) with a central fissured non-enhanced area and the absence of rim enhancement.

follicular spindle cells without expression of CD21, CD23, CD68 and cytokeratin. These immunohistological results confirmed the diagnosis as stroma-rich variant of Castleman's disease of the hyaline-vascular type.

Discussion

The stroma-rich variant of Castleman's disease of the

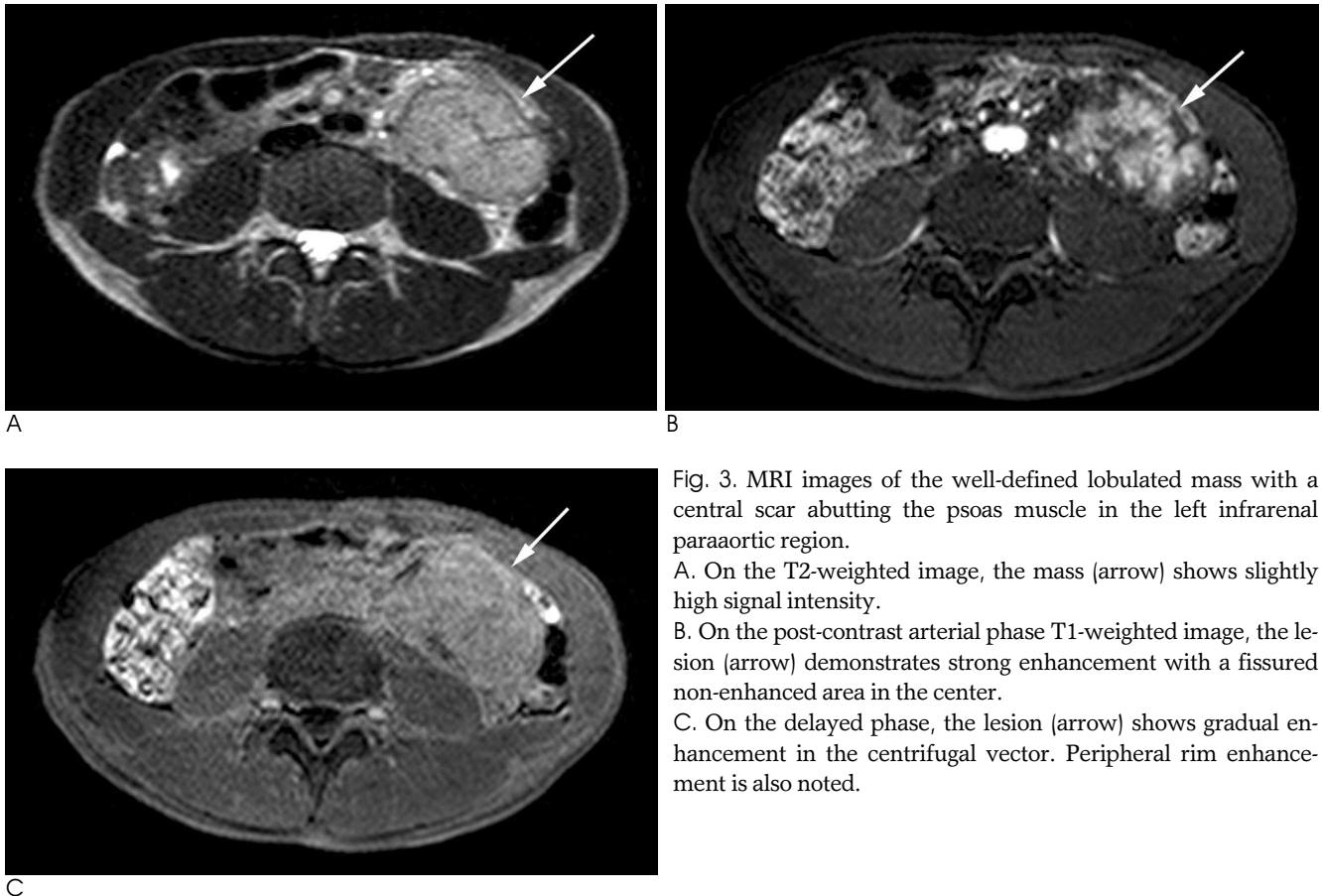


Fig. 3. MRI images of the well-defined lobulated mass with a central scar abutting the psoas muscle in the left infrarenal paraaortic region.

A. On the T2-weighted image, the mass (arrow) shows slightly high signal intensity.

B. On the post-contrast arterial phase T1-weighted image, the lesion (arrow) demonstrates strong enhancement with a fissured non-enhanced area in the center.

C. On the delayed phase, the lesion (arrow) shows gradual enhancement in the centrifugal vector. Peripheral rim enhancement is also noted.

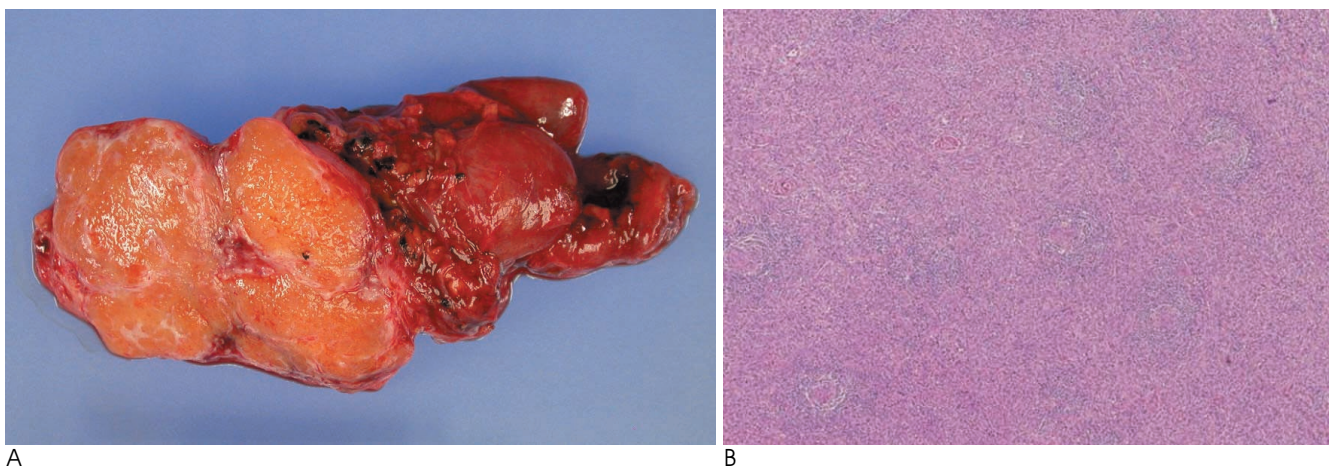


Fig. 4. The gross specimen and microscopic features of the tumor.

A. The photograph of a cut section of the resected specimen demonstrates a well-encapsulated retroperitoneal mass that is characterized by its solid, homogeneous and yellowish features.

B. The follicle shows an abnormal germinal center with hyalinized capillaries surrounded by concentrically arranged lymphocytes, which resembles the appearance of an onion skin. The widening of the interfollicular area appears to create an ill-defined nodule. Inside it, there is prominent proliferation of stromal cells (H-E stain, ×100).

hyaline-vascular type is a recently identified entity that shows overgrowth of a variety of stromal cells. In 1993, Danon et al. reported a histological review and analysis of 102 cases of Castleman's disease of the hyaline-vascular type; these cases were divided into three subtypes according to the differences in the proportion of follicles to interfollicular tissue. Cases with near equal proportion of follicles and interfollicular tissue were classified into the classic variant, those with the follicular component exceeding 50% were classified into the follicular variant, and those with the interfollicular tissue component exceeding 50% were classified into the stroma-rich variant. The stroma-rich variant, in contrast to the other subtypes, occurred exclusively in the adult population, frequently involved the intra-abdominal lymph nodes rather than the peripheral lymph nodes or mediastinum, and tended to form a large mass. Moreover, this histological variant demonstrated vessel proliferation in the interfollicular areas along with excessive proliferation of either SMA positive spindle cells or CD68 positive histiocytic reticulum cells (3).

The localized hyaline-vascular Castleman's disease of the abdomen and pelvis most commonly manifests on CT as a single, well-defined, homogeneous soft-tissue attenuation mass with moderate to marked enhancement, with the presence of calcifications in approximately 30% of the cases (4). When the mass exceeds 5 cm in diameter, the interior of the tumor can exhibit distinct radial or fissured non-enhanced areas during the early stage of enhancement. The non-enhancing areas are reduced or vanish entirely on a delayed CT scan (5). Zheng et al. (6), in a report on the newly discovered features of localized Castleman's disease using multi-detector helical CT, recently described a peripheral 'rim-like' enhancement of the mass at the arterial phase and/or the portal phase on contrast-enhanced CT as another characteristic feature.

In our case, the mass demonstrated strong enhancement with a distinct radial or fissured non-enhanced area in the central portion. However, the mass failed to show peripheral rim enhancement on CT at either the arterial or portal phases. Instead, centrifugal enhancement of the mass was shown on MRI with gradually progressive enhancement from the center towards the periphery, and the 'rim-like' enhancement was seen on the delayed-phase MRI.

To the best of our knowledge, there are currently no reports on the MRI findings of this specific stroma-rich variant. The peculiar progressive centrifugal enhance-

ment of this tumor is noteworthy and it may be a representative feature of this subtype. Although more studies are needed to clarify the cause of this enhancement pattern, it is probably the outcome of gradual centrifugal diffusion of the contrast medium from the central vascular core throughout the abundant and thick interfollicular stromal component comprising the mass.

The differential diagnosis of Castleman's disease that presents as a retroperitoneal mass includes lymphoma, sarcoma, fibrohistiocytoma, neurogenic tumor and granulomatous disease, all of which can share similar imaging characteristics.

Lymphoma, another common retroperitoneal malignancy, is very difficult to distinguish from Castleman's disease because of its homogeneity (7). However, the hyaline-vascular type of Castleman's disease tends to exhibit intense enhancement on CT and angiography. Leiomyosarcoma presents as a heterogeneous mass that contains cystic and solid components. Liposarcoma will often, but not always, have an identifiable amount of fat. Malignant fibrohistiocytoma has a complex appearance and it can contain amorphous calcifications (8).

Castleman's disease should be considered in the differential diagnosis of a well-enhancing mass that is found in an area of normally distributed lymphoid tissue (9). An arborizing pattern of central calcifications may help suggest the diagnosis (10).

In conclusion, the characteristic features of the stroma-rich variant of Castleman's disease localized in the retroperitoneum are hypervascularity and a centrifugal enhancement pattern on imaging studies. A general understanding of the imaging findings of this variant may be helpful to narrow down the preoperative differential diagnosis.

References

1. Demirpolat G, Pourbagher A, Hekimgil M, Elmas N, Kitis O, Korkut M, et al. Mesenteric Castleman's disease: case report. *Abdom Imaging* 2000;25:551-553
2. Libson E, Fields S, Strauss S, Bloom RA, Okon E, Galun E, et al. Widespread Castleman disease: CT and US findings. *Radiology* 1988;166:753-755
3. Danon AD, Krishnan J, Frizzera G. Morpho-immunophenotypic diversity of Castleman's disease, hyaline-vascular type: with emphasis on a stroma-rich variant and a new pathogenetic hypothesis. *Virchows Arch A Pathol Anat Histopathol* 1993;423:369-382
4. Kim TJ, Han JK, Kim YH, Kim TK, Choi BI. Castleman disease of the abdomen: imaging spectrum and clinicopathologic correlations. *J Comput Assist Tomogr* 2001;25:207-214
5. Zhou LP, Zhang B, Peng WJ, Yang WT, Guan YB, Zhou KR. Imaging findings of Castleman disease of the abdomen and pelvis.

- Abdom Imaging* 2008;33:482-488
6. Zheng X, Pan K, Cheng J, Dong L, Yang K, Wu E. Localized Castleman disease in retroperitoneum: newly discovered features by multi-detector helical CT. *Abdom Imaging* 2008;33:489-492
7. Moon WK, Kim WS, Kim IO, Yeon KM, Han MC. Castleman disease in the child: CT and ultrasound findings. *Pediatr Radiol* 1994;24:182-184
8. Singletary LA, Karcnik TJ, Abujudeh H. Hyaline vascular-type Castleman disease: a rare cause of a hypervascular retroperitoneal mass. *Abdom Imaging* 2000;25:207-209
9. Yamashita Y, Hirai T, Matsukawa T, Ogata I, Takahashi M. Radiological presentations of Castleman's disease. *Comput Med Imaging Graph* 1993;17:107-117
10. Luburich P, Nicolau C, Ayuso MC, Torra R, Clavero JA. Pelvic Castleman disease: CT and MR appearance. *J Comput Assist Tomogr* 1992;16:657-659

대한영상의학회지 2009; 61 : 329-333

후복막에서 발생한 초자-혈관형 Castleman병의 간질 풍부 변종: 증례 보고¹

¹연세의대 세브란스병원 영상의학과

강정호 · 김명진 · 김영철 · 정용은 · 최진영

초자-혈관형 Castleman병의 간질 풍부 변종은 드문 질병으로 확장된 여포 사이 공간에 과증식하는 간질 세포를 갖는 것이 특징이다. 우리는 후복막의 종괴를 주소로 내원한 21세 여자 환자의 증례를 보고하고자 한다. 내원 당시 이 종괴는 지방육종, 악성섬유조직구종 또는 신경성종양으로 의심되었으나 수술 후 조직 병리상 초자-혈관형 Castleman병의 간질 풍부 변종으로 최종 확인되었다. 수술 전 시행한 전산화단층촬영술 및 자기공명영상에서 가지를 치는 모양의 중심성 석회화와 원심성 조영증강이 확인되었다. 본 증례에서는 간질 풍부 변종의 자기공명영상소견에 대해서 고찰하고자 한다.