Case Report

Bilateral Vision Loss in an Adult Patient with Woakes’ Syndrome: An Unprecedented Case

Kaveh Abri Aghdam1, Ali Aghajani1,2, Sara Hemmati1, Mostafa Soltan Sanjari1

1Eye Research Center, Eye Department, The Five Senses Health Institute, School of Medicine, Iran University of Medical Sciences, Tehran, Iran,
2Isfahan Eye Research Center, Department of Ophthalmology, Isfahan University of Medical Sciences, Isfahan, Iran

Abstract

Purpose: To report a rare case of Woakes’ syndrome presented with bilateral vision loss.

Methods: A 28-year-old male with a 1-year history of vision loss in the left eye was referred to the neuro-ophthalmology clinic after sudden vision loss in his right eye. A detailed review of clinical findings and the presumed pathophysiological basis of vision loss was performed.

Results: Neuroimaging revealed bilateral massive nasal polyps, sphenoid sinus mucocele formation, and optic nerve dehiscence inside the sphenoid sinus. The vision in the right eye was restored after pulse corticosteroid therapy; however, the left eye remained severely visually compromised even after nasal polypectomy and mucocele drainage.

Conclusion: Sinonasal disorders should be sought for patients with unexplained vision loss, as prompt intervention could be vision-saving in these patients.

Keywords: Sphenoid sinus mucocele, Vision loss, Woakes’ syndrome

Address for correspondence: Ali Aghajani, Department of Ophthalmology, Eye Research Center, The Five Senses Health Institute, Rassoul Akram Hospital, Niayesh Avenue, Sattarkhan Street, Tehran 1445613131, Iran.
E-mail: aliaaghajani_y@yahoo.com
Submitted: 09-Oct-2021; Revised: 30-Jan-2022; Accepted: 01-Feb-2022; Published: 26-Jul-2022

Introduction

The insult toward the orbital structures from sinonasal pathologies is a well-known entity. Everything from chronic sinusitis to malignant tumors have been reported to cause orbital structure deformity and visual impairments.1 Nasal polyps are benign intranasal and sinus lesions; however, they could cause serious complications in some patients. Orbital deformities and vision loss to various degrees have been reported in patients with sinonasal polyps. One rare presentation of sinonasal polyps is Woakes’ syndrome which is characterized by facial disfigurement and nasal pyramid deformity.2 The present case demonstrates the unprecedented bilateral vision loss in a patient with Woakes’ syndrome.

Case Report

A 28-year-old male was referred to the neuro-ophthalmology clinic with a 4-day history of painless decreased vision in the right eye. He also had a similar history of vision loss in his left eye 1 year before. Visual acuity was 20/400 in the right eye and hand motion in the left eye, and a left relative afferent pupillary defect was present. External examination revealed telecanthus and 2 mm proptosis of the left eye. The ocular movements, anterior segment examination, and intraocular pressure in both eyes were within the normal limits. No abnormality was found in the right eye on fundus examination; however, the optic disc pallor was found in the left eye. The Humphrey visual field test depicted a cecocentral scotoma in the right eye and severe visual field
loss in the left eye [Supplementary Figure 1]. With the initial impression of the right optic neuritis, the patient received 3 g of intravenous methylprednisolone over 3 days, and the vision improved dramatically to 20/25 in the right eye. However, magnetic resonance imaging (MRI) at the time of admission demonstrated the fullness of the whole nasal cavity with the expansion of ethmoidal sinuses and multiple polypoid lesions in the maxillary and sphenoid sinuses [Figure 1]. Morphology of the intraorbital optic nerves appeared normal on MRI without pathological enhancement after contrast injection; however, the posterior orbit was remodeled, and the optic nerve passage through the misshaped sphenoid sinus was visible [Figure 2]. The computed tomography scan showed the dehiscence of the bony wall of the optic canal as passing through the sphenoid sinus on the left side [Figure 1 and Supplementary Figure 2]. The ENT consultation revealed anosmia (in paper-strip test), bilateral intranasal polyps, and disfigurement of the nasal bridge. Thus, based on the clinical and imaging findings, the diagnosis of Woakes’ syndrome was made, and endoscopic polypectomy of the nasal cavity and paranasal sinuses was performed 1 week afterward. The histopathologic study of the sinus specimen revealed the inflammatory polyps with mucocele formation without any neoplasia. Unfortunately, visual acuity in the left eye showed no improvement even after the sinus surgery. Informed consent including publication of photographs in medical journals was obtained from the patient.

**DISCUSSION**

Optic nerve insult and visual compromise are well-known complications of sinonasal pathologies such as tumors and infections. Mucocele formation and inflammation, especially in posterior ethmoidal and sphenoid sinuses could cause optic nerve damage either by direct compression or inflammation. The formation of mucocele and mucopyocele is a consequence of sinus drainage obstruction by pathologies such as nasal polyps, especially when bilateral. Woakes’ syndrome is characterized by severe recurrent nasal polyps with consecutive destruction of the nasal pyramid, leading to the broadening of the nose, hypertelorism, and perioseal resorption with bony fibrosis. Most patients with this disorder are young children. This syndrome in adults is a rare condition, and its ocular complications are even more unusual. Unlike silent sinus syndrome that polyps lead to the inward displacement of the sinus wall and consequent enophthalmic appearance, massive polyps in this syndrome cause facial disfigurement and orbital deformities such as hypertelorism and frank proptosis (as in the index case) through their pressure effect and continuous inflammation; however, to the best of our knowledge, there has been no previous report of vision loss in these patients. In our case, the bony deformity was expanded toward the orbital apices and led to optic canal

![Figure 1: Facial, orbital, and paranasal sinus study.](image)

![Figure 2: Optic nerve study.](image)
deformity and passage of the optic nerve through the deformed sphenoid sinus. Optic nerve protrusion into the sphenoid sinus predisposes it to insults such as compression, ischemia, and toxic effect of intrasinus materials that have been described in patients with pneumosinus dilatans syndrome and sphenoid sinus mucocele.5-7 At least, some inflammatory processes have been involved in the pathogenesis of vision loss in this patient, which responded well to corticosteroid therapy and led to vision restoration in the right eye. The pattern of visual field loss in the right eye (ceccentrical scotoma) was consistent with inflammatory optic neuropathy that was most probably due to exposure to the toxic materials inside the sphenoid sinus mucocele. Polyps in Woakes’ syndrome are inherently resistant to corticosteroid therapy due to their fibrotic nature,2 thus, reduction of the direct (as in Onodi cells) or indirect (negative pressure inside the obstructed sphenoid sinus) compression effect of inflamed polyps is a less likely explanation for treatment success in this patient. On the other hand, we cannot assure whether the dehiscence of the bony structure of the left optic canal was due to the destructive effect of sinonasal polyps or was a normal anatomical variation; in either of which, prolonged direct contact of the optic nerve with the intrasinus (mucopyocele) materials seems to be a major cause of axonal damage and irreversible vision loss in the left eye.

In conclusion, a clinician must be aware of the devastating effect of sinonasal pathologies on the orbital structures and optic nerve. Looking for possible sinonasal disorder clues in clinical examination, such as telecanthus and proptosis, and paraclinical studies (posterior ethmoidal and sphenoid sinus abnormalities) is crucial when facing a patient with unexplained vision loss, as early diagnosis and intervention in these patients could be vision-saving before the optic atrophy ensues.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES
1. Kim YH, Kim J, Kang MG, Lee DH, Chin HS, Jang TY, et al. Optic nerve changes in chronic sinusitis patients: Correlation with disease severity and relevant sinus location. PLoS One 2018;13:e0199875.
2. Caversaccio M, Baumann A, Helbling A. Woakes’ syndrome and albinism. Auris Nasus Larynx 2007;34:245-8.
3. Schoenenberger U, Tasman AJ. Adult-onset woakes’ syndrome: Report of a rare case. Case Rep Otolaryngol 2015;2015:857675.
4. Cruz AA, Chahud F, Akaishi PM, Enz TJ. Woakes syndrome: A rare cause of proptosis due to sinonasal polyposis. Ophthalmic Plast Reconstr Surg 2019;35:e102-4.
5. Li Z, Zhang Y, Liao Y, Zeng R, Zeng P, Lan Y. Comparison of efficacy between anti-vascular endothelial growth factor (VEGF) and laser treatment in Type-1 and threshold retinopathy of prematurity (ROP). BMC Ophthalmol 2018;18:19.
6. Voglewede AT, Justice JM. Bilateral pneumosinus dilatans of the sphenoid sinuses causing visual loss. Int J Pediatr Otorhinolaryngol Extra 2015;10:79-83.
7. Aghdam KA, Aghajani A, Sanjari MS. Bilateral visual loss caused by pneumosinus dilatans: Idiopathic cases are not always reversible. J Curr Ophthalmol 2021;33:197-200.
Supplementary Figure 1: Visual field report

Supplementary Figure 2: (a-i) Represents different orbital computed tomography axial scans of the patient