CASE REPORT

Diffuse large B-cell lymphoma masquerading as orbital cellulitis

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Proptosis is commonly encountered in clinical practice. We report on a patient with acute proptosis, eyelid swelling, and chemosis, which was initially treated as cellulitis. After radiological and pathological assessments, a diagnosis of large B-cell lymphoma was made. The patient died within 2 months of presentation. Not all patients with proptosis have cellulitis. Proptosis, especially unilateral proptosis, should always lead to a radiological scan in case of malignancy.

Introduction

Orbital lymphoma and lymphoma of the orbital adnexae are relatively rare lymphomas, representing 0.1% of all lymphomas.[1] Lymphomas of B-cell lineage are more likely to be associated with symptoms related to the eyes and to have extension to the orbit than are lymphomas of T- or NK-cell lineage.[2] The most common clinical signs of orbital lymphoma are proptosis, a slow-growing palpable mass, and painless swelling of the eyelids.[3] This report is of a patient with orbital large B-cell lymphoma presenting with acute proptosis and eyelid swelling. The initial diagnosis was orbital cellulitis. The correct diagnosis was made after referral to the Department of Ophthalmology.

Case report

In late July 2009, a 70-year-old previously healthy man presented to the Department of Ophthalmology, Caritas Medical Centre, Hong Kong, with left eye swelling for 10 days and acute proptosis for 1 day. He had initially been treated elsewhere for orbital cellulitis and was given oral ampicillin and cloxacillin, and topical chloramphenicol eye drops, with no improvement in symptoms. At presentation to the Department of Ophthalmology, his visual acuity was 6/20 in the right eye and hand movements in the left eye. Left extra-ocular movements were limited in all directions. There was left eyelid swelling, proptosis, and chemosis (Figs 1a, 1b). Hertel measurements were 12 mm in the right eye and 23 mm in the left eye. Slit-lamp examination revealed bilateral cataracts. Fundus examination was normal for the right eye, but the optic disc was swollen in the left eye. The vessels were tortuous and choroidal folds were present in the left fundus (Fig 1c).

Computed tomography (CT) of the orbit showed a large soft tissue mass (5 x 3 cm) with homogeneous contrast enhancement extending from the left upper nasal cavity to the left maxillary antrum, left orbit, and left ethmoid and frontal sinuses. Bony destruction was seen at the left ethmoid and frontal sinuses, the medial wall of the left orbit, and the medial wall of the left maxillary antrum. The left globe was displaced anteriorly and laterally, and deformed, resulting in proptosis (Figs 2a to 2d).

Biopsy of the nasal mass was performed by ear, nose, and throat surgeons. Microscopic examination showed large lymphoid cells with round nuclei and prominent nucleoli. The cells were positive for B-cell marker CD20, but negative for T-cell marker CD3, NK-cell marker CD56, and epithelial marker AE1/3. A diagnosis of diffuse large B-cell lymphoma was confirmed. Systemic survey with brain and abdominal CT and bone scan showed brain metastasis with a 5 x 6–cm hypodense lesion in the left frontal lobe and midline shift. Orbital involvement of the tumour was progressive (Figs 2e, 2f). The visual acuity of the left eye further deteriorated to no light perception. The patient and his relatives preferred debulking surgery to radiotherapy or chemotherapy. However, the disease was too advanced for either radical or palliative treatment. The general condition of the patient deteriorated rapidly and, before any treatment could be offered, he died in September 2009, less than 2 months since presentation.

Discussion

Diffuse large B-cell lymphoma (DLBCL) is a high-grade lymphoma that commonly

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presents with systemic involvement.[4] More than half of the orbital and ocular adnexal lymphomas are of the mucosa-associated lymphoid tissue (MALT) lymphoma subtype, whereas DLBCL predominates intra-ocularly.[5] In a study of 128 South Korean patients with ocular lymphoid tumours, DLBCL represented 4.7% of all lymphoproliferative diseases and was the second most common lymphoma.[6] Orbital lymphoma typically presents as proptosis or a palpable mass causing eyelid swelling.[3,4,7] Other symptoms include restricted extra-ocular movement, diplopia, and pain. Decrease in visual acuity is uncommon unless there is optic nerve infiltration or compression caused by the

擬似眶蜂窩組織炎的彌漫大B細胞淋巴瘤

眼球突出在臨床上很常見。本文報告一名患有急性眼球突出、眼瞼腫脹及球結膜水腫的病人，他初時被診斷為患有眶蜂窩組織炎。後經影像及病理學檢查確診為大B細胞淋巴瘤。病人於病發後兩個月死亡。眼球突出並不一定代表有眶蜂窩組織炎。如果遇上眼球突出的病人，尤其是單眼球突出，應為病人進行影像學檢查以確定是否患有惡性腫瘤。

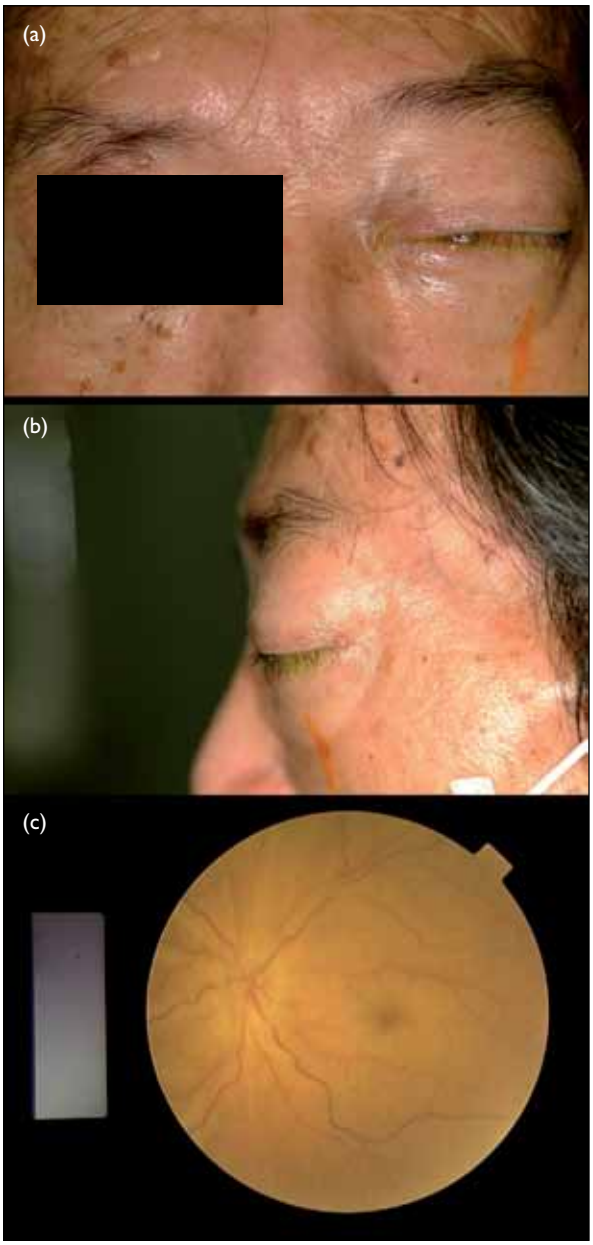

FIG 1. (a, b) A 70-year-old man with left proptosis, eyelid swelling, and chemosis. (c) Fundus photograph of the patient's left eye showing optic disc swelling, tortuous vessels, and choroidal folds

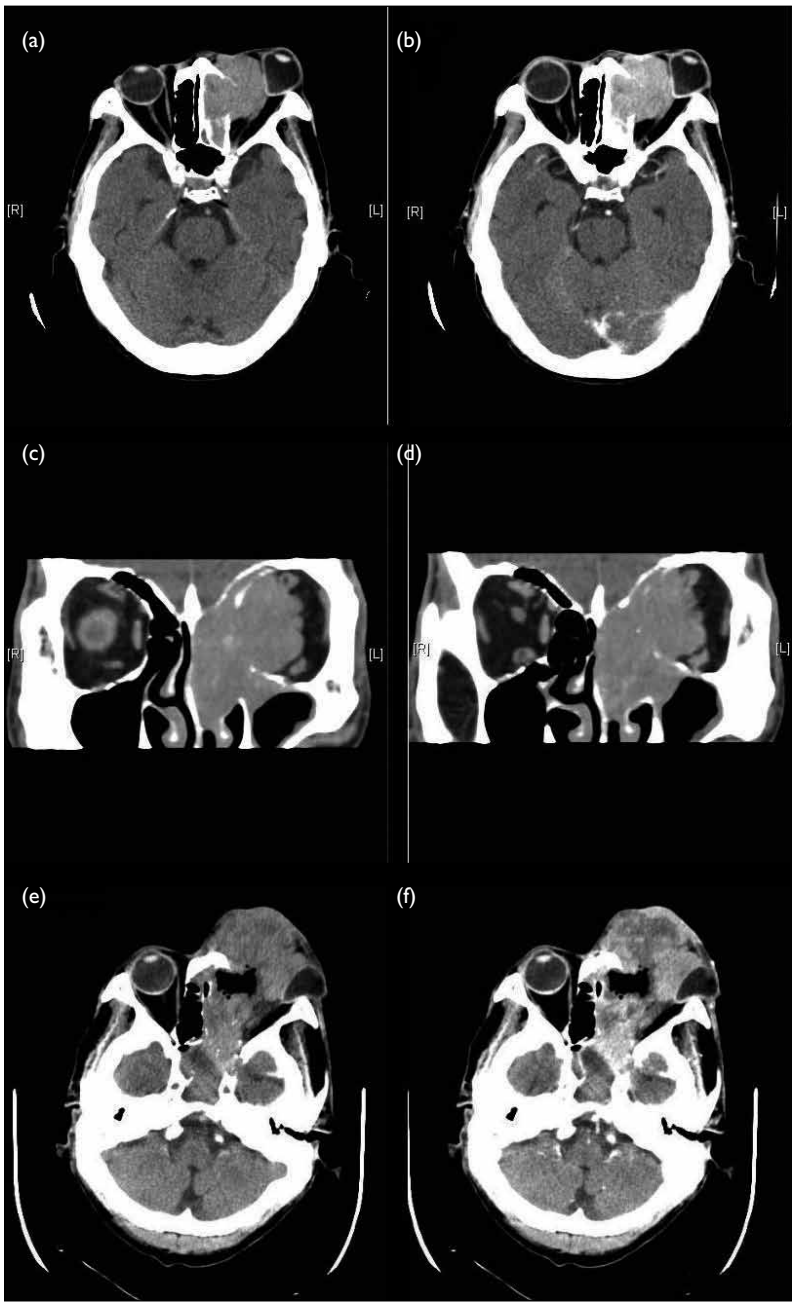

FIG 2. (a) Axial plain cut, (b) coronal plain cut, (c) axial cut with contrast, and (d) coronal cut with contrast computed tomographic scan of the orbits at presentation showing a large soft tissue mass extending from the left upper nasal cavity to the left maxillary antrum, left orbit, and left ethmoid and frontal sinuses. (e) Plain cut and (f) with contrast computed tomographic scan of the orbits 7 weeks after presentation showing progression of the tumour

mass.[8] Imaging is necessary to aid the diagnosis and to delineate the extension of the lesion. Diagnosis relies on pathological examination of a biopsy specimen because the most appropriate treatment modality requires both grading and staging of the disease. It is generally accepted that local control can be achieved with radiotherapy for lymphoma localised to the ocular region.[9,10] Chemotherapy or a combination of chemotherapy and radiotherapy is given to patients with systemic spread.[11] Different subtypes of lymphoma are diverse in clinical behaviour and prognosis, with patients with MALT lymphoma having more favourable outcomes than patients with other types of lymphoma.[6]

Orbital lymphoma may be misdiagnosed as cellulitis.[12-14] Orbital cellulitis is an infection of the orbital soft tissues posterior to the orbital septum. In most patients, cellulitis occurs as a direct extension of infection of the paranasal sinuses. This patient was initially diagnosed with and treated for orbital cellulitis. Proptosis, eyelid swelling, and chemosis may mislead clinicians to diagnose orbital cellulitis. However, the absence of response to antibiotics implied other pathology, which explained the persistent eyelid swelling and proptosis, and later resulted in complete loss of sight in his left eye and finally death within 2 months of presentation.

Proptosis, especially unilateral proptosis, should always lead to a CT or magnetic resonance imaging scan to exclude malignancy. A biopsy will be needed to make a diagnosis of lymphoma. Clinical alertness, together with radiological investigation and adequate pathological sampling, are necessary for correct diagnosis and treatment. In case of uncertainty, timely referral to an ophthalmologist is mandatory.

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