CASE REPORT

Unusual eye signs in Wegener's granulomatosis

Alice YK Chan 陳綺琪
Edmund K Li 李國銘
Paul CL Choi 蔡祥龍
David TL Liu 劉大立
Philip TH Lam 林子顯
Dennis SC Lam 林順潮

Wegener's granulomatosis is a rare necrotising vasculitis not easily diagnosed due to the obscurity of its diverse clinical features. Despite its comparatively low incidence, the unusual ophthalmic manifestations seen in this disease warrant extra caution from attending rheumatologists. In this case, bilateral peripheral ulcerative keratitis preceded any systemic symptoms. Timely recognition of the significance of this ophthalmic complaint and prompt ophthalmological consultation can help achieve early diagnosis and treatment of this potentially fatal rheumatological disease.

Introduction

Wegener's granulomatosis is a rare inflammatory disease of unknown aetiology.[1] Clinical manifestations are diverse and sometimes ambiguous. Early diagnosis is difficult. Ocular involvement may be the sole clinical presentation prior to systemic symptoms.[2] Common ocular presentations of Wegener's granulomatosis are proptosis, orbital pseudotumour, and nasolacrimal duct obstruction. They are believed to be the result of contiguous spread of long-standing granulomatous sinusitis. Uncommonly, patients can have focal vasculitis involving either the anterior or posterior segment of the eye in the form of conjunctivitis, episcleritis, scleritis, peripheral ulcerative keratitis, uveitis and, rarely, optic nerve vasculitis or retinal artery occlusion. Recognition of these unusual ophthalmic features coupled with prompt investigations enables early diagnosis and treatment. We report bilateral corneal ulcers as the manifestation heralding Wegener's granulomatosis in a Chinese patient.

Case report

A 62-year-old woman presented to the Prince of Wales Hospital, Hong Kong in June 2005 with hypertension, hyperlipidaemia, and diabetes mellitus. She was treated with insulin, lisinopril, felodopine, and simvastatin. Two weeks prior to admission, she had painful red eyes and a loss of vision. Nasal blockage, thick rhinorrhoea, weight loss, and fever followed afterwards. Both eyes showed chemosis, striking round-the-clock peripheral corneal ulceration and furrowing (Fig). The central corneas were unaffected. Background diabetic retinopathy was the only ophthalmoscopic finding. Her visual acuity was 20/200 in the right eye and 20/70 in the left eye. Her nasal mucosa was swollen but a systemic review was unremarkable. A full blood count found a normochromic normocytic anaemia and thrombocytosis (haemoglobin, 76 g/L; mean corpuscular volume, 80.4 fL; platelet count, 796 x 10^9 /L; white cell count, 12.9 x 10^9 /L). Her erythrocyte sedimentation rate was elevated (130 mm/h) and the anti-nuclear antibodies were positive (1:160). The c-anti-neutrophil cytoplasmic antibodies (C-ANCA) were positive with anti-proteinase 3 greater than 200 RU/mL. Cultures of corneal scrapings grew no organisms. A corneal biopsy showed chronic inflammatory cells infiltrating lamellar stromal tissue only. A plain radiograph of the lung revealed bilateral air space shadowing in the upper zone predominantly affecting the left side. Computed tomography of the head, neck, and chest revealed enlarged lacrimal glands, pansinusitis, multiple lung parenchymal soft tissue lesions, and a 3-mm hypodense pulmonary embolus in the right pulmonary artery. The nasal cavity was aspirated and lavaged but cultures grew no bacteria or fungus. A nasal biopsy showed inflammatory exudates with no evidence of vasculitis or malignant change. A bronchoscopy showed oedematous and erythematous mucosa and whitish yellow patches in the mucosa of the left upper lobe. The vocal cords, trachea, and carina were normal. A culture of bronchoalveolar lavage for bacteria, mycobacterium tuberculosis, and pneumocystic pneumonia were all negative. A transbronchial biopsy revealed focal capillaritis but no definite necrotising granuloma. An echocardiogram showed an elevated pulmonary arterial systolic pressure of 45 mm Hg. Anticoagulants were started to manage the pulmonary embolism. The clinical picture was compatible with Wegener's granulomatosis and oral prednisolone 1 mg/kg/d and oral cyclophosphamide 2 mg/kg/d were commenced. She showed gradual improvement after treatment. Twenty-four months after her initial presentation, the visual acuity in both eyes had returned to 20/40. The peripheral corneal ulcer had healed but shallow peripheral corneal thinning (gutters) resulted.

Key words
Vasculitis; Wegener's granulomatosis

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Social Hygiene Service, Department of Health, Hong Kong
AYK Chan, MRCP
Prince of Wales Hospital, The Chinese University of Hong Kong, Shatin, Hong Kong:
Department of Medicine and Therapeutics
EK Li, FRCP
Department of Anatomical and Cellular Pathology
PCL Choi, FRCPA
Department of Ophthalmology and Visual Sciences
DTL Liu, MRCS
PTH Lam, FRCS
DSC Lam, MD, FRCOphth

Correspondence to: Dr DTL Liu
E-mail: david_tlliu@yahoo.com

韋格納肉芽腫導致眼部不尋常的徵狀

韋格納肉芽腫是一種罕見的壞死血管炎，由於不同病例會出現不同的臨床病徵，此病較難診斷。雖然肉芽腫發生的可能性相對較低，此病症引致眼睛出現的反常徵狀，會使主診的風濕病科醫生加倍留意。在本文的病例中，兩眼周邊潰瘍性角膜炎便較其他系統性病徵較早發生。及時識別這個眼疾背後的原因並且即時諮詢眼科專家的意見，可以及早診斷和治療這個潛在的致命性風濕病。

Discussion

In our patient, bilateral corneal ulcers were an initial manifestation of Wegener's granulomatosis, and markedly preceded the systemic changes. They were caused by sterile peripheral ulcerative keratitis as a result of vasculitic involvement of the limbal vessels at the corneal-scleral junction.[3] The presence of such pathology, coupled with multiple vague complaints, may serve as a diagnostic hint for this potentially fatal necrotising vasculitis.

The clinical manifestations of Wegener's granulomatosis are diverse and upper airway disease is considered the most common presenting feature.[4] Conversely, it is very uncommon to have an ocular manifestation as the initial presentation. Duna et al[5] have shown that ocular involvement is the presenting feature in 16% of patients. Any compartment or layer of the globe may be affected and it is much rarer for peripheral ulcerative keratitis or necrotising scleritis to be the initial presentation.[5] The importance of these conditions lies in the fact that they may indicate an underlying, generalised vasculitic process since their presence involves small vessel vasculitis of the anterior ciliary arteries or perilimbal arteries.[2] It is interesting to note that a relapse of scleritis in Wegener's granulomatosis is not preceded by a significant rise in the ANCA titre. Therefore, unlike systemic vasculitis, ANCA levels do not correlate with ocular disease activity.[6] Anti-neutrophil cytoplasmic antibodies appear to be specific to but not sensitive for Wegener's granulomatosis–associated vasculitic ocular involvement.

In summary, prompt recognition of the keratoscleral manifestation aids early treatment for Wegener's granulomatosis, highlighting the importance of early ophthalmological referral in patients with an unusual corneal presentation and vague systemic complaints.

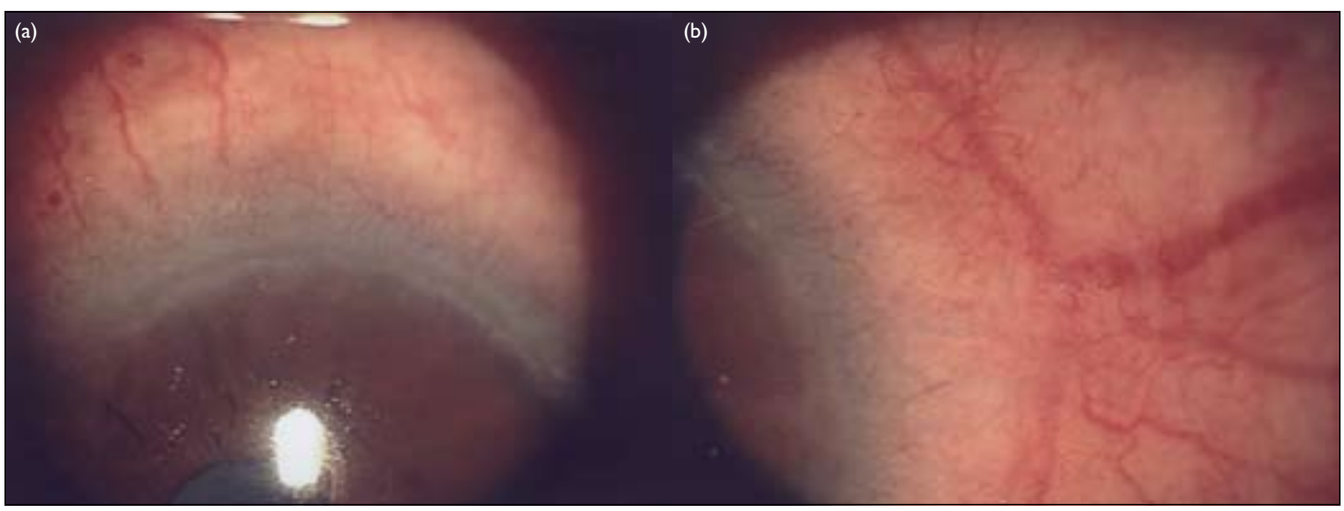

FIG. (a) Right eye and (b) left eye: slit-lamp photomicrographs showed distinctive bilateral circumferential peripheral corneal ulceration with furrowing and guttering. The surrounding conjunctiva was injected and oedematous. Engorgement of the limbal vasculature is evident

References

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