

## Ocular Changes in a Limited Form of Wegener's Granulomatosis: Patient with Cutaneous Ulcer of Upper Eyelid

Toshinobu Kubota and Hiroshi Hirose

*Department of Ophthalmology, Nagoya National Hospital, Nagoya, Japan*

**Background:** We report a patient with a limited form of Wegener's granulomatosis (WG) who presented with ocular changes and an eyelid ulcer in her left eye.

**Case:** A 56-year-old woman complained of ocular pain and discharge. Upon examination, we found evidence of ulcerative keratitis, a mass in the orbit, and an eyelid skin ulcer in her left eye.

**Observations:** The antineutrophil cytoplasmic antibody test was negative, and systemic evaluations showed no specific changes. A biopsy specimen of the left orbit demonstrated necrosis, granulomatous inflammation, and vasculitis. A diagnosis was made of a limited form of WG, while the biopsy specimen of the left upper lid demonstrated granulomatous inflammation without vasculitis.

**Conclusion:** Although the eyelid ulcer was contiguous with the orbital mass, histopathology showed they had distinctive pathological features. **Jpn J Ophthalmol 2003;47:398–400** © 2003 Japanese Ophthalmological Society

**Key Words:** Antineutrophil cytoplasmic antibody, eyelid ulcer, Wegener's granulomatosis.

### Introduction

Wegener's granulomatosis (WG) is a necrotizing granulomatous vasculitis characterized by the involvement of the upper airways, lungs, and kidneys. Orbital involvement and cutaneous ulcers are the most common manifestations of WG. Patients who have cutaneous granulomatous lesions without vasculitis have neither renal nor pulmonary manifestations of WG, and the disease progresses at a slower rate than in patients with vasculitis.<sup>1</sup> We report a patient with a limited form of WG with initial ocular changes and a cutaneous ulcer in the left upper eyelid.

### Case Report

A 56-year-old Japanese woman presented with a 6-month history of swollen left upper eyelid, discharge, and ocular pain. Antibacterial treatment at another hospital failed to arrest the worsening discharge and ocular pain. Visual disturbance and a skin ulcer developed, and she was referred to us.

Her best-corrected visual acuity was 20/20 OD and 6/600 OS. The left upper eyelid was ulcerated, and motility was restricted during upward and rightward gaze, with the left eye displaced temporally. The conjunctiva was markedly hyperemic, and the cornea showed ulcerative keratitis (Figure 1).

The left posterior segment was unremarkable. Computed tomography demonstrated an enlargement of the lacrimal gland and a soft-intensity mass in the left orbit that appeared contiguous to the upper eyelid. The right orbit appeared normal.

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Correspondence and reprint requests to: Toshinobu KUBOTA, MD, Department of Ophthalmology, National Nagoya Hospital, 4-1-1, Sannomaru, Naka-ku, Nagoya-shi, Aichi-ken 460-0001, Japan. Tel.: 52-951-1111; E-mail: ganiky@nnh.hosp.go.jp

The results of chest radiography, serum biochemical screening, and urinalysis were negative. Antineutrophil cytoplasmic antibody (c-ANCA) tests of repeated samples were consistently negative. Cultures of bacteria and fungi were also negative.

A biopsy specimen of the left orbit demonstrated necrosis, granulomatous inflammation, and vasculitis (Figure 2).

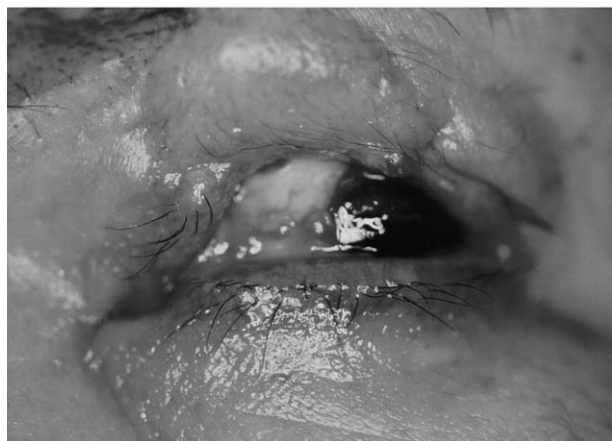
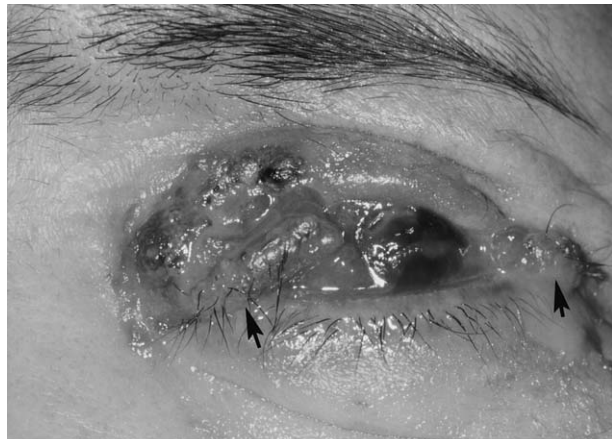
A biopsy of the left upper lid, on the other hand, demonstrated granulomatous inflammation and necrosis but no vasculitis.

A diagnosis of Wegener's granulomatosis was made, and oral treatment was initiated with 100 mg/day cyclophosphamide and 60 mg/day prednisolone. The discharge and ocular pain dramatically lessened. At the 10-month follow-up examination, the visual acuity remained as hand motion. The strabismus and restricted ocular movement also persisted (Figure 1). However, the patient was free of ocular pain, discharge, and orbital granulation.

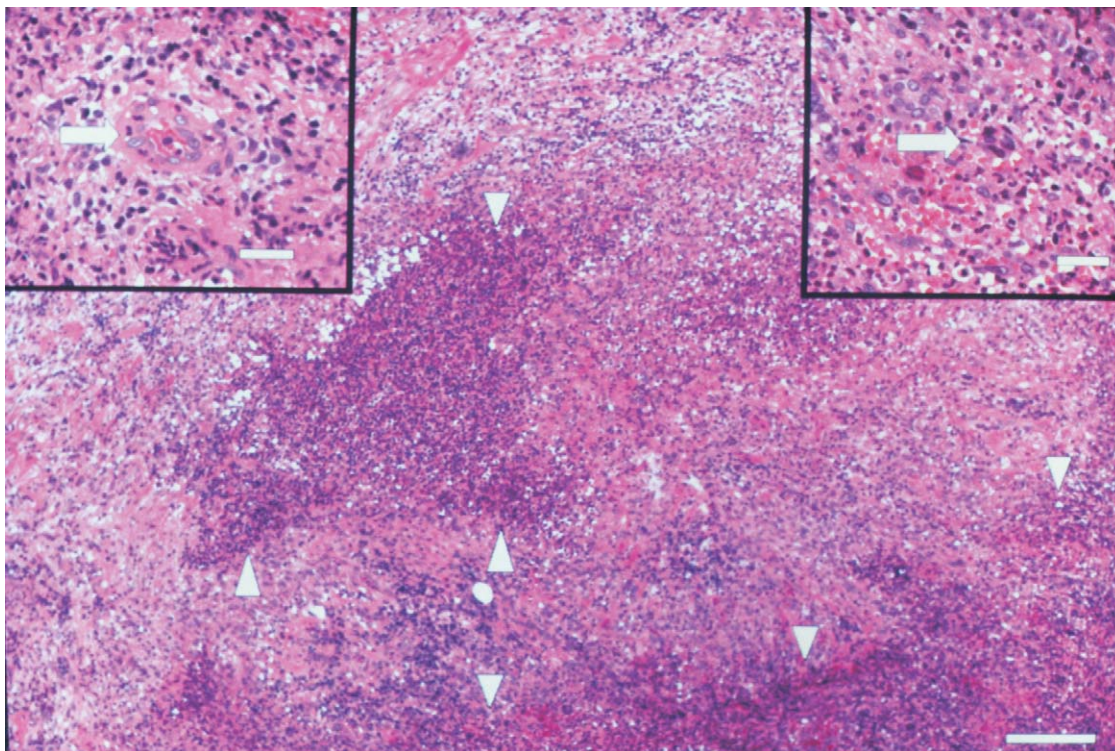
## Discussion

Cutaneous ulcers are subdivided into two histopathological subgroups: granulomatous inflammation and vasculitis.<sup>1,2</sup> WG patients with vasculitis skin lesions have a more rapid progressive course and the lesions are more widespread than in patients with granulomatous skin lesions.<sup>1</sup> Our case demonstrated ocular changes in a limited form of WG complicated by eyelid ulcer. The eyelid ulcer was contiguous with the orbital mass, but a biopsy specimen of the orbit demonstrated necrosis, granulomatous inflammation, and vasculitis, while a biopsy specimen of the left upper lid demonstrated granulomatous inflammation without vasculitis. This distinctive cutaneous pathological feature might be a guideline to whether a lesion will become widespread or not.

Ocular manifestations of limited forms of WG present with a variety of clinical symptoms,<sup>3</sup> and the disease in our case was initially misdiagnosed as an orbital infection on the basis of the patient's eyelid inflammation and discharge. To prevent irreversible ocular complications through failure to make an early diagnosis, WG should be considered whenever ocular pain, discharge, and eyelid inflammation are present. Our case presented with atypical manifestations including negative c-ANCA and ulcerated upper eyelid. This suggests that the initial symptoms of WG can include ocular pain, discharge, corneal ulcer, orbital mass, eyelid ulcer, and negative c-ANCA.



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**Figure 1.** Top: The left upper eyelid is grossly swollen and indurated with discharge in a 56-year-old woman found to have a limited form of Wegener's granulomatosis. The conjunctiva of the left eye is markedly hyperemic and keratitis is present. The middle of the tarsus is not present, and the edge of the tarsus is drooping (bow). Bottom: After reconstructive surgery of the upper eyelid, strabismus and restricted ocular movement persisted.



**Figure 2.** Result of histopathological analyses (hematoxylin-eosin). Biopsy specimen shows geographic necrosis (arrowheads, bar = 500  $\mu$ m), giant cells (top right) and vasculitis (top left). Bars = 40  $\mu$ m.

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