Clinical Case Report

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Imaging diagnosis of orbital Wegener granulomatosis

A rare case report

Bin Yang, MS[a,b], Zhijian Yin, MS[c], Shuai Chen, MS[a], Feng Yuan, MS[a], Wei Zhao, MS[a], Yaying Yang, MS[a,*]

Abstract

Introduction: Wegener granulomatosis (WG) is a rare idiopathic autoimmune disease causing necrotizing granulomatous vasculitis. Whether as the first symptom or as part of systemic changes, ocular manifestations in WG patients are not specific. Any part of the eyes can be affected, with the anterior segment and orbit most commonly involved. So, early diagnosis and treatment are essential for controlling the progression of the disease and improving the quality of life for patients.

Clinical findings/Patient concerns: Here we present a rare case of orbital WG of a 22-year-old woman was admitted to the hospital because of intense pain associated with decreased visual acuity in her right eye since 1 day. She had been previously diagnosed with WG at our hospital.

Imaging diagnosis: Orbital computed tomography imaging showed diffuse swelling of intraorbital muscles, and space-occupying lesions were present in both eyes. Most postnasal anatomical structures were absent, appearing as a massive cavity shadow. Orbital magnetic resonance imaging showed a shadow of orbital soft tissues.

Conclusion: WG is a serious, fatal disease. Early diagnosis and treatment are essential for controlling the progression of the disease and improving the quality of life for patients.

Abbreviations: ANCA = antineutrophil cytoplasmic antibody, CT = computed tomography, MRI = magnetic resonance imaging, WG = Wegener granulomatosis.

Keywords: case report, imaging diagnosis, orbital, Wegener granulomatosis

1. Introduction

1.1. Patient

A 22-year-old woman was admitted to the hospital because of intense pain associated with decreased visual acuity in her right eye since 1 day. She had been previously diagnosed with Wegener granulomatosis (WG) at our hospital. Admission examination showed no improvement in corrected vision in the eyes, enophthalmos, and slight conjunctival hyperemia (Fig. 1A). White lesions with neovascularization, central corneal perforation, and iris incarceration were observed around the inferior nasal corneal limbus in her right eye (Fig. 1B). Further, massive neovascularization was observed at the corneal limbus in her left eye, and whitish gray lesions associated with neovascularization were visualized on the superior nasal and temporal sides (Fig. 1C).

2. Materials and methods

2.1. Imaging examination

All the materials and methods were carried out in accordance with guidelines and regulations set by the Affiliated Hospital of Kunming Medical University. All the protocols were approved by the Department of Radiology and Medical Imaging of Affiliated Hospital of Kunming Medical University. And patient provided the written consent on standard forms. Orbital computed tomography (CT) imaging showed diffuse swelling of intraorbital muscles, and space-occupying lesions were present in both eyes. Most postnasal anatomical structures were absent, appearing as a massive cavity shadow (Fig. 1D and E). Orbital magnetic resonance imaging (MRI) showed a shadow of orbital soft tissues (Fig. 1F and G).

3. Results

3.1. Surgery and pathology

The surgically removed conjunctiva was approximately 2×2 mm, dark red in color, and brittle. Histologically, light microscopy revealed invasion by a large number of eosinophils and plasma cells, fibrin-like degeneration, and hyperplasia in the capillary walls (Fig. 1H and I). The pathological diagnosis was conjunctival necrotizing granulomatous lesions with fibrinoid vasculitis.

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BY, WZ, and YY have contributed equally to this work.

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[a] Department of Radiology, The First Affiliated Hospital of Kunming Medical University, Kunming, [b] Department of Radiology, The First Affiliated Hospital of Dali University, Dali, [c] Department of Ophthalmology, The First Affiliated Hospital of Dali University, Dali, China.

[*] Correspondence: Yaying Yang, Department of Radiology, The First Affiliated Hospital of Kunming Medical University, Kunming, China (e-mail: yayingyang@163.com).

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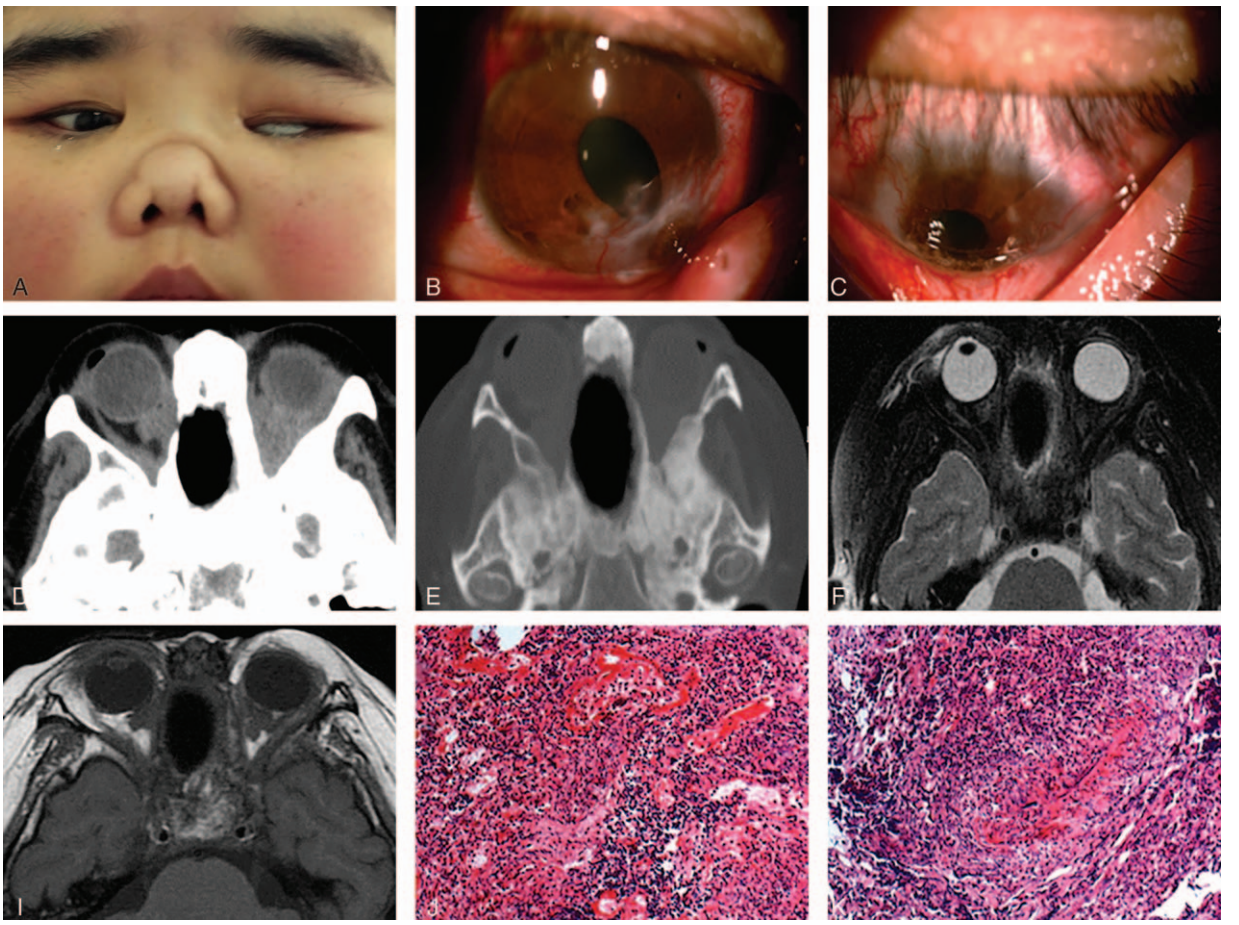

Figure 1. (A) Facial deformity and bilateral enophthalmos. (B) Whitish gray lesions with neovascularization, central corneal perforation, and iris incarceration found around the inferior nasal cornea limbus in the right eye. (C). Massive neovascularization found at the corneal limbus in the left eye, and whitish gray lesions associated with neovascularization found on the superior nasal and temporal sides. (D) CT scan showing bilateral extraocular muscle thickening, orbital soft tissue masses, and a postnasal cavity shadow. (E) CT bone window scan showing bone destruction on the right inner orbital wall and postnasal bones. (F) Bilateral short T2 orbital space-occupying lesions with a signal slightly higher than extraocular muscle and a postnasal cavity. (G) Bilateral iso-T1 orbital space-occupying lesions not distinguishable from extraocular muscles, and a postnasal cavity. (H) Microscopy showing invasion by a large number of eosinophils and plasma cells, fibrinoid degeneration, and hyperplasia in the capillary walls. (I) Microscopy showing invasion by a large number of eosinophils and plasma cells. CT = computed tomography.

4. Discussion

WG is a rare idiopathic autoimmune disease causing necrotizing granulomatous vasculitis.[1] It is more common in Caucasians, with a reported incidence of approximately 3 in 100,000 in the United States. The average age of onset is 40 to 55 years, with no significant sex differences.[2] Upper respiratory tract symptoms are usually the main symptoms, followed by symptoms related to the lungs, eyes, ears, kidneys, and skin.

WG involving the nose can cause chronic rhinitis, sinusitis, or multiple nasal polyps. Ultimately, WG can present as a typical saddle nose due to nasopharyngeal ulcers, bone and cartilage destruction, and nasal septum perforation. Ocular involvement suggests spread from neighboring periorbital and upper respiratory tract lesions, while isolated lesions are caused by local necrotizing vasculitis. Literature reports show that ocular manifestations are the first symptoms in 8% to 16% of patients, and WG will eventually involve the eyes in 50% to 87% of patients.[3] Whether as the first symptom or as part of systemic changes, ocular manifestations in WG patients are not specific. Any part of the eyes can be affected, with the anterior segment and orbit most commonly involved.

Necrotizing granulomatous vasculitis is the main feature of WG, often involving small arteries, veins, and capillaries, and occasionally involving the aorta. The disease usually starts with granulomatous inflammation of nasal and lung tissue and then progresses to diffuse vascular necrotizing inflammation.[3] Serum antineutrophil cytoplasmic antibody (ANCA) can be measured in these patients, and it has a high sensitivity and specificity for diagnosis. Erythrocyte sedimentation rate, C-reactive protein, and other tests may be used to monitor the progress of the disease and response to treatment.

CT and MRI examination can accurately demonstrate the location and extent of the lesions and thus provide the basis for treatment. CT can clearly show adjacent bone destruction and thickening. Early signs generally present as those of sinusitis, with even multiple biopsies of nasal mucosa showing chronic inflammation. Therefore, many cases are misdiagnosed as general inflammation. With the progression of WG, the nasal septum, lateral wall of the nasal cavity, and even the ethmoid bone and other midline structures are completely destroyed and resorbed, forming a large cavity. This finding is characteristic and plays a role in differential diagnosis. MRI has an advantage in evaluating soft tissue, and it can clearly show the relationship among the

muscles, optic nerve, and involved soft tissue. However, in early stages, MRI cannot differentiate mucosal inflammation from granulation tissue. In late stages, WG can present as a typical orbital granulomatous pseudotumor. Compared with the ocular muscles, lesions generally show slightly lower T1WI and higher T2WI signals, and enhanced scans show obvious homogeneous enhancement with indistinct boundaries. When a relatively homogeneous mass is associated with destruction of paranasal sinus or sinus bone, it should be highly suspected as a WG mass. However, some orbital WG cases are not associated with the paranasal sinus or sinus bone destruction, and therefore, orbital WG diagnosis should not be completely excluded.

5. Conclusion

In summary, WG is a serious, fatal disease. Early diagnosis and treatment are essential for controlling the progression of the disease and improving the quality of life for patients. Therefore, suspected patients who are first examined in the ophthalmology clinic with only ocular manifestations should undergo nasal biopsies, sinus imaging, and laboratory ANCA as part of a routine examination, due to frequent involvement of the nose and sinuses in WG. This would facilitate correct and timely diagnosis for such patients.

References

[1] Rodrigues CE, Callado MR, Nobre CA, et al. Wegener's granulomatosis: prevalence of the initial clinical manifestations report of six cases and review of the literature. Rev Bras Reumatol 2010;50:150–64.
[2] Tarabishy AB, Schulte M, Papaliodis GN, et al. Wegener's granulomatosis: clinical manifestations, differential diagnosis, and management of ocular and systemic disease. Surv Ophthamol 2010;55:429–44.
[3] Pakrou N, Selva D, Leibovitch I, et al. Wegener's granulomatosis: ophthalmic manifestations and management. Semin Arthritis Rheum 2006;35:284–92.