

Unicentric Castleman's disease in the orbit: A case report

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A 53-year-old man presented with a palpable mass on the left lower eyelid and occasional diplopia. Under suspicion of orbital lymphoma, an excisional biopsy was performed, and

histopathology revealed Castleman's disease. Castleman's disease is a rare disorder of the lymphoid system, and only a few cases of Castleman's disease in the orbit have been reported.

Key words: Castleman's disease, lymphoid system, orbit

Castleman's disease is a rare atypical lymphoproliferative disorder with three histopathological types: Hyaline vascular, plasma cell, and mixed (transitional) cell type. The disease also has two clinical types: Unicentric and multicentric.^[1,2]

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The mediastinum is the most common site of occurrence and the first reported site by Castleman and Towne in 1954.^[3] Intra-orbital involvement of Castleman's disease is rare. This report discusses a hyaline vascular type of Castleman's disease in the orbit.

Case Report

A 53-year-old male presented with a painless, hard, palpable mass around the left lower lid with 4 months duration. On examination, bilateral best corrected visual acuity was 20/20. Under slit lamp examination, mild chemosis was seen on the left bulbar conjunctiva. There was mild eye movement limitation of the lateral downward gaze of the left eye. Hertel exophthalmometric measurements were 19 mm oculus dexter and 21 mm oculus sinister [Fig. 1]. During the visit, the patient did not complain of diplopia but described occasional episodes of double vision in the recent past. His medical history was otherwise unremarkable. Computed tomography (CT) scan revealed well enhancing, lobular hyperdense mass in left intraconal space [Fig. 2]. With a high clinical and imaging suspicion of lymphoma, an excisional biopsy was performed.

A relatively well circumscribed solid mass [Fig. 3] was excised and sent for histological examination. The specimen consisted of a fragment of soft tissue measuring 3.2 cm × 2.6 cm × 1.3 cm. On section, the cut surface appeared whitish and smooth. The sample had a 0.1 cm resection margin. The biopsy showed proliferation of lymphoid follicles with depletion of germinal center lymphocytes and proliferation of dendritic cells and prominent vascularity of the germinal centers. One follicle contained two or more three germinal centers ("twinning") [Fig. 4a-d]. Immunohistochemical staining for CD3 and CD20 demonstrated follicular hyperplasia, and CD21 and CD23 staining showed dendritic cell proliferation. The CD5 and cyclin D-1 stains were applied to exclude the possibility of malignant lymphoma, including mantle cell lymphoma. The Ki-67 proliferation marker highlighted some remaining germinal centers. Overall, the mass was revealed to be Castleman's disease of the extranodal, hyaline vascular variant type.

The patient was referred to the Hematology-Oncology Department for systemic workup and treatment. Under the diagnosis of Castleman's disease of the orbit, chest, abdomen and pelvis CT scans were obtained, which were unremarkable. The patient's lab findings (complete blood count, liver function test, lactate dehydrogenase) were normal. The patient is on the 13 months of follow-up till the writing of this report and radiotherapy will be considered if there is recurrence of mass. Follow-up CT scans have been obtained every 3 months, and no specific findings including recurrence of mass have been observed.

Discussion

In 1954, Castleman published a case report that described the eponymous disease for the 1st time.^[3] Castleman's disease within the orbit is extremely rare, the exact incidence unknown, and there have been few reported cases. Until now, only a few cases of Castleman's disease in the orbital area have been published, with the most common being the unicentric, hyaline vascular type.^[4-8]



Figure 1: Initial external photograph. Mild chemosis and exophthalmos were seen with the left eye



Figure 2: Computed tomography revealed a solid, lobular hyperdense mass conforming to the shape of the posterolateral aspect of the left globe in left intraconal space. There were lack of bony involvement, molding around normal structure



Figure 3: A solid, well-circumscribed, 3.2 cm × 2.6 cm × 1.3 cm solid mass was excised

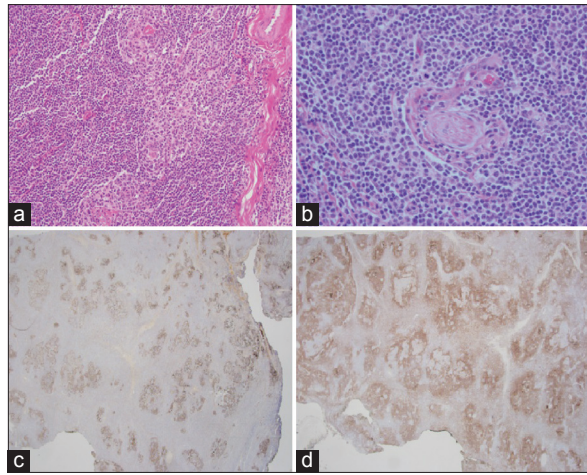


Figure 4: (a) Histology of the tissue showed proliferation of lymphoid follicles with depletion of germinal center lymphocytes and proliferation of dendritic cells (H and E, $\times 200$). (b) The "onion-skinning" of mantle zone lymphocytes, together with the prominent central vessel, has been likened to the appearance of a lollipop (H and E, $\times 400$). (c and d) Immunohistochemical stains of CD21 and CD23 demonstrated follicular hyperplasia

Histopathologically, Castleman's disease can be classified as a hyaline vascular, plasma cell, or mixed cell type neoplasm, with the hyaline vascular type being the most common.^[1,2] The hyaline vascular type is characterized by hyalinized germinal centers with penetrating blood vessels with surrounding mantle zones of concentric lymphocytes and vascular proliferation of the interfollicular tissue. The plasma cell type is characterized by interfollicular proliferation of plasma cells and is more commonly multicentric than unicentric.

The disease is divided into two clinical subtypes, unicentric and multicentric. The localized type of Castleman's disease commonly presents as the hyaline vascular type. Clinically, most cases present as a solitary mass without other symptoms.^[2] Most multicentric Castleman's disease presents histologic features of the plasma cell type. Recently, a few case of the disease most frequently found in immunosuppressed individuals, especially in HIV-positive patients, is described as human herpes virus-8 (HHV-8) associated type.^[9]

The multicentric type appears as lesions throughout the body, is commonly associated with fever, chills, night sweats, fatigue and may also accompany lymphadenopathy and organomegaly. The prognosis of this type is usually poor. While the unicentric form can be cured with surgical excision, surgery usually isn't an option for multicentric Castleman's disease because of the number of lymph nodes involved.^[7] When treating the multicentric form, treatments such as corticosteroids, chemotherapy, or immunotherapy

should be reach all parts of the body. One or a combination of these methods is selected to try and put the disease into remission.^[7,10] Anti-viral drugs including anti-HIV treatment may also be beneficial in the HHV-8 associated type, thus histopathology and immunohistochemistry are important in reaching a diagnosis and exact classification of this rare disease.^[9]

The currently described patient had a hyaline vascular type mass with no evidence of other tumor foci or systemic spread, and underwent excisional biopsy. For most unicentric cases, no additional treatment is required. Up to the time of this case report, there has been no evidence of tumor recurrence or other complications. Although relatively rare, ophthalmologists should consider 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.

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