Case Report

Retroperitoneal Unicentric Castleman Disease: Surgery is Curative

Doença de Castleman Unicêntrica Retroperitoneal: A Cirurgia é Curativa

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ABSTRACT

Castleman disease (CD) comprises a group of rare nonclonal lymphoproliferative disorders.

Unicentric Castleman disease (UCD) typically presents as isolated lymphadenopathy with progressive enlargement and an indolent course. Most patients are asymptomatic or may present symptoms related to a localized mass effect on organ function. Multicentric Castleman disease (MCD) affects various lymph node stations and leads to systemic constitutional symptoms and systemic cytokine dysregulation, resulting in laboratory abnormalities, hepatosplenomegaly, and complex organ dysfunction.

We report a case of retroperitoneal UCD. Imaging studies showed a solitary, solid, right pararenal lesion causing anterior deviation of the inferior vena cava and renal pedicle. Complete surgical resection was performed, and histopathologic analyses confirmed the diagnosis of the hyaline-vascular subtype of UCD.

Patients with UCD generally have an excellent prognosis following curative surgical resection. However, recurrence may occur, and long-term follow-up is advised.

Keywords: Castleman Disease/diagnostic imaging; Castleman Disease/surgery

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RESUMO

A doença de Castleman é um grupo de doenças raras linfoproliferativas não-clonais.
A doença de Castleman unicêntrica (DCU) manifesta-se tipicamente como uma linfadenopatia isolada com crescimento progressivo e indolente. A maioria dos doentes são assintomáticos ou apresentam sintomas compressivos dos órgãos adjacentes. A doença de Castleman multicêntrica afecta várias regiões ganglionares e cursa com sintomas sistémicos constitucionais, desregulação sistémica das citoquinas, alterações laboratoriais, hepatoesplenomegalia e disfunção de órgão complexa.
Apresentamos um caso clínico de DCU retroperitoneal. Os estudos imagiológicos revelaram uma lesão sólida para-renal direita que causa desvio anterior da veia cava inferior e do pedículo renal. O exame histopatológico da peça operatória confirmou DCU do subtipo hialino-vascular.
O prognóstico após ressecção cirúrgica completa é favorável. A possibilidade de recidiva tardia preconiza um *follow-up* prolongado.

Palavras-chave: Doença de Castleman/cirurgia; Doença de Castleman/diagnóstico por imagem

INTRODUCTION

Castleman disease (CD) comprises a group of rare nonclonal lymphoproliferative disorders. Benjamin Castleman first described the unicentric form in 1954 as a localized hyperplasia of lymphoid tissue.[1] In the 1980s, a variant of multicentric disease with multiple lymph node station involvement was described. CD Classification divides the diseases into two forms, Unicentric and Multicentric, based on morphologic features. MCD is further divided according to histopathogenetic classification into peripheral sensorimotor neuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin lesions (POEMS)-associated MCD (POEMS-MCD), HHV8+ MCD, and idiopathic MCD (iMCD).[2,3] Histopathologically, there are three main patterns described in Castleman disease: hyaline-vascular (90%-91% of cases), plasma cell, and mixed type.[4]

UCD can be diagnosed at any age, but typically occurs in the third or fourth decade of life and has an equal gender distribution. There are no recognized epidemiological risk factors. UCD is characterized by the involvement of a single lymph node region, 90% of the time localized in the chest, neck, abdomen, or retroperitoneum. Most cases of UCD are hyaline-vascular type histology, and usually, these patients are asymptomatic or may present symptoms related to localized mass effect on organ function. Approximately 10% to 25% of patients have plasma cell type histology that can be associated with constitutional symptoms (e.g., fever, sweat, weight loss) and laboratory abnormalities (e.g., anemia, elevated erythrocyte sedimentation rate and/or C-reactive protein, hypergammaglobulinemia, and hypoalbuminemia) reflecting excess interleukin-6 secretion.[5,6]

Surgical resection provides simultaneous histological diagnosis and potential for curative treatment. Complete surgical resection is the preferred treatment of UCD. For unresectable lesions with compression symptoms, debulking surgery or local radiotherapy are alternative therapies. Given the generally indolent nature of the disease, a watch-and-wait strategy can be considered in asymptomatic patients with low-burden disease. Still, paraneoplastic syndromes, such as bronchiolitis obliterans and pemphigoid eruptions, may develop rarely.[7] In symptomatic patients who are not eligible for surgery or local radiotherapy, systemic therapeutic options for MCD, such as chemotherapy, rituximab, or anti-IL-6 therapy, can constitute an alternative strategy.[5]

MCD affects various lymph node stations associated with marked systemic constitutional symptoms and systemic cytokine dysregulation, resulting in laboratory abnormalities, hepatosplenomegaly, and complex organ dysfunction. In the majority of patients, the cause is idiopathic. In HHV8+ MCD, the primary risk factor is immunocompromised status, often associated with HIV infection.[4,6] MCD more commonly develops in the sixth decade of life and tends to have an unfavorable prognosis. MCD therapeutic options are multiple, and treatment is complex. Unlike UCD, surgery in MCD does not have the potential to be curative. Instead, surgery is typically reserved for tissue harvesting for diagnostic purposes or to alleviate compressive symptoms through debulking surgery. Multiple therapeutic strategies have been used, including chemotherapy, antiviral agents, glucocorticoids, thalidomide, interferon-alpha, and molecular target therapies.[8,9]

CD harbors two distinct entities, unicentric and multicentric disease, with very different clinical characteristics, therapeutic strategies, and prognoses.[9]

We report a case of retroperitoneal unicentric Castleman disease in a 70-year-old woman who presented with abdominal pain.

CASE REPORT

A female patient in her 70s presented to the Emergency Department with abdominal pain for the past four days, localized in the right lower quadrant. She denied fever, anorexia, nausea, vomiting, or any other symptomatology. Past medical history revealed well-controlled hypertension. Blood and urine analyses were within normal limits. Physical examination was unremarkable. Abdominal and pelvic ultrasound identified an echogenic expansive process next to the right kidney, approximately 5-6 cm in diameter, with irregular contours and a hypoechoic central area.

Abdominal and pelvic computed tomography (CT) scans show a 4.6 cm fatty mass with a solid component, anterior to the right kidney, with renal pedicle compression (Fig. 1A). This mass causes anterior deviation of the inferior vena cava; there is no invasion of contiguous structures, particularly the kidney (Fig. 1B).

Abdominal magnetic resonance imaging (MRI) described a solid nodule that appears to be in the right adrenal gland dependence; it measures 35 x 34 x 22 mm, is not delimited by a capsule, and presents diffusion restriction and contrast enhancement.

18F-fluorodeoxyglucose positron emission tomography-computed tomography ([^{18}F]FDG PET/CT) confirmed the existence of a right pararenal lesion with elevated SUV (SUV=5), suggesting the diagnosis of a neoplastic retroperitoneal lesion (Fig. 2), and excluded disease in other locations.

Based on imaging features, the main differential diagnoses included liposarcoma (primary retroperitoneal lesion) and right adrenal myelolipoma.

In short, this is a resectable retroperitoneal solid mass in a fit patient (ECOG-PS 0) with low surgical risk (ACS NSQIP Surgical Risk Calculator).

After multidisciplinary discussion, surgical excision was recommended for histopathological diagnosis and tailored treatment. A right-sided lateral transcostal approach was used to access the retroperitoneal space, providing optimal exposure of the lesion, right kidney, renal hilum, and inferior vena cava. The lesion caused compression of the inferior vena cava and right renal hilum, but careful dissection enabled complete en bloc removal without rupture (Fig. 3A).

The postoperative course was uneventful, and the patient was discharged on the third day after surgery.

Gross examination of the surgical specimen revealed a well-defined, pink, elastic nodular mass measuring 3.5 × 2.5 × 2 cm within an 11 × 8.5 × 3.5 cm resection specimen (Fig. 3B).

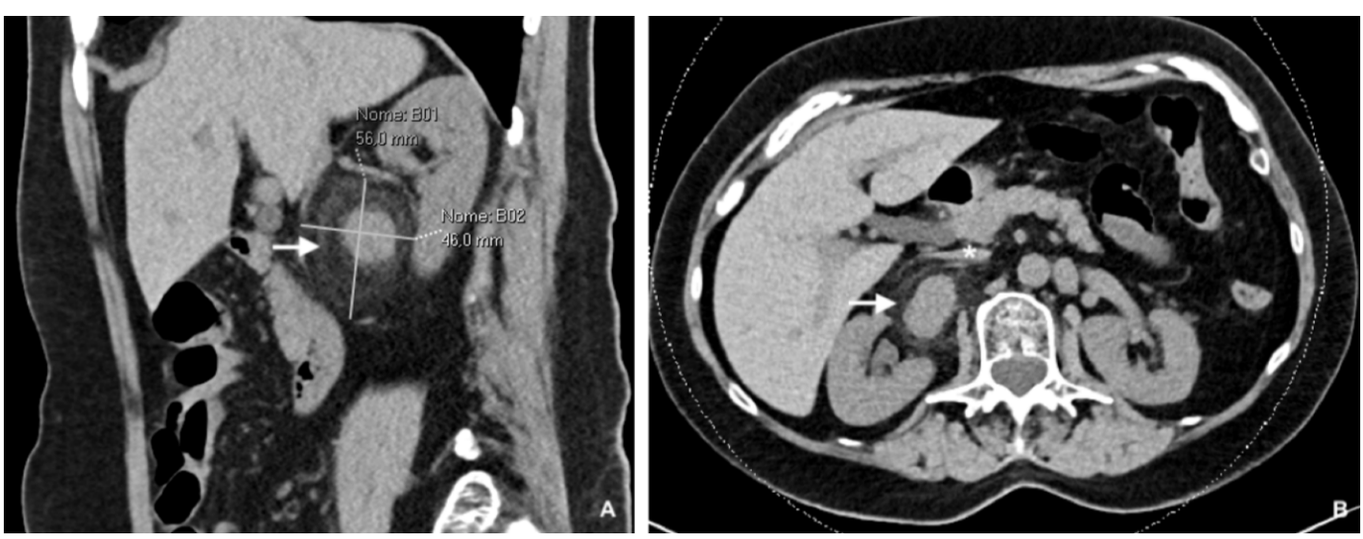

Figure 1 – Abdominal and pelvic computed tomographic (CT) scans. (A) The sagittal CT scan section shows a fatty mass (56x46 mm) with a solid component (arrow) anterior to the right kidney. (B) The axial CT scan section shows the lesion (arrow) causing anterior deviation of the inferior vena cava (*).

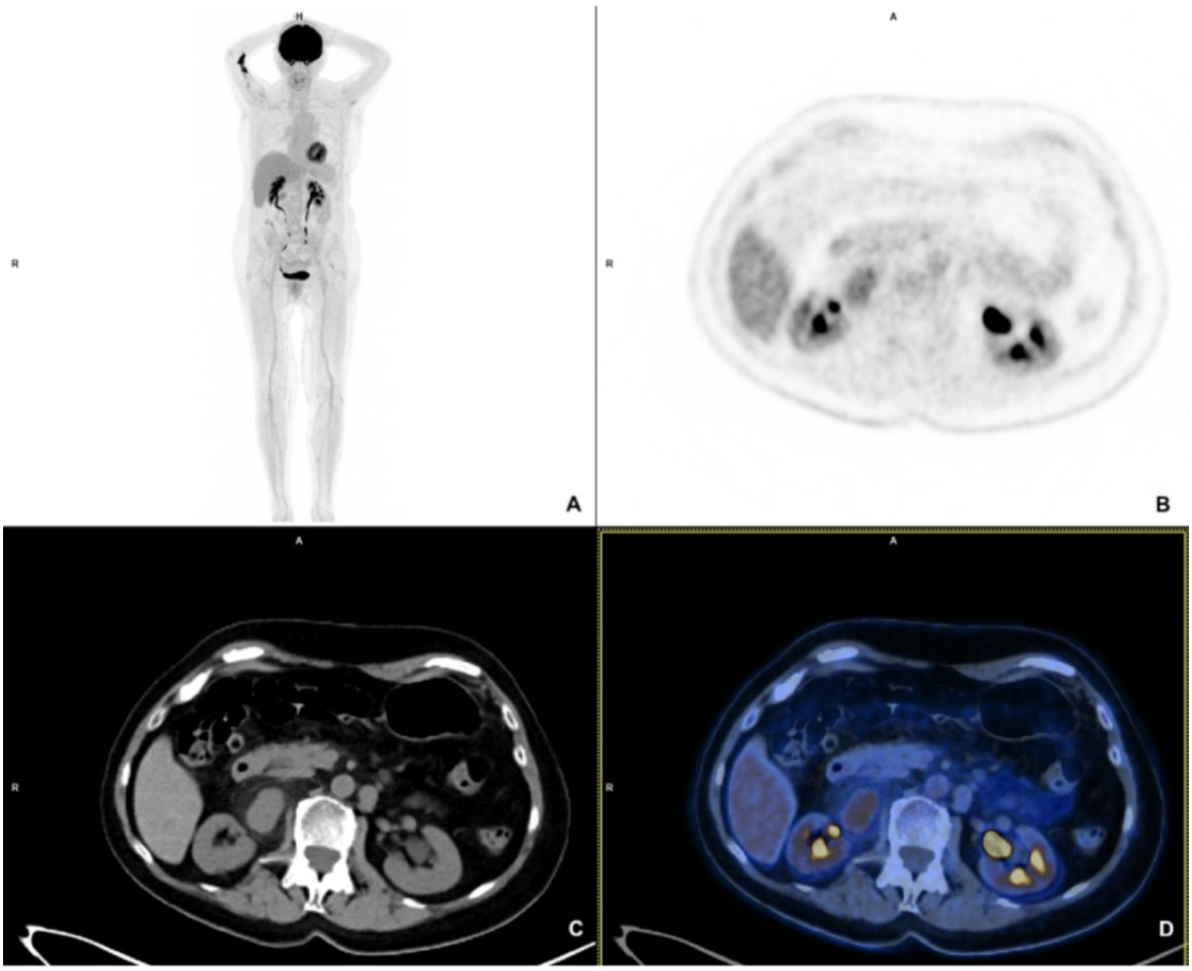

Figure 2 – [^{18}F]FDG PET/CT. (A) Whole body evaluation shows a unique lesion located near the right renal hilum. (B) PET image shows [^{18}F]FDG uptake in the lesion. (C) CT image shows the anatomical location and structural features of the lesion. (D) PET/CT fusion image.

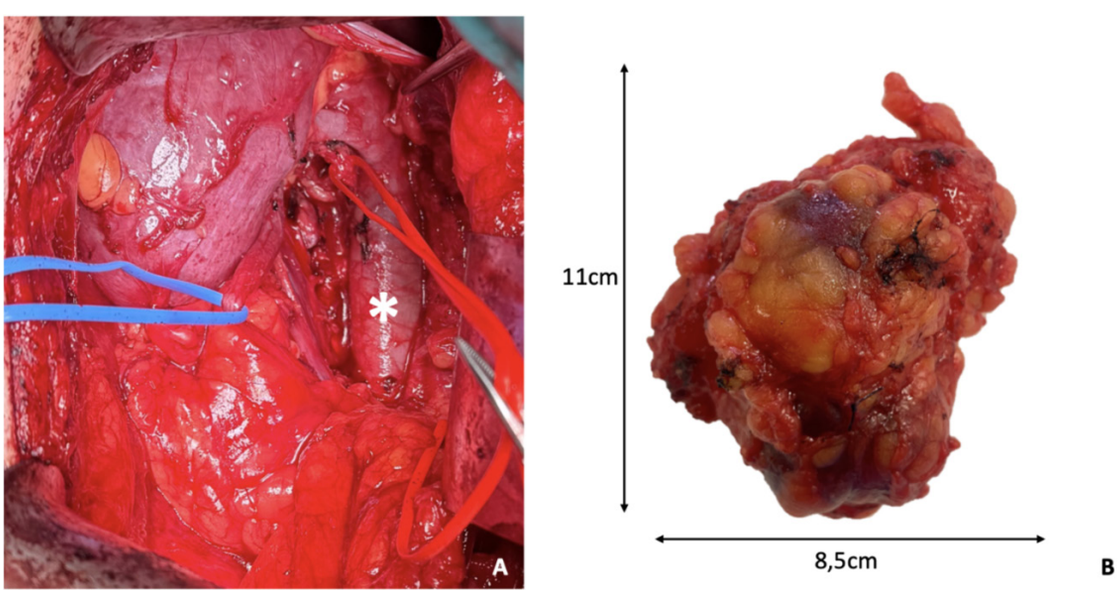

Figure 3 – (A) Surgical site after specimen removal. Right ureter (blue vascular silicone tie). Renal artery branch (red vascular silicone tie). Inferior vena cava (*). (B) Macroscopic photograph of the surgical specimen.

Microscopic analysis showed a lymph node with altered architecture, characterized by numerous atretic follicles and an expanded mantle zone (Fig. 4A). Hyalinized blood vessels penetrating atrophic germinal centers were observed (Fig. 4B), along with slight proliferation of follicular dendritic cells (CD21+ and CD23+) (Fig. 4C). Immunostains for IgD and Bcl2 highlighted the mantle zone (Fig. 4D). Germinal centers contained few CD10- and Bcl6-positive B lymphocytes, while the interfollicular area showed a predominance of small CD3-positive T lymphocytes and marked vascular proliferation without endothelial atypia. HHV8 immunostaining was negative. CD138 highlighted sparse plasma cells without kappa or lambda light chain predominance.

These findings were consistent with unicentric Castleman disease, hyaline-vascular subtype.

After one year of follow-up, the patient remains asymptomatic and disease-free, with no abnormal [^{18}F]FDG PET/CT uptake.

DISCUSSION

Histologic examination is essential for diagnosing Castleman disease. Complete surgical removal is possible in most cases of UCD and serves both as a diagnostic and curative procedure. Fine-needle aspiration (FNA) biopsy was not considered, given the resectability of the lesion and the risk of tumoral seeding. Moreover, in previously described cases, fine-needle aspiration was not helpful, mainly because the histological diagnosis of CD is based on cell architecture.[10]

In a systematic review that included 278 patients with UCD, resective surgery with no further multimodal approach is considered a safe therapeutic option and should be regarded as the gold standard for treating suspected Castleman disease.[9] The authors of a retrospective study that included 71 Patients with biopsy-proven UCD enrolled in the French National Registry for CD over 20 years corroborate these conclusions.[11]

Patients with UCD have a good prognosis if a curative complete resection can be achieved, with a 5-year disease-free survival (DFS) rate higher than 80% and an overall survival (OS) rate higher than 90%. Although rare, recurrence has been reported up to 14 years after resection, making it imperative to conduct long-term follow-up. Surgical excision and/or radiotherapy can be considered in the presence of a

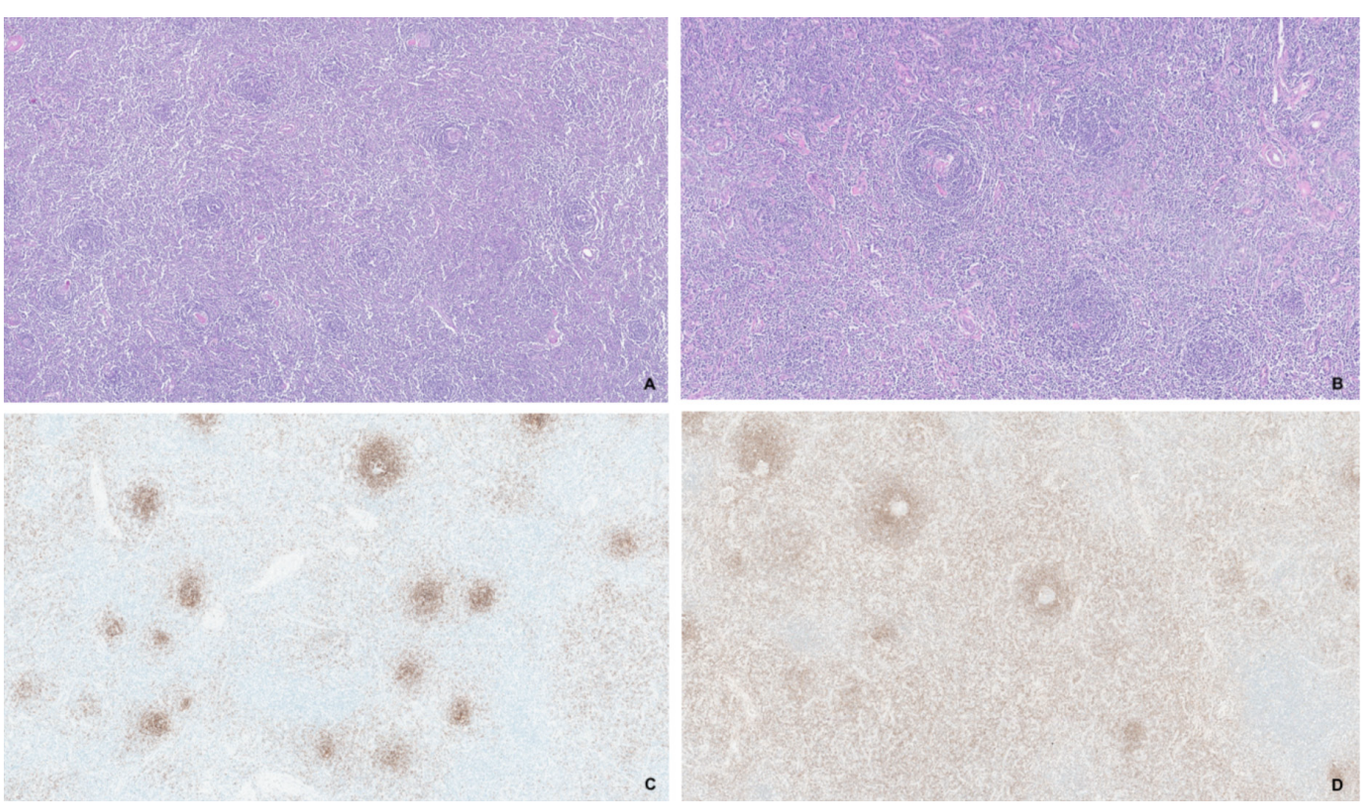

Figure 4 – Histopathological presentation of the patient's lesion (A) Atrophic germinal centers with expanded mantle layer (H&E; x40). (B) Hyalinized blood vessels in the center of the atrophic follicle (H&E; x100). (C) Positive CD23 showing atrophic follicular centers (x40). (D) Immunostains for IgD in mantle cells (x100).

recurrence. Incomplete resection was associated with a worse OS than complete resection but still approximately 70% at ten years.[4,7]

There are medical conditions associated with UCD that must be carefully monitored, such as osteosclerotic plasma-cell myeloma and POEMS syndrome in patients with the plasma-cell type and follicular dendritic-cell sarcoma and paraneoplastic pemphigus among patients with the hyaline-vascular type.[4] Plasma-cell UCD has a worse prognosis than the hyaline-vascular type.[5]

To date, a follow-up protocol for UCD patients has not been defined. In case of complete resection with negative margins, some centers recommend routine CT yearly, within the first three years, and alternating abdominal ultrasound with CT until year 5.[4] Additionally, [^{18}F]FDG PET/CT can detect abnormal uptake in non-enlarged lymph nodes and is more sensitive than contrast-enhanced CT in evaluating and monitoring CD.[5]

In summary, unicentric Castleman disease arising in the retroperitoneum, although rare, should be included in the differential diagnosis of solid retroperitoneal lesions. Complete surgical excision remains both diagnostic and curative.

LEARNING POINTS/TAKE-HOME MESSAGES

- The term Castleman Disease comprises distinct lympho-proliferative disorders with different clinical characteristics, therapeutic strategies, and prognoses
- Unicentric Castleman disease (UCD) is a rare, slow-growing lymphoproliferative condition involving a single anatomic site
- Most UCD cases are of the hyaline-vascular subtype and remain asymptomatic
- Complete surgical excision, when achievable, is the gold standard treatment providing both diagnosis and cure
- UCD carries an excellent prognosis, but long-term follow-up is recommended
- When facing an isolated retroperitoneal mass, UCD should be considered in the differential diagnosis

ETHICAL DISCLOSURES

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CONTRIBUTORSHIP STATEMENT

SBM: Design of the study, data collection and analysis, writing of the manuscript, critical revision of the article, linguistic revision and final formatting of the article.
FCA: Design of the study, participation in the imaging analysis, and critical revision of the article.
PT: Design of the study, critical revision of the article.
JSL: Design of the study, data collection pot and analysis, critical revision of the article.

DECLARAÇÃO DE CONTRIBUIÇÃO

SBM: Desenho do estudo, recolha e análise de dados, redação do manuscrito, revisão crítica do artigo, revisão linguística e formatação final do artigo.
FCA: Desenho do estudo, participação na análise de imagens e revisão crítica do artigo.
PT: Desenho do estudo, revisão crítica do artigo.
JSL: Desenho do estudo, recolha e análise de dados, revisão crítica do artigo.

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