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Castleman's syndrome: Main sonographic findings

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ABSTRACT
Castleman's disease is a rare, usually benign, lymph node disorder that can manifest as cervical or mediastinal adenopathy. There are three histological variants: unicentric hyaline-vascular, unicentric plasma cells, and multicentric plasma cells, each with different symptoms and radiological features. The hyaline vascular form has a predilection for the thorax, and can simulate other conditions such as thymoma or lymphoma. Imaging tests, such as CT scans and MRIs, are useful for diagnosis, revealing characteristic patterns of lymph node enhancement and involvement. Treatment varies depending on the variant of the disease and may include surgery, steroids, chemotherapy, or antiviral therapy. Accurate diagnosis is challenging, requiring attention from radiologists to differentiate Castleman's disease from other lymphoproliferative conditions. This report aims to describe the clinical case of a patient admitted to the emergency department with pain in the left hypochondrium and to warn about the need for early diagnosis and follow-up for appropriate therapy.

Keywords: Castleman's disease, Castleman's tumor, Lymphoproliferative disorders, Lymphadenopathy, Radiology, Diagnostic imaging, Doppler ultrasonography of vessels.

1 INTRODUCTION

Castleman's disease (CD) is a rare disorder, also known as angiofollicular lymph node hyperplasia and giant lymph node hyperplasia, with a generally benign clinical course [1] . It is the benign lymphoproliferative disease that most frequently involves the mediastinal lymph nodes and may present in the head and neck as a cervical adenopathy of unknown etiology.

It appears to be related to multiple causes, including Human Herpes Virus-8 (HHV-8) and paraneoplastic pemphigus 2,3. CD was first described in 1956 by CASTLEMAN et al, as a localized mediastinal lymph node hyperplasia mimicking a thymoma.

According to the histological type, CD can be classified into three variants: unicentric hyaline-vascular (VHU), which corresponds to 72%; unicentric plasma cells (CPU), 18%; and multicentric plasma cells (MPC); 10%. Clinical manifestations vary according to the predominant histological type. The VHU form is usually asymptomatic, and is most often discovered in routine examinations; it usually occurs in young, healthy people, with 70% of cases occurring before the age of 30, with no predominance of sex. In the CPU and MPS forms, most patients present constitutional symptoms, such as fever, weight loss, elevated erythrocyte sedimentation rate (ESV) and anemia, and in the MPS variant, there seems to be a certain preference for males.4

The objective of this report is to describe the clinical case of a patient admitted to the emergency department with pain in the left hypochondriasis and to warn about the need for early diagnosis and follow-up for appropriate therapy.

2 DESCRIPTION

A 60-year-old male patient was admitted to the service due to a report of sudden pain in the left hypochondrium for 2 days. Severe pain with a characteristic stinging condition, unrelated to position or factors of improvement or worsening. Undergoing treatment for COPD infected for 4 days with Moxifloxacin. She denied cough, dyspnea, runny nose or other respiratory symptoms. He did not use any painkillers.

She was in consultation with an assistant hematologist who indicated hospitalization for propaedeutics. Total abdominal Doppler ultrasonography of hepatic vessels was performed, followed by multislice computed tomography of the thorax and total abdomen, assuming Castleman's Disease or Pneumonia as a diagnostic hypothesis. Among the differential diagnoses is Splenic Thrombosis. On physical examination, the patient presented with pain fascies, pale, hydrated at the threshold, anicteric, and acyanotic. HR 101 bpm FR 18irpm PA 90x60mmHg.

Image 1 - Total abdominal Doppler ultrasonography of hepatic vessels: nonspecific hepatosplenomegaly, simple hepatic cyst, simple renal cyst on the left.

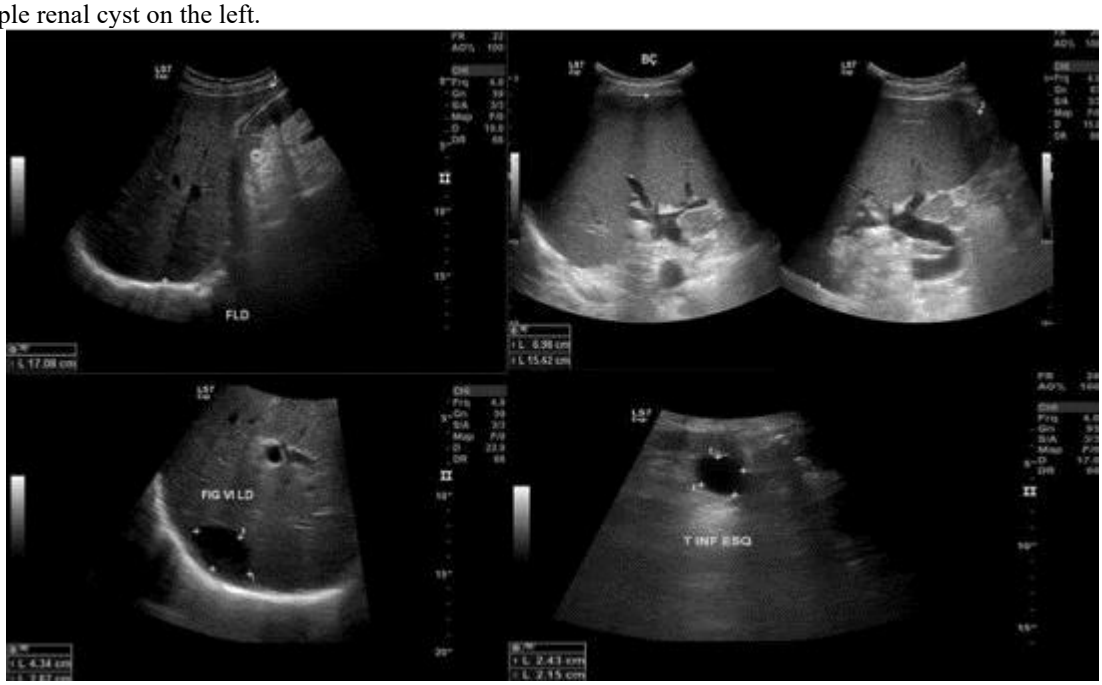


Image 2 - Total abdominal Doppler ultrasonography of hepatic vessels: Mesenteric and peripancreatic lymph node enlargement near the hepatic and splenic hila, and paraaortic lymph node enlargement.

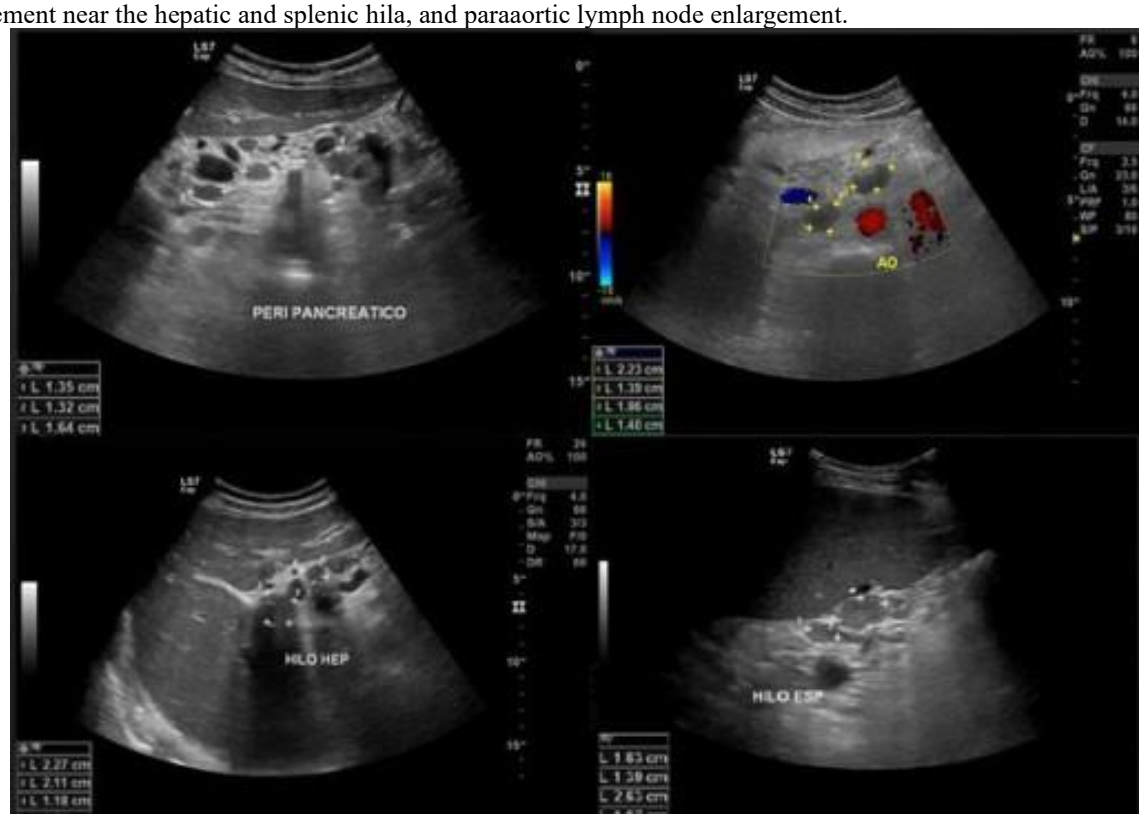


Image 3: Increase in the caliber of the splenic vein, possibly compensatory to the decrease in the caliber of the superior mesenteric vein. Doppler study showing preserved splenic flow.

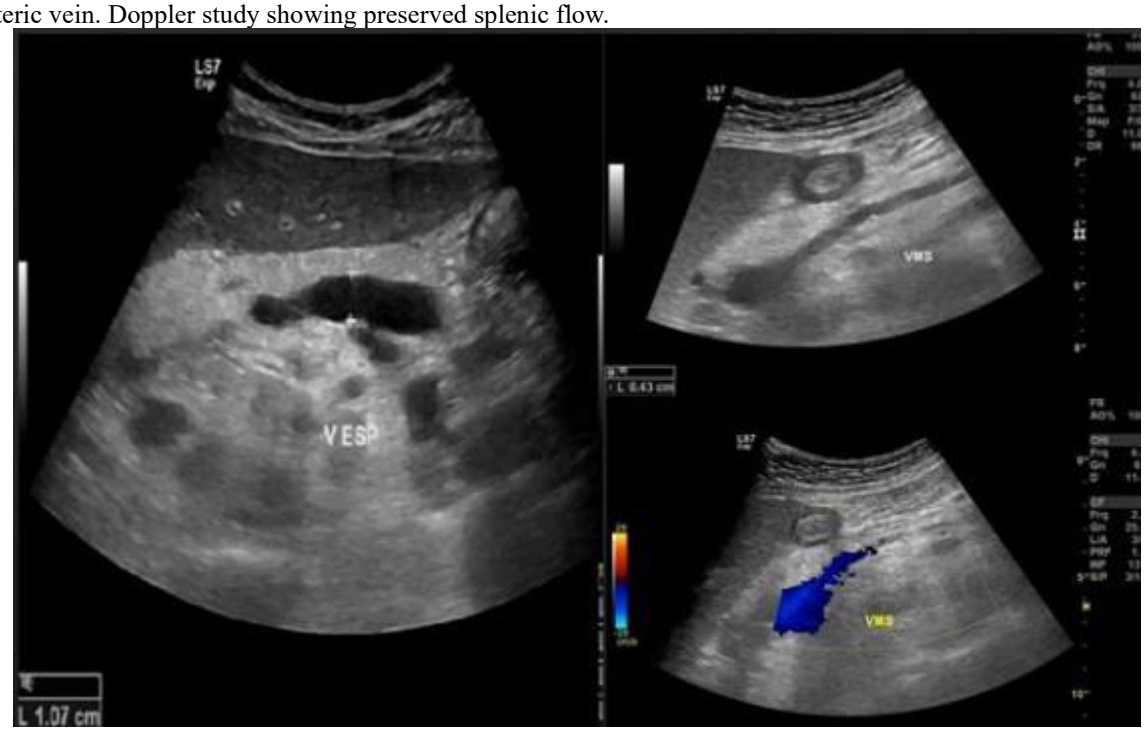


Image 4: Doppler study showing preserved splenic flow. Absence of thrombosis of the splenic and portal veins.

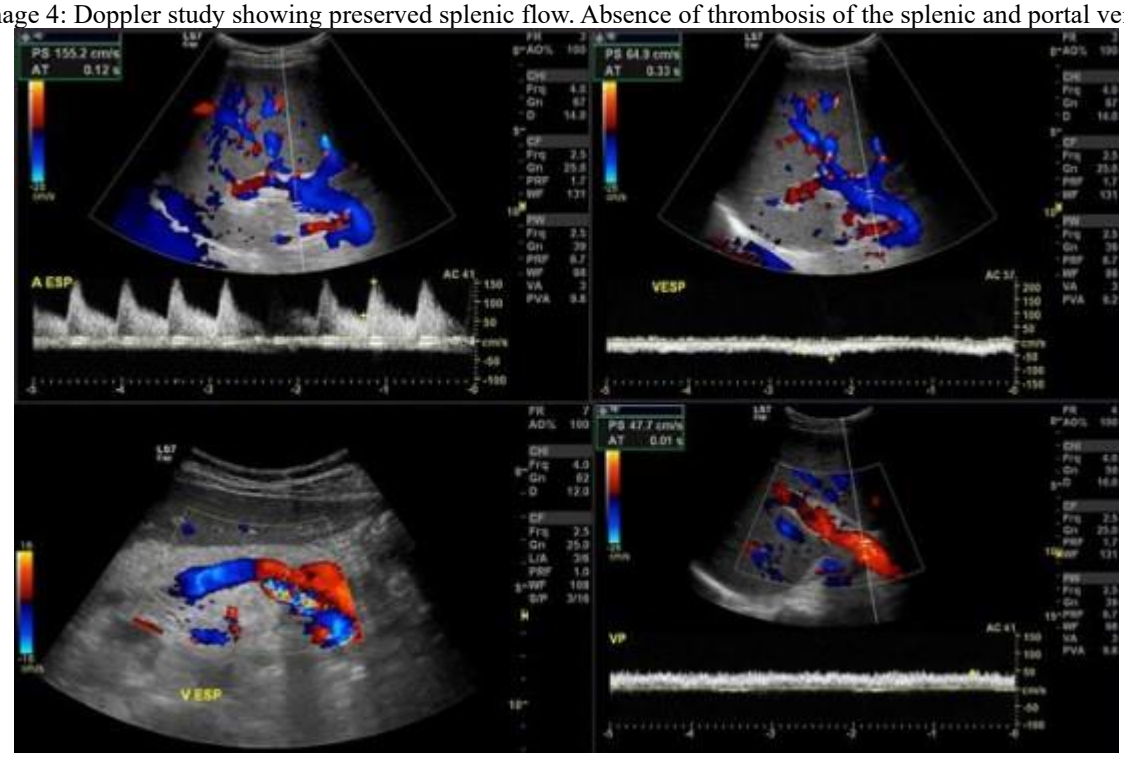


Image 5 – Multislice computed tomography of the chest: Mediastinal and axillary lymph node enlargement. Bilateral, right-sided and small left laminar pleural effusion with compressive atelectasis of the adjacent lung parenchyma in the left lower lobe. Simple hepatic cyst in the right lobe.

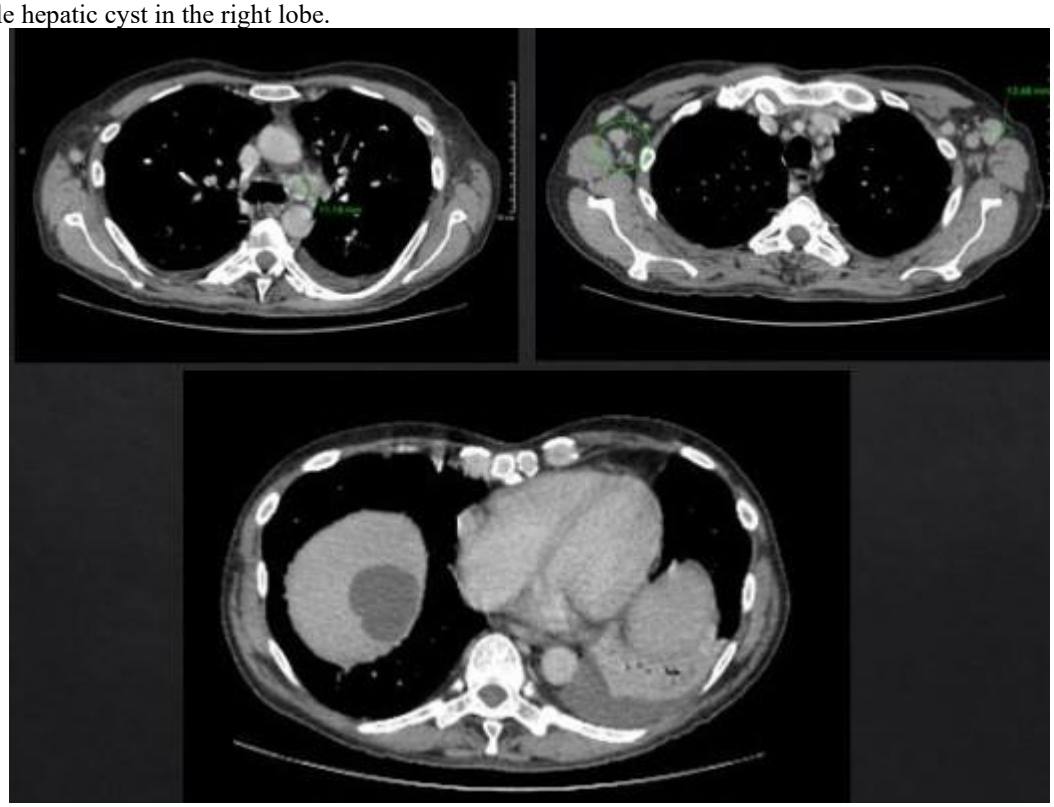
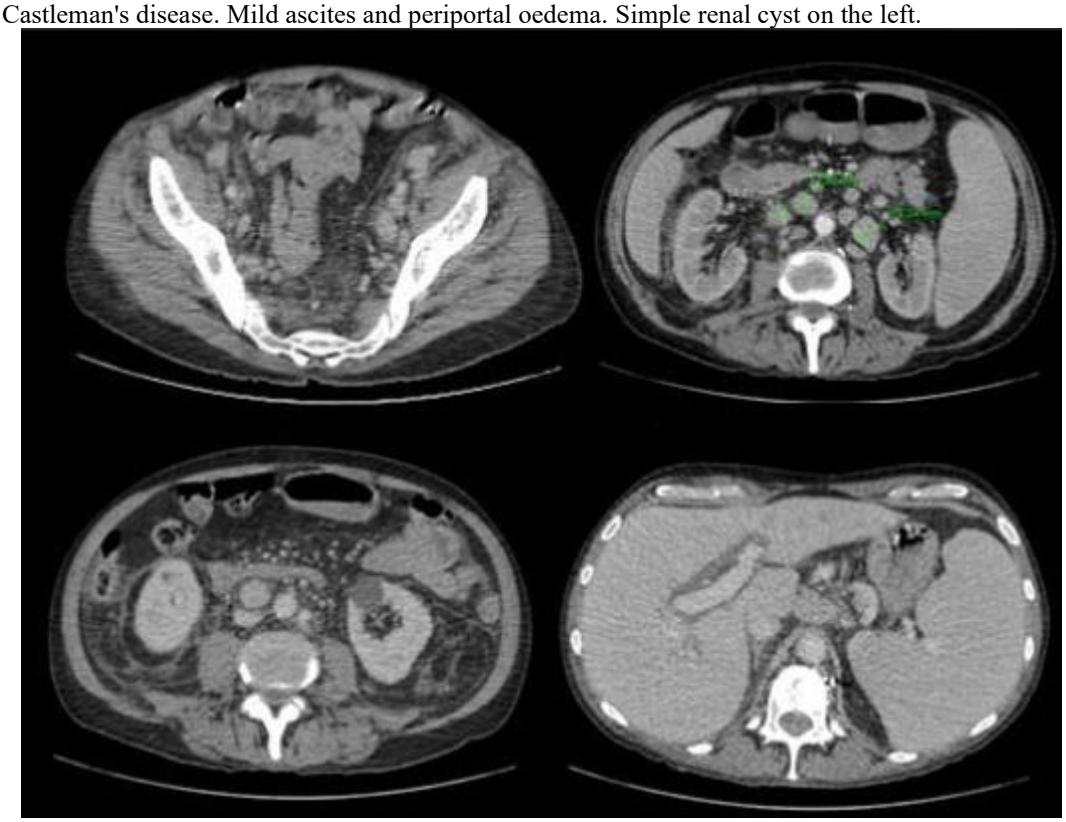


Image 6: Hepatosplenomegaly associated with diffuse abdominal and pelvic lymph node enlargement, possibly related to suspected Castleman's disease. Mild ascites and periportal oedema. Simple renal cyst on the left.



3 DISCUSSION

Hyaline vascular Castleman's disease has a considerable predilection for chest involvement, where it usually manifests as a long-enlarged mediastinal nodal mass. Mediastinal Castleman disease may mimic thymoma, lymphoma, sarcoma, hemangiopericytoma, neural crest-derived neoplasms such as paraganglioma, neurofibroma, or schwannoma, and chest wall tumors. 5

Among the radiological features, the classic CT appearance of hyaline vascular Castleman's disease is that of a single enlarged lymph node or localized nodal masses that demonstrate intense homogeneous enhancement after contrast administration. Three patterns of involvement have been described, including a non-invasive solitary mass (most common: 50% of cases), a dominant infiltrative mass with associated lymphadenopathy (40% of cases), and tangled lymphadenopathy without a dominant mass (10% of cases). 6

Hyaline Castleman's vascular disease may manifest as a mesenteric or retroperitoneal mass with mild contrast enhancement, with imaging mimicking retroperitoneal adenopathy and carcinoid tumor. Hyaline vascular Castleman disease has a considerable predilection for chest involvement, where it typically manifests as a eagerly enlarging mediastinal nodal mass. Mediastinal Castleman's disease can mimic thymoma, lymphoma, sarcoma, hemangiopericytoma, neural crest-derived neoplasms such as paraganglioma, neurofibroma, or schwannoma, and chest wall tumors. Hilar Castleman's disease can be confused with bronchial adenoma.5 Pleural Castleman's disease is uncommon and may manifest as a well-defined mass or with associated pleural effusion. Castleman's pericardial disease can mimic a pericardial cyst.

Castleman's intercostal disease may show erosion of the ribs and mimic other chest wall masses. Prominent feeding vessels in the vicinity of a nodal mass are a clue to diagnosis and are predominantly seen in hyaline vascular Castleman disease.

Approximately 10% of lesions have internal calcifications, which are characteristically coarse or have a distinct branching pattern. More commonly, however, nonspecific calcifications are observed. Central hypoattenuation in nodal masses is uncommon but may be seen in a few cases.

On magnetic resonance imaging (MRI), hyaline vascular Castleman disease lesions classically exhibit heterogeneous T1 and T2 hyperintensity compared to skeletal muscle. 6 Prominent flow voids can be seen, which identify the feeding vessels. Magnetic resonance imaging is well suited to describe the extent of the disease and the relationship to adjacent structures, although evaluation of calcifications is limited.

Systemic manifestations of hyaline vascular Castleman disease are rare compared to the other forms and include pleural and pericardial effusions, hepatomegaly, and diffuse lymphadenopathy. Such an aggressive course can mimic that of an aggressive lymphoma. As discussed in the following section, associated diseases are less commonly seen in hyaline vascular Castleman disease than in the other

forms, but they do occur with hyaline vascular Castleman disease. The combination of adenopathy and lytic or sclerotic bone lesions may be a clue to the diagnosis of hyaline vascular Castleman disease with associated POEMS syndrome. Hyaline vascular Castleman's disease can occur in unusual sites, such as the presacral region, mimicking a nerve sheath tumor, inflammatory pseudotumor, desmoid tumor, or lymphoma. Plasma cell Castleman disease typically demonstrates less avid enhancement after administration of contrast material compared to hyaline vascular Castleman disease, which makes differentiation of reactive or neoplastic nodal involvement more difficult.

Castleman's disease of unicentric plasma cells is not uncommon and manifests as a focal mass. Near the pancreas, unicentric plasma cell Castleman's disease can mimic pancreatic lymphoma, adenocarcinoma, or neuroendocrine tumor. 5

Castleman's disease includes a broad spectrum of pathological findings, manifestations, and associations. It most commonly manifests as a single-center disease with a hyperenhancement lymph node mass and should be considered in the differential diagnosis of lymphoma, metastatic adenopathy, and infectious and/or inflammatory causes of adenopathy. Due to the diverse manifestations of Castleman's disease and its ability to affect any region of the body, Castleman's disease is a great mimicry of benign and malignant findings in the neck, chest, abdomen, and pelvis. Castleman disease includes a spectrum of pathological variants, including classic hyaline vascular Castleman disease and the less common plasma cell Castleman disease, multicentric Castleman disease, and HHV-8-associated Castleman disease.

Castleman's disease may be associated with HIV/AIDS, lymphoma, POEMS syndrome, paraneoplastic pemphigus, and plasma cell dyscrasias. HHV-8-associated Castleman's disease occurs predominantly in patients with immunosuppression and/or HIV infection and commonly has an aggressive and fatal course with a poor prognosis. Aggressive forms with systemic manifestations may occur in plasmacytic Castleman disease and are rare in unicentric hyaline vascular Castleman disease. Unicentric hyaline vascular Castleman disease is often curable with surgery; treatment of multicentric Castleman disease may require steroid therapy, chemotherapy, antiviral medication, or the use of antiproliferative regimens. 5,6 Although rare, Castleman's disease should always be included in our differential diagnoses of lymph node enlargement in daily medical practice. It is worth mentioning that the clinical and anatomopathological diagnosis are often difficult and inconclusive in relation to Castleman's disease, for this reason the radiologist must be attentive and use different imaging methods to elucidate the diagnosis and possible atypical complications to favor the appropriate therapeutic conduct.

REFERENCES

Yeh CM, Chou CM, Wong LC. Castleman's disease mimicking intrapulmonary malignancy. Ann Thorac Surg. 2007;84(2):e6-7.

Hengge UR. Update on Kaposi. Sarcoma and other HHV8 associated diseases.

Part 2: pathogenesis, Castleman Disease, and pleural effusion lymphoma. Lancet Infect Dis, 2(5): 281-292, 2002.

Twist CJ. Asessment of lymphadenopathy in children. Pediatr Clin North Am 49(5):1009-1025,2002.

Caprio JS, Leitão LS, Simão, MNS. Castleman's disease: case report. Rev. Soc. Bras. Clín. Méd ; 14(3): 163-165, jul. 2016.

Bonekamp D, Horton KM, Hruban RH, Fishman EK. Castleman Disease: The Great Mimic. RadioGraphics 2011; 31:1793–1807

Castleman Disease of the Thorax: Clinical, Radiologic, and Pathologic Correlation: From the Radiologic Pathology Archives RadioGraphics 2016; 36:1309-1332.

Zhag S, Wan Y, Huang Z. Imaging and clinical features of Castleman Disease. Cancer Imaging 19, article number 53,2019.

Mishra A, ElHousieni EA, Ehtuish EF. Retropharyngeal Castleman´s disease: US, CT and MR imaging findings: Case report and review of literature.

Meadow T, McLarney J. Features of Castleman Disease of the Abdomen and Pelvis. AJR Online.