Saudi Journal of Ophthalmology (2018) xxx, xxx-xxx

Case Report

# Fungal endophthalmitis in a case of granulomatosis with polyangitis

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#### **Abstract**

A 70-year-old immuno-compromised man, due to multiple comorbidities, particularly granulomatosis with polyangitis (GPA) and its related treatment, presented with generalized weakness, odynophagia and loss of taste sensation. After a complete evaluation, a diagnosis of right frontal lobe brain abscess was made. The patient then developed headache and sudden painful loss of vision in the right eye. Clinical examination revealed anterior chamber cells and flare, vitreous haze and cells, and hemorrhagic chorioretinitis with severe vasculitis in the right eye. Culture from the drained pus of the frontal brain abscess came positive for Aspergillus fumigatus. Incidental echocardiogram showed large vegetation in the mitral valve. Pars plana vitrectomy was done and a specimen was sent for culture that came positive for Aspergillus fumigatus. Although all the necessary medical and surgical interventions were timely carried out in the affected right eye, the patient's vision worsened due to retinal damage.

**Keywords**: Fungal endophthalmitis, Endogenous endophthalmitis, Aspergillus fumigatus, Granulomatosis with polyangitis, GPA, Wegener's granulomatosis

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https://doi.org/10.1016/j.sjopt.2018.01.001

#### Introduction

Granulomatosis with polyangitis (GPA), also known as Wegener's granulomatosis (WG), was first identified as a clinical entity by Friedrich Wegner in 1936.<sup>1</sup> It is an autoimmune disorder of unknown cause characterized by necrotizing granulomatous inflammatory and pauci-immune vasculitis affecting small to medium sized vessels in different systems of the body, most commonly the respiratory system.<sup>1–4</sup> Both ocular and orbital involvements can manifest in GPA patients at the time of diagnosis or later on during the course of the disease.<sup>1–3</sup> Ocular involvement occurs in 50–60% of patients with GPA, and it can affect any structure of the eye, from the eyelid and orbit to the optic nerve.<sup>2,5,6</sup>

Retinal and choroidal involvement in GPA is rare.<sup>3</sup> Recognized retinal manifestations of GPA include retinitis, chorioretinitis, macular edema, exudative retinal detachment and retinal necrosis.<sup>6</sup> Endogenous fungal endophthalmitis (EFE) is a serious ocular infection and a medical emergency that requires prompt diagnosis and medical intervention to save vision.<sup>7,8</sup> EFE is preceded by fungemia that occurs with systemic fungal infections.<sup>8</sup> The incidence of EFE in patients with a systemic fungal infection, according to different studies, varies between 2% and 45%.<sup>8</sup> The most common organism implicated in EFE is Candida albicans, followed by Aspergillus species.<sup>9</sup>

We report a case of EFE secondary to fungal endocarditis in a known case of GPA.

Received 29 June 2017; received in revised form 9 January 2018; accepted 15 January 2018; available online xxxx.

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Peer review under responsibility of Saudi Ophthalmological Society, King Saud University



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#### Case report

A 70-year-old man, presented to our emergency medicine with generalized weakness, odynophagia and loss of taste sensation. He was on antihypertensive medications for the last 10 years and underwent angioplasty for ischemic heart disease 2 years back. The patient had prostate carcinoma treated with chemotherapy and radiotherapy 2 years back and was on androgen deprivation therapy on presentation. He underwent cataract extraction with intraocular lens implantation in both eyes more than 5 years ago.

Chart review of the patient showed an ICU admission six months back due to hemoptysis. On investigations he was found to have alveolar hemorrhage, high P-ANCA and C-ANCA levels, therefore, the diagnosis of granulomatosis with polyangitis was made. He was treated with pulse methylprednisolone, plasmapheresis and cyclophosphomide. Following discharge, the patient developed steroid-induced diabetes, and, because of that, oral steroids were slowly tapered with cyclophosphomide dose increased from 25 mg to 50 mg daily. One month ago, the patient was started on rituximab and was due for the second dose.

A thorough neurological evaluation with MRI imaging revealed right frontal subcortical lesion suggestive of brain abscess [Fig. 1], thus, the patient was admitted to the neurosurgery ward. Cyclophosphamide and rituximab were held. While waiting for surgery, the patient developed sudden painful loss of vision in the right eye associated with head-

ache that warranted ophthalmic consultation. On examination, the patient's visual acuity was counting finger at 1 m in the right eye and 20\40 in the left eye. Intraocular pressure was 26 in the right eye and 22 in the left eye. On ocular examination, the right eye showed conjunctival chemosis, clear cornea and grade 4 cells in the anterior chamber with grade 3 flare. The right eye pupil was 3 mm in diameter and not reactive to light. Right eye intraocular lens was seen in place with pigment dusting present on the lens. In addition, there was grade 2 vitreous haze and grade 3 cells in the right eye. Fundus examination revealed right eye hemorrhagic chorioretinitis with severe vasculitis. The patient was started on prednisolone eye drops every hour for anterior chamber reaction and cyclopentolate eye drops every 8 h for mydriasis, and for cycloplegia to relieve pain in the right eye. Lab investigations showed elevated ESR and CRP levels, high WBC count, thrombocytopenia and low hemoglobin. After two days, the patient developed right facial palsy with left hemiparesis, and imaging showed acute small infarcts suggestive of cerebrovascular accident. Two days later, the patient underwent right frontal craniotomy and excision of the brain lesion. Culture from the drained pus was positive for Aspergillus fumigatus, and the patient was put on 250 mg of intravenous voriconazole every 12 h. Blood culture was negative. As a part of the workup, an echocardiogram was done that showed a  $1.7 \times 1.7$  cm large vegetation on the anterior mitral leaflet. After 1 week, the patient's condition of the right eye worsened with the retina showing dense

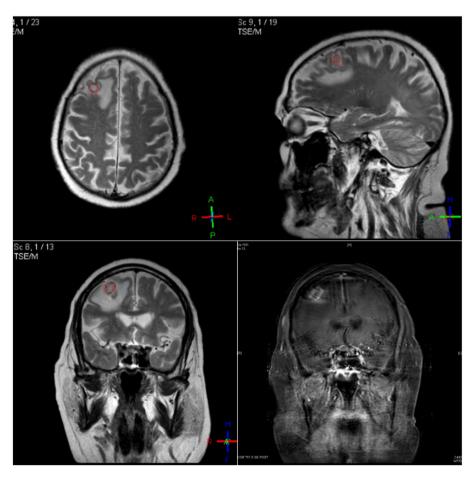


Fig. 1. Brain MRI showing right frontal lobe abscess which showed Aspergillus in aspiration of abscess.

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