Orbital Castleman Disease: Rare Case

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Abstract

Castleman's disease is a lymphoproliferative disease of the lymph node follicles known as angiofollicular lymph node hyperplasia. Introduced by Dr. Benjamin Castleman in 1956. Tree type of histopathology Castleman Disease Hyaline is Vascular Cell, Plasma Cell and Mix type. The incidence of orbital cavities is very rare, with less than 17 case reports in the world. The cause is unknown, it resembles other tumors. Herein, we reported a case of orbital Castleman's disease followed by reviewing the literature to provide an optimal diagnostic approach.

Keywords: Castleman's disease, Hialine Vascular Type, Orbital

Introduction

In general, Castleman disease is a disease that attacks the lymph nodes. However, several case reports state that Casteman's disease can occur outside the lymph nodes. The classification of Castleman disease is divided into solitary or unicentric and scattered/multicentric. Histopathologically, Castleman disease is divided into hyaline-vascular type, plasma cell type, and mixed type. Castleman disease affecting the orbit is a rare case. Cases of Castleman disease involving the orbit have been reported in less than 15 cases in the world. The first case reported was of the multicentric type of Castleman disease, affecting the orbits bilaterally and involving systemic disease. Clinical symptoms of Castleman disease affecting the orbit include proptosis, ptosis, and mass in the conjunctiva. Unicentric Castleman disease involving the orbit is clinically mild, but multicentric Castleman disease involving systemic disease can have a poor prognosis. Herein, we reported a case of Orbital Castleman disease followed by reviewing the literature to provide an optimal diagnostic approach [(1–6)].

Case Presentation

A 61-year-old man complained that his left eye felt proptosis for 1 year before he was admitted to the hospital (figure 1). Complaints are felt to be getting worse because the

eyes are protruding bigger. The patient's complaints were not accompanied by painful eyes, red eyes, blurred vision or double vision. The patient had no complaints of lumps or tumors anywhere else on his body. CT scan results showed extraconal masses in bilateral orbits, suspicious for lymphoma, lacrimal gland tumor, metastasis or dermoid (Figure 2). Then the patient underwent the first biopsy at Cicendo Eye Hospital with Reactive Hyperplasia a/r Left superior ocular palpebra and a differential diagnosis of low grade malignant lymphoma. Then this patient was advised to check IHC CD20, CD3, BCL2, BCL6, Cyclin D1, CD10, Ki67 (figure 4). With normal results, the conclusion is that the lesion is benign. Then the patient underwent resection of the tumor mass in the right orbit and a histopathological examination result was a relatively well circumscribed solid mass was excised and sent for histological examination. The specimen consisted of a fragment of soft tissue measuring 4,5 cm × 3,2 cm × 1,7 cm. On section, the cut surface appeared whitish and smooth. The biopsy showed Reactive Hyperplasia with thick capsule, prominent proliferation of blood vessels accompanied by hyalinization of the blood vessel walls. The germinal center is penetrated by vessels to form a lollipop appearance. The mantle zone is thickened with lymphocyte cells arranged in layers like onion skin (onion skin appearance). Among them is visible proliferation of follicular dendritic cells (figure 3). Suggestions for immunohistochemical examination of CD21, CD23, CD138 (figure 4). The preparation was subjected to immunohistochemical examination at Hasan Sadikin Hospital with the conclusion of Castleman disease, hyaline vascular type.

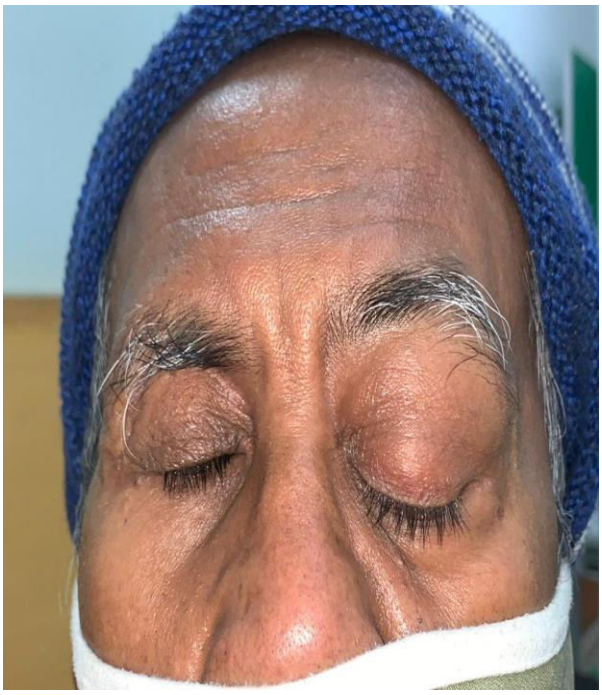

Figure 1: proptosis in left eye

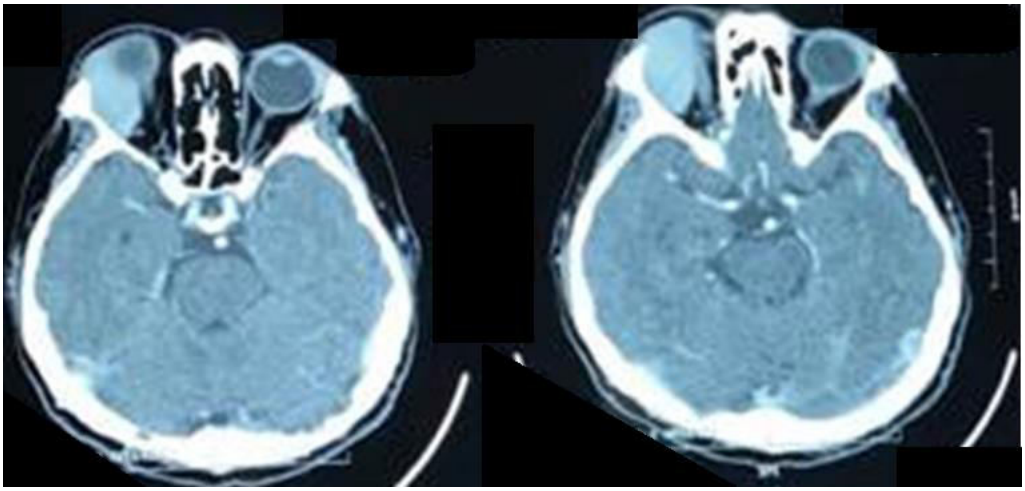

Figure 2. masses in bilateral orbits, suspicious for lymphoma, lacrimal gland tumor, metastasis or dermoid.

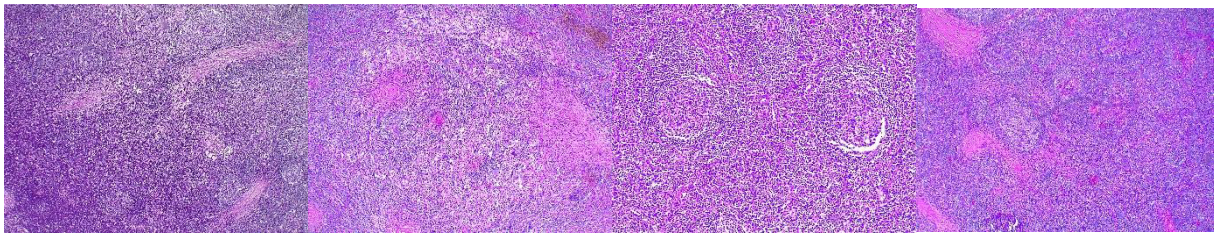

Figure 3. lollipop and union skin appearance in castlemant disease hialin vascular type

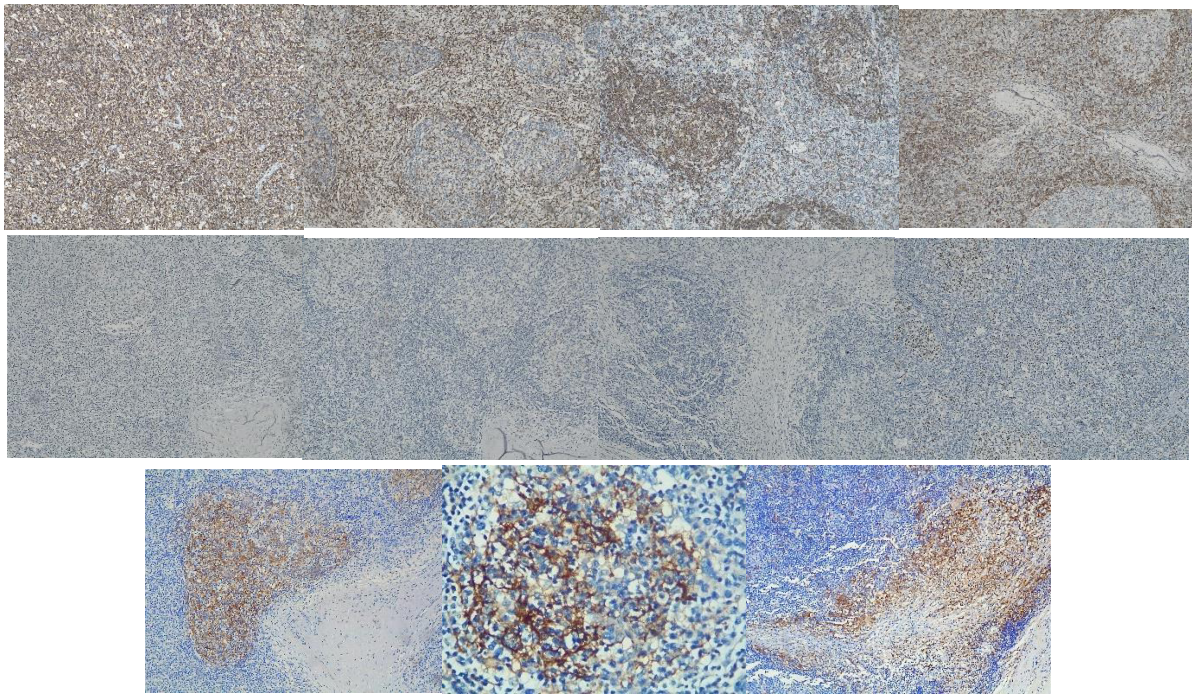

Figure 4. CD45; normal positive, CD 20: normal positive, CD3: normal positive, BCL2: negative, BCL6: Negative, CD10 : normal positive, CylinD1: negative, Ki67: normal positive, CD21 :positive, CD23: positive, CD138: positive serounding vaskulare

Discussion

Castleman disease is a rare lymphoproliferative disease that attacks the lymph nodes. Research and case reports report the occurrence of Castleman disease outside the lymph nodes. Less than 17 cases have been reported of Castleman disease involving the orbit. Castleman disease involving the orbits can occur at any age. Castleman disease involving bilateral orbits usually occurs in old age with a predilection for men more often than women. Castleman disease involving the orbit is one of the 27% incidence of lymphoproliferative disease in the orbit. As many as 90% of Castleman disease that occurs in the orbit is of the hyaline-vascular type. The hyaline-vascular type is usually asymptomatic, limited to the infraorbital area, or can involve the extraorbital without systemic disease involvement. Symptoms that can appear in Castleman disease in the eye socket are proptosis, ptosis, enlargement of the lacrimal gland, perioptic edema, and eye cavity masses. The pathogenesis of Castleman disease is still unclear[1–5]. Some literature mentions the aberrant production of secondary interleukin 6 (IL-6) stimulated by HHV-8 or the presence of unknown exogenous and endogenous factors that can cause plasma cell proliferation and inhibit the apoptosis process[1]. This patient is a 60 year old man who complained of proptosis in the left eye. These complaints are not accompanied by red eyes, blurry or double vision. This is in accordance with the case report reported in Mexico[3]. In this patient, there was also limited eye movement in the left eye, especially upper and lateral eye movement, in accordance with the case reported in Korea[6].

CT scans with contrast in Castleman disease generally show a well-defined mass with stinging after contrast injection. This picture is not typical so it can resemble the appearance of other tumors in the orbit. The presence of tumor destruction or expansion depends on the size of the tumor itself. This patient from CT scan results showed an extraconal mass in the bilateral orbits, suspicious for lymphoma, lacrimal gland tumor, metastasis or dermoid. However, the infrequent occurrence of CD has led to alimited analysis of its imaging characteristics as well as susceptibility to misdiagnosis during diagnostic imaging[2,5].

Histopathologically, Castleman disease is divided into two types, namely hyaline type and plasma cell type Castleman disease. The histological type most often found by pathologists is the hyaline vascular type. The hyaline vascular type is characterized by hyalinization in the germinal center area accompanied by dilatation of blood vessels, as well as concentric lymphocyte infiltration in the mantle zone. Apart from that, vascular proliferation was also seen in the interfollicular tissue. The plasma cell type of Castleman disease is characterized by proliferation of interfollicular plasma cells[5]. In this patient, the microscopic picture shows a tumor mass with a thick capsule at the edges, prominent proliferation of blood vessels accompanied by hyalinization of the blood vessel walls. The germinal center is penetrated by vessels to form a lollipop appearance. The mantle zone is

thickened with lymphocyte cells arranged in layers like onion skin (onion skin appearance). Among them is visible proliferation of follicular dendritic cells and moderate infestation of plasma cells and histiocytes. So this patient fits the description of Castleman's hyaline vascular type disease.

Immunohistochemical examinations carried out to confirm Castleman disease include CD21, CD23, CD35, EGFR which show an increase in follicular dendritic cells in the germinal center. Apart from that, CD123, TCL1 can be examined to show plasmacytoid dendritic cells. In this patient, CD21 and CD23 tests were positive in the germinal centrum. The differential diagnosis of Castleman disease includes other lymphoproliverative lesions, malignant lymphoma, or other solid lesions in the orbital cavity. In other lymphoproliverative lesions, the typical features found in Castleman disease are not found. Lymphoid follicles with centrum germinativum were within normal limits. If granulomatous infection is suspected, signs of granulomatous inflammation will be seen on the microscopic appearance. The next differential diagnosis is lymphoma. The type of lymphoma that is most often used in the differential diagnosis of Castleman disease, especially the hyaline vascular type, is angioimmunoblastic T-Cell Lymphoma, mantle cell lymphoma. Follicular lymphoma shows follicles of various sizes that replace the architecture of the centrum germinativum, there is no appearance of vascular hyalinization, and displays positive immunohistochemical expression on CD10 and Bcl6 staining. Mantle cell lymphoma is microscopically characterized by the presence of prominent lymphoid follicles surrounded by lymphocyte cells. There was no vascular hyalinization or vascular proliferation into the stroma, and showed positive CD5 and Cyclin D1 images. Angioimmunoblastic T Cell Lymphoma is characterized by proliferation in the paracortex, sometimes accompanied by many eosinophil cells. There was also proliferation of venule endothelial cells, and the presence of atypical cells with clear cytoplasm[1–6].

Therapy for Castleman disease of the orbit is surgery followed by administration of neoadjuvant chemotherapy. If the ocular is involved, steroid administration is necessary followed by radiotherapy. The prognosis for patients with unicentric Castleman disease is excellent, with a 10-year survival rate of more than 95%. However, if the patient does not receive radiotherapy, the average survival rate in 10 years is 80%. This patient has had surgery, and currently the patient is being observed every 6 months to see if there is any recurrence[7].To conclude, CD must be included as a differential diagnosis of a well-defined orbital lesion. To the best of our knowledge, this is the first case report of a multicentric, Hialinvasculare variant of orbital CD in the absence of systemic features.Castleman's disease among the differential diagnoses including lymphoma, hemangioma, and other masses. Understanding the histopathologic and clinical feature of Castleman's disease is important and from an accurate diagnosis, appropriate treatment

methods can be selected. A step-wise multidisciplinary approach is crucial for an early diagnosis and appropriate treatment.

Ethics

Informed Consent: Obtained.

Authorship Contributions

Concept, Design, Data Collection or Processing, Analysis or Interpretation, Literature Search, Writing: RJ

Conflict of Interest: No conflict of interest was declared by the authors.

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