CASE REPORT
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Optic neuritis as ocular manifestation of Wegener's granulomatosis — case report

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ABSTRACT

Wegener's granulomatosis (WG) is a systemic, autoimmune, granulomatous necrotising inflammation of unknown aetiology. WG is described by the triad, which includes: rhinitis, and pulmonary and kidney dysfunction. The ocular signs are inflammatory orbital disease, nasolacrimal ducts obstruction, conjunctivitis, episcleritis, scleritis uveitis, retinal vasculitis, and optic neuritis. This article describes the case of a patient with optic neuritis associated with WG.

KEY WORDS: Wegener's granulomatosis, ophthalmic manifestations, c-ANCA

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INTRODUCTION

Wegener's granulomatosis (WG) is a systemic, autoimmune, granulomatous necrotising inflammation and vasculitis (inflammation of the small to the medium vessels). The aetiology of this disease is unknown. The first case of this disease was described in 1897 by Peter McBride who was a Scottish otolaryngologist. Heinz Karl Ernst Klinger was a medical student who added information on the anatomical pathology in 1931. Frederick Wegener was a German pathologist who defined the full picture of WG in two reports in 1936 and 1939.

The primary role of the vessels inflammation is production of granulomas with a tendency to disintegration. WG is diagnosed in any age but generally on the fourth decade of life. WG is characteristic of the triad, which includes: rhinitis, pulmonary and kidney dysfunction. It is usually located in the upper respiratory tract, ear, salivary glands, larynx, trachea, bronchi, and lung. The progressing disease comprises the other organs, e.g. kidney, liver, skin, eye, and nerves. The acute condition occurs with fever, weight loss, pain and swelling of joints, and deteriorating patient's condition. The diagnostics of WG are based on patient examination and laboratory tests, which include kidney parameters, radiogram of the lungs, and histopathological verification. The specific marker of this disease is antibodies against cytoplasmic neutrophils (c-ANCA), which increases by 95% in the active phase and only 10-20% in the inactive phase of the disease. The positive c-ANCA is a useful tool for screening. The general treatment is based on glucocorticoids and cyclophosphamide [1, 3]. The ophthalmic manifestations are present in 60% of cases.

This article describes the case of a patient with optic neuritis as ocular manifestation of WG.

CASE REPORT

A 62-year-old female reported to the General Medicine Department with severe and chronic cough and fever in 2013. X-rays showed nodules in the lungs. She was given oral and intravenous corticosteroids and antituberculosis drugs. The cANCA test was positive. The left thoracotomy was performed and granulation tissue was found in a histopathological test. A diagnosis of WG was made.

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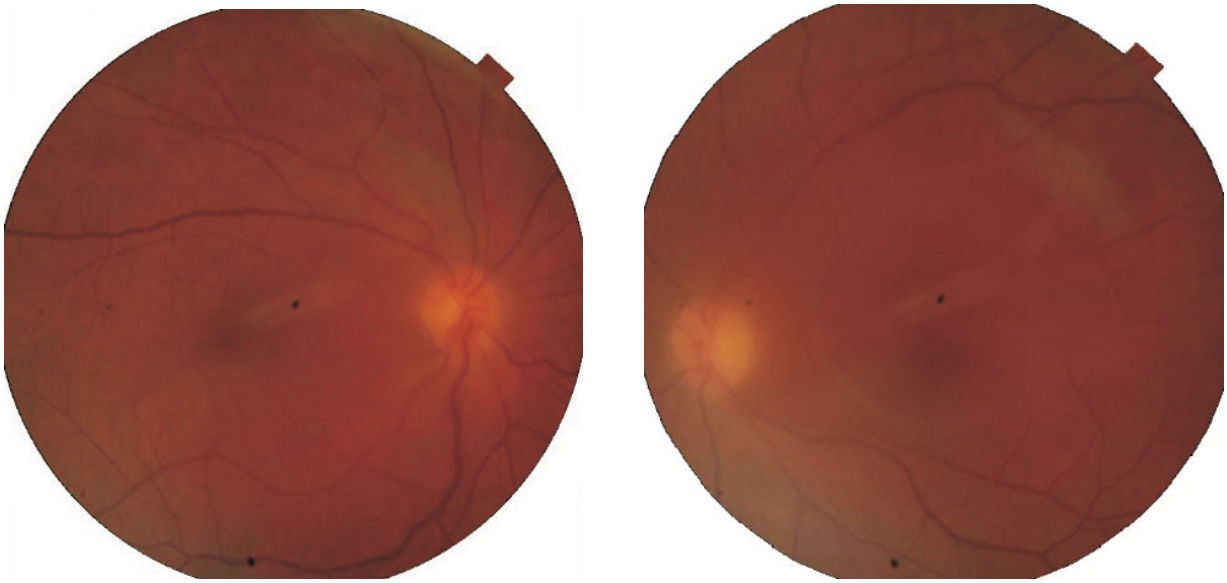

FIGURE 1. Fundus photography of both eyes (right on the right side, left on the left side)

This patient showed the systemic manifestations, e.g. impaired hearing on the left ear, small scars on the skin, nodules and cavities in lungs, kidney illness, hypertension, and weight loss (18 kg).

This woman reported to the Department of Ophthalmology in May 2014 with a pain in the orbital region and redness for more than two months in both eyes. Her visual acuity was 0.32 in the right eye and 0.8 in the left. The intraocular pressure was 20 mm Hg in both eyes. There was a stare in the right eye. Ophthalmologic examination showed normal optic discs in both eyes. Fluorescent angiography was normal. The result of brain magnetic resonance was: "contrast enhancement in the area of the temporal lobe which corresponds to the inflammatory process". The patient was given oral steroids, which brought improvement.

In March 2015 the patient reported to the Department of Ophthalmology with visual loss, diplopia, pain in the temporal area, and pain on ocular movement. Her visual acuity was 0.2 in the right eye and 0.5 in the left. The fundus examination of the both eyes was normal. Intraocular pressures were normal. The patient was given oral steroids, which brought improvement.

The patient reported to the Department of Ophthalmology in July 2016 again with visual loss. Her visual acuity was 0.1 s.c. in the right eye and she counted fingers before the left eye. Relative afferent pupillary defect (RAPD) was noticed in her left eye. Ophthalmologic examination showed pallor of the left optic nerve disc (Fig. 1). The Ocular Coherence Tomography (OCT) (Cirrus, Zeiss) of the macula was correct. There was attenuation of the peripapillary retinal nerve fibre layer (RNFL) in the left eye (Fig. 2). There was a visual field defect in the left eye — retinal nerve fibre layer in the lower hemisphere (HFA 754, Carl Zeiss) (Fig. 3).

The patient received an increased dose of steroids and she began to have better vision. The vision acuity was counted fingers from three metres in the left eye during the control examination.

DISCUSSION

The ophthalmic manifestations are found with or without systemic symptoms of WG. The ocular signs are inflammatory orbital disease, nasolacrimal ducts obstruction, conjunctivitis, episcleritis, scleritis sometimes with keratopathy, uveitis, retinal vasculitis, and optic neuritis. Visual loss occurs in 8% of patients with WG. The ocular symptoms are nonspecific, so the diagnosis of WG only with ophthalmic signs is difficult.

Orbital disease involves the nose and paranasal sinuses. The patients complain about orbital symptoms e.g. diplopia, pain on ocular movement, proptosis, periorbital swelling, red eyes, and visual loss. Eyelids are rarely involved with WG. The ocular signs are: ptosis or xanthelasma despite the patient having normal lipid metabolism, dacryoadenitis (ocular sicca syndrome), and conjunctivitis. In some patients corneal ulceration and ocular perfora-

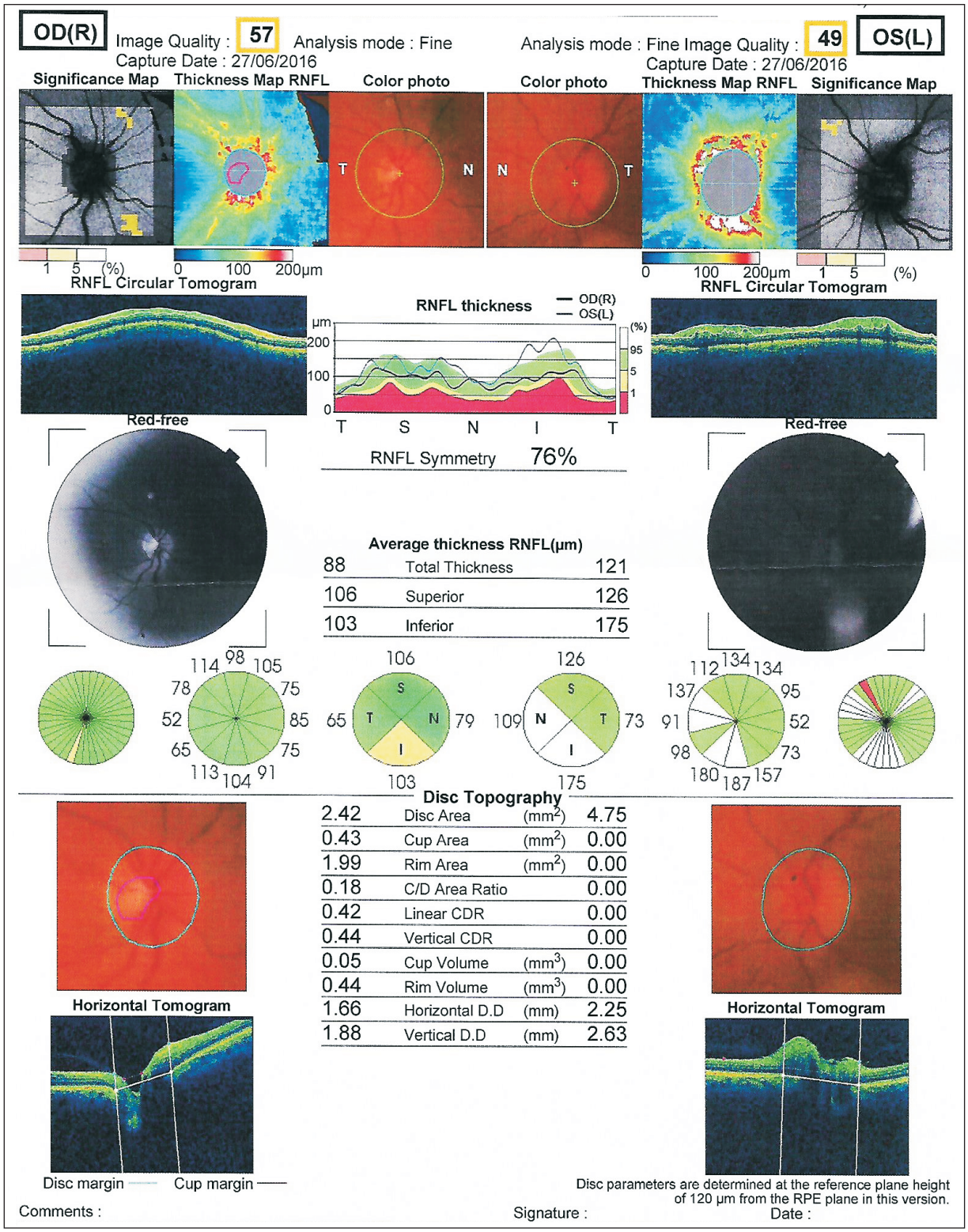

FIGURE 2. Peripapillary nerve fibre layer in optical coherence tomography of both eyes (right on the right side, left on the left side)

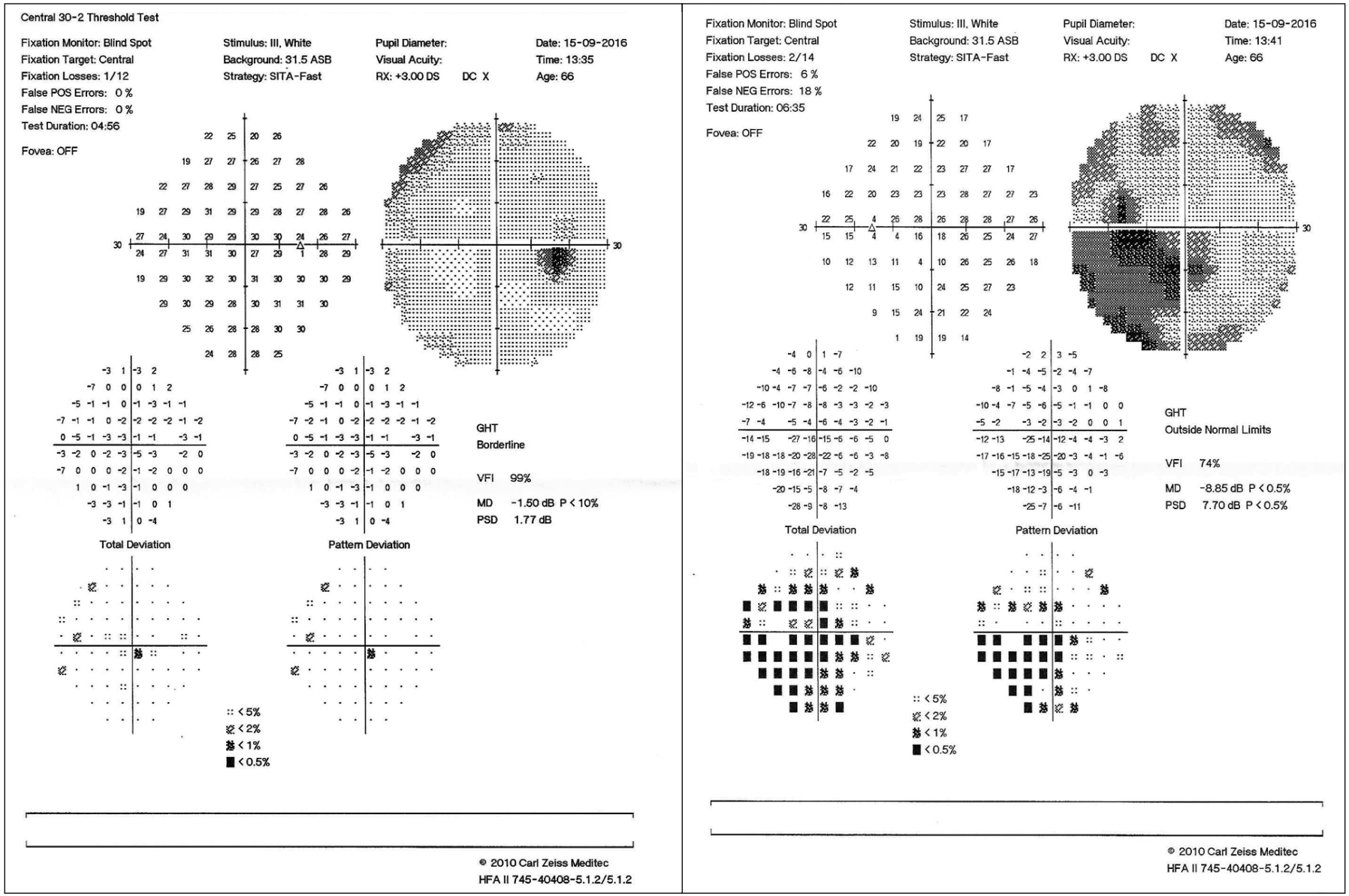

FIGURE 3. Visual field results of both eyes (right on the right side, left on the left side)

tion develop. Keratitis is a common ophthalmology sign of WG, which includes ocular pain, tearing, photophobia, and visual loss. It may result in corneal perforation. Episcleritis is presented as red eye with mild discomfort and epiphora. Scleritis is the most frequent symptom of WG (50% of patients). The patients feel pain during inflammation. The vasculitis can lead to ocular hypertension or glaucoma. Uveitis and retinitis are rare manifestations in WG. This includes chorioretinitis, macular oedema, exudative retinal detachment, and retinal necrosis (vein, artery occlusion by the tissue granulomas).

Optic neuritis is rare manifestation of WG, but among the cranial nerves this nerve is the most frequently affected. Optic nerve compression and ischaemia from vasculitic involvement are common mechanisms. Cyclophosphamide and corticosteroids are successfully used to improve visual activity. If steroids help in optic neuritis, suspicion of WG should be raised. In our patient diagnosis of optic neuritis was based on clinical signs and results of OCT of the optic nerve and visual field.

Prevention of visual loss is through knowledge about the ocular signs [1, 3].

The diagnosis and treatment of patients with WG may be a big challenge due to the possible nonspecific manifestations in the early stages of the disease. There is a need for strict monitoring of patients with WG because ophthalmological signs may be diverse.

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