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MISCELLANEOUS

CASE REPORT

Castleman's disease: atypical pathology – a case report

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Abstract

Castleman's disease is a rare, benign pathology characterized by lymph node hyperplasia. Clinically, it is classified as either multicentric or unicentric disease. The choice of treatment depends on the subtype of the disease, with surgical intervention being preferred for unicentric cases and a combination of chemotherapy, corticosteroids, or immunomodulators being used for multicentric cases. Given the rarity of this pathology, the objective of this study is to present a case report of a male patient with an anatomopathological diagnosis of Castleman's disease.

Keywords: Castleman's disease; cervical mass; head and neck surgery; case report.

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Introduction

Castleman's disease is an atypical, non-oncological condition that presents in two clinical forms[1].

The unicentric variant affects one or more lymph nodes in a single region of the body, and the majority of cases are asymptomatic[1]; in contrast, the multicentric variant involves lymphadenopathies across multiple regions of the body. Patients with this form of disease typically exhibit systemic inflammatory symptoms, generalized pathological lymph nodes, hepatosplenomegaly, cytopenia, and in some cases, organ dysfunction due to an excessive proinflammatory response to interleukin 6 (IL-6)[1,2].

Case report

A 47-year-old male patient with no history of prior diseases sought medical evaluation for a slow-growing mass in the submaxillary region, with no associated symptoms. Upon physical examination, a mass, mobile, non-painful left submaxillary mass, approximately 5 cm in diameter, was identified, along with a second mass at the right submaxillary level (Figure 1). A biopsy of the left submaxillary mass reported a lymph node with partial loss of architecture, primary and secondary lymphoid follicles with hyalinized germinal centers, and blood vessels with thickened walls. An expanded mantle area, observed in concentric layers in several follicles, and a slightly expanded interfollicular area due to the presence of reactive plasma cells were also noted. An immunohistochemical

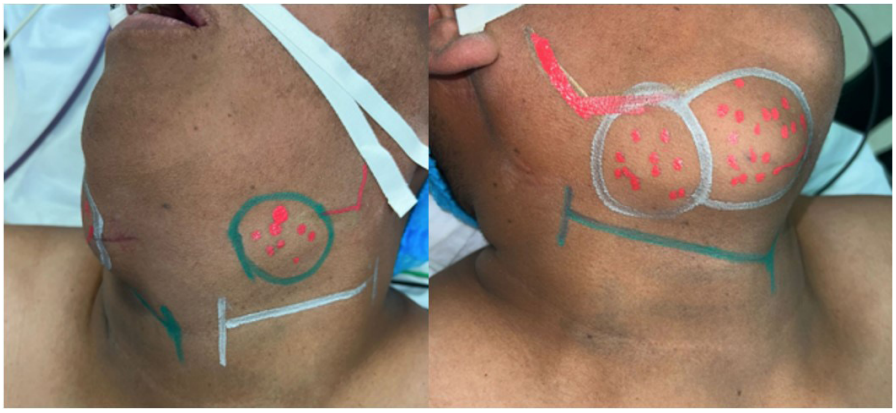

Figure 1. 47-year-old man with presence of cervical masses at the submental, right submaxillary, and left submaxillary levels.

study revealed CD3-positive in membrane of reactive T lymphocyte membranes, CD20-positive reactive B lymphocyte membranes, BCL-2 positivity in a reactive pattern, CD138-positive reactive plasma cell membranes in the zone-enhancing area, and CD23-positive expanded dendritic cells arranged in a concentric pattern. The biopsy results were consistent with mixed Castleman's disease (vascular hyaline and rich in plasma cells).

Further examinations—brain, thoracic, abdominal, and pelvic tomography—were carried out to assess the disease's extent, revealing no pathological changes. Neck tomography, however, revealed an adenomegaly approximately 4 cm in diameter at the IA level, a lymphadenopathy approximately 3 cm in diameter at the right IB level, and another approximately 3 cm at the left IB level. (Figure 2).

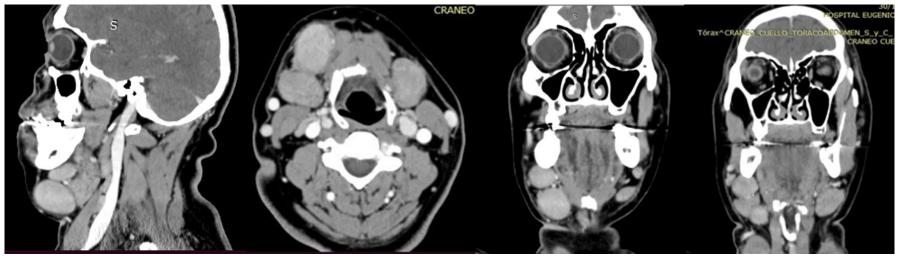

Figure 2. Tomography shows a homogeneous mass of 4 cm in diameter located at the cervical IA level, a homogeneous mass of approximately 3 cm located at the cervical right IB level, and a homogeneous mass of 3 cm at the left IB level.

Laboratory tests showed no signs of anemia, infectious or active inflammatory process. Given the diagnosis of localized Castleman's disease, a decision was made to perform surgical resection. This involved the complete resection of cervical adenopathies at the IA level and the bilateral IB level. During the surgery, a submandibular cervical adenopathy approximately 3x2x2 cm in size was found, along with a right submandibular cervical adenopathy approximately 3x2x2 cm in size, which was firmly attached to the submandibular gland, and a left submandibular adenopathy approximately 4x3 cm in size (Figure 3).

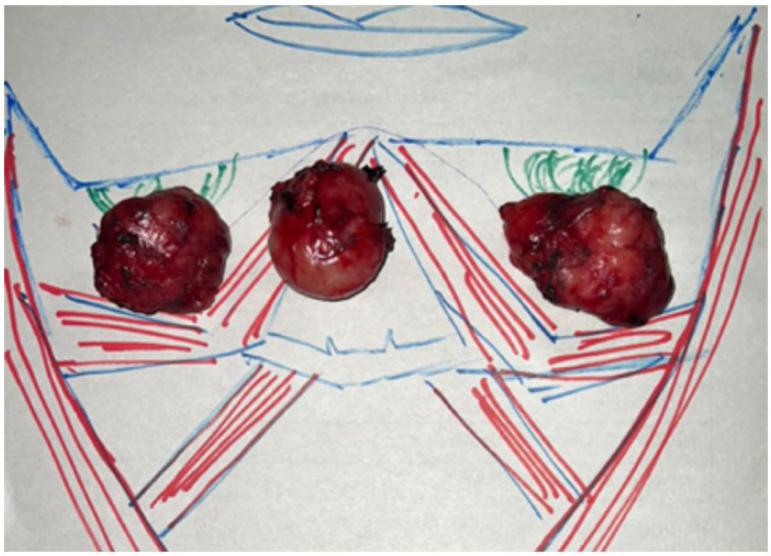

Figure 3. Complete resection of cervical adenopathies at the bilateral IB level and IA level.

The patient's post-surgical recovery was favorable, with adequate pain control and vital signs within normal limits. The patient tolerated the diet well and was discharged 24 h post-surgery. At one-year postoperative checkup, the patient was asymptomatic with no evidence of disease recurrence.

Discussion

Castleman's disease is a rare benign lymphoproliferative pathology, first described in 1956 by Benjamin Castleman, and is characterized by antigenic overstimulation of unknown origin. The clinical presentation can be either localized unicentric or multicentric disease[2,3].

The etiology remains unclear, but the pathophysiology of the disease often involves the proliferation of B lymphocytes and plasma cells in the lymphoid organs of any body region. Most often, it is identified in the mediastinum (30-70%), neck (20%), abdomen (23%), and retroperitoneum (14%)[4].

In some cases, multicentric disease is related to the human herpes virus 8 (HHV-8) and the human immunodeficiency virus (HIV), both of which can cause abnormal production of interleukin 6 (IL-6)[1]. The symptomatology in multicentric cases is associated with systemic disease resulting from an increase in IL-6, leading to B lymphocyte proliferation and vasculogenesis. This can manifest as constitutional symptoms such as diaphoresis, fever (>38 °C), weight loss, fatigue, hepatosplenomegaly, fluid retention with pleural effusion, peripheral edema and ascites, cutaneous findings like violet papules and angiomas, or respiratory pathologies such as lymphocytic interstitial pneumonitis[2,3]. The age of onset varies, but cases have been more frequently reported in age groups in the fifth decade of life without gender predilection. Imaging studies may reveal homogeneous diffuse lymphadenopathies with lymph nodes >1 cm in more two lymph node stations; paraclinical findings

may include PCR >10 mg/dL, anemia, and alterations in platelet count and ESR (erythrocyte sedimentation rate) (>15 mm/h). The diagnosis is confirmed through biopsy. Histologically, the disease can differ into three groups when idiopathic multicentric: hypervascular histological subtype, plasma cell subtype, and mixed subtype. If related to HHV-8, the subtype is plasmablastic. [1,3]

For treatment, various therapies can be combined, including radiotherapy, chemotherapy, steroid use, and/or surgical resection[1]. The unicentric variant is often asymptomatic, manifesting as a slow-growing mass identified incidentally in imaging studies, as in the patient of the present study. Symptoms can sometimes develop, associated with a mass effect compressing adjacent structures[1]. This variant appears more frequently in the fourth decade of life, emphasizing the versality of age at presentation, and is seen more frequently in women[2]. Laboratory studies are typically normal, and tomography may reveal a mass that can be either homogeneous or heterogeneous, with calcifications present in 5 to 10% of cases, showing hyper uptake on contrast-enhancing studies[1]. Fine-needle aspiration is not recommended because of the risk of bleeding and the possibility of obtaining an insufficient sample for a definitive diagnosis. According to histopathological studies, classification can include plasma subtype, vascular hyaline, and mixed, as determined in the patient in this case, who presented with mixed Castleman's disease (vascular hyaline and rich in plasma cells)[3].

Management in localized disease consists of complete surgical resection, which is considered the gold standard. Recurrence is rare and is usually attributed to incomplete initial resection or lymphadenopathies not identified during the initial assessment, leading to surgical intervention as the chosen treatment for our patient. Occasionally, angioembolization may be used to reduce the risk of hemorrhage, particularly in large and vascularized adenomegaly[4,5]. A mass may be considered unresectable when its location involves vital structures such as major blood vessels; if asymptomatic and unresectable, management consists of follow-up, or in symptomatic unresectable cases, treatment may involve immunomodulation, immunosuppression, angioembolization, or radiotherapy[3,4].

Post-treatment follow-up should occur one month after treatment to check for symptoms or findings on physical examination and laboratory and imaging studies. Subsequent follow-up is carried out annually, and imaging studies may be discontinued after five years if the patient remains disease-free. In cases of disease recurrence, positron emission tomography and computed tomography (PET/CT) scans may be requested to assess the potential development of lymphoma[5].

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