



Plasma Cell Type Localized Intraorbital Castleman's Disease: A Case Report

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Abstract

A 76-year-old male presented with a 3-month history of progressive mass in the right eye area. Excisional biopsy revealed a well-circumscribed mass, histopathological and immunohistochemical studies of the orbital mass reveal the features consistent with plasma cell type Castleman's disease (CD). Plasma cell type is not commonly found than hyaline-vascular type in the two types of CD. In the orbit, CD is extremely rare with few reported cases. We report this patient with localized intraorbital Castleman's disease.

Keywords: Castleman's disease; Orbit; Magnetic resonance imaging

Introduction

Castleman's disease (CD) is a rare atypical lymphoproliferative disorder. It was first reported by Benjamin Castleman in 1954 [1]. CD usually presents in the mediastinum (60-75%). In the orbit, CD is extremely rare with few reported cases [2-5]. We report this patient with localized intraorbital Castleman's disease.

Case Presentation

A 76-year-old Chinese male presented with a 3-month history of progressive mass in the right eye area. Ophthalmic examination revealed a soft tissue mass, which was well-circumscribed and unmovable in the inferior eyelid.

Magnetic resonance imaging (MRI) demonstrated a soft tissue mass in the right orbit located in the posterior and the lower aspect of the eyeball. MRI presented T₁ and T₂ isointensity and slightly hyperintensity in fat suppressed sequence. The mass which grew around the eyeball, extruded beyond the orbit and involved in the internal and external pyramidal muscle. The eyeball was compressed and dislocated. There was obvious and homogeneous enhancement (Figure 1). The MRI diagnosis suggested that the benign lesion located in the orbit maybe inflammatory pseudotumor.

Pathological examination: Microscopically, the soft tissue mass were found to be irregular and grey-red in color. Part of the tissue showed node-like structures. Characteristically, the interfollicular zones showed numerous plasma cells and Russell bodies. Additionally, immunohistochemical staining revealed that the majority of the plasma cells expressed CD38 positive stain (Figure 2). Histopathological and immunohistochemical studies of the orbital mass reveal the features consistent with plasma cell type CD.

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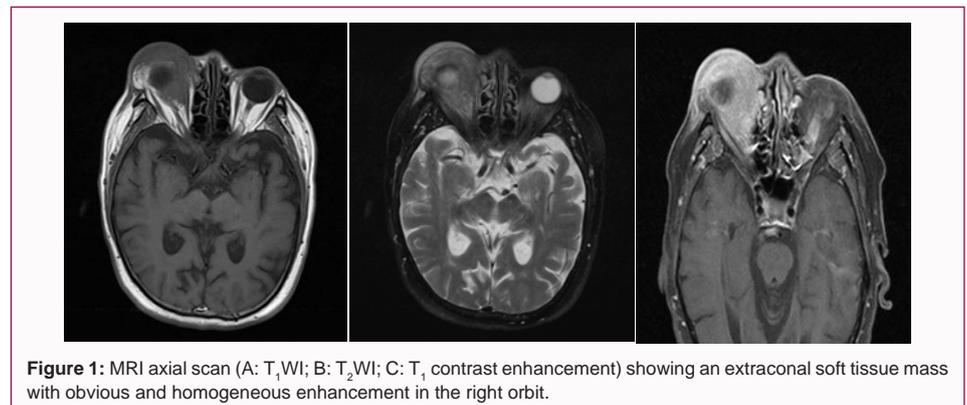


Figure 1: MRI axial scan (A: T₁WI; B: T₂WI; C: T₁ contrast enhancement) showing an extraconal soft tissue mass with obvious and homogeneous enhancement in the right orbit.

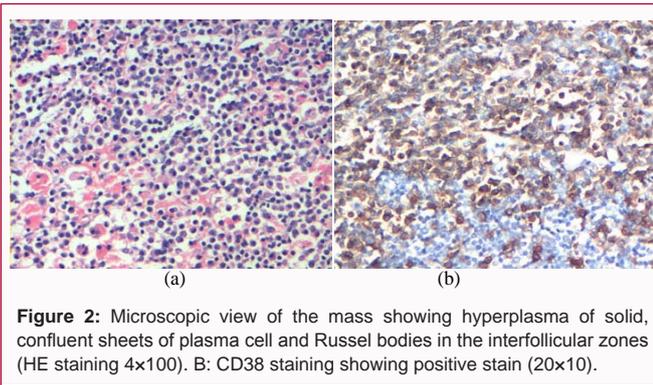


Figure 2: Microscopic view of the mass showing hyperplasia of solid, confluent sheets of plasma cell and Russel bodies in the interfollicular zones (HE staining 4x100). B: CD38 staining showing positive stain (20x10).

Discussion

Clinically, CD occurs in localized and multicentric forms. Pathologically, it is subdivided into two forms: hyaline-vascular type and plasma cell type. Hyaline-vascular type is most commonly found (90%). The characteristic histopathological features of this form are the presence of abnormal follicles with atrophic hyalinized follicular centre and a broad mantle zone of small lymphocytes in a concentric or onion skin arrangement.

The characteristic histopathological feature of the plasma cell type form is the presence of solid, confluent sheets of plasma cells in the interfollicular areas. The follicular centres are usually enlarged and hyperplastic.

The unique sign of this case is the intraorbital soft mass. The differential diagnosis with other orbital mass consist of: 1. Diffuse inflammatory pseudotumor: The mass presents medium and

hypointensity in T_1 weighted image and medium and hyperintensity in T_2 weighted image, which shows the medium enhancement and presents thickening of optic nerves and swelling of lacrimal gland. 2. Cavernous hemangioma. The mass presents hypointensity or isointensity in T_1 weighted image and hyperintensity in T_2 weighted image, which shows the character of gradual enhancement.

The case reported in this paper presented isointensity in T_1 and T_2 weighted image, which was different from the other mass in the orbit. The enhancement was different from cavernous hemangioma. Optic nerve and cellulitis gland were not involved, which was different from inflammatory pseudotumor. It is therefore suggested that CD should be considered when the atypical mass was found in the orbit. Histopathological examination needs to be conducted in order to confirm the diagnosis.

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