Case Report

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

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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

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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.

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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

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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.

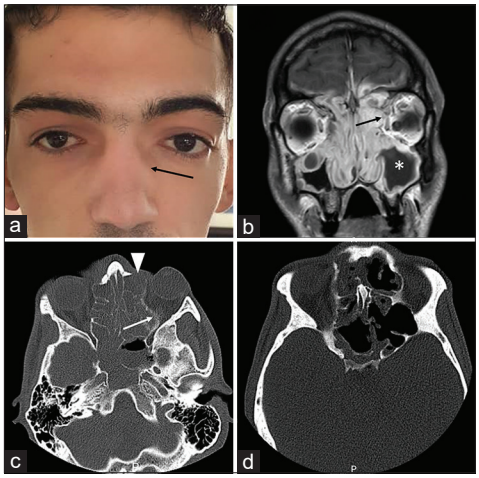

Figure 1: Facial, orbital, and paranasal sinus study. (a) The face photograph of the patient shows telecanthus and deformity of the nasal bridge (black arrow). (b) Coronal noncontrast T1-weighted magnetic resonance image of the orbits and paranasal sinuses reveals the remodeling of the medial orbital walls with an expansion of the ethmoid sinus (white arrow); note the hyperintense lesions that occupy the nasal cavity and the ethmoidal sinuses with expansion to the frontal sinuses. Maxillary sinus fullness with polypoidal lesions and mucocele formation is also notable (black asterisk). (c) Axial noncontrast computed tomography (CT) scan of the orbits depicts the destruction of the nasal pyramid and expansion of the ethmoidal sinuses toward the left orbit. (d) Axial noncontrast CT scan of the orbits and paranasal sinuses after endoscopic polypectomy shows the intact bony wall of the right optic canal through the sphenoid sinus

Discussion

Optic nerve insult and visual compromise are well-known complications of sinonasal pathologies such as tumors and infections.[1] 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 periosteal resorption with bony fibrosis.[2] Most patients with this disorder are young children. This syndrome in adults is a rare condition, and its ocular complications are even more unusual.[3] 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;[4] 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

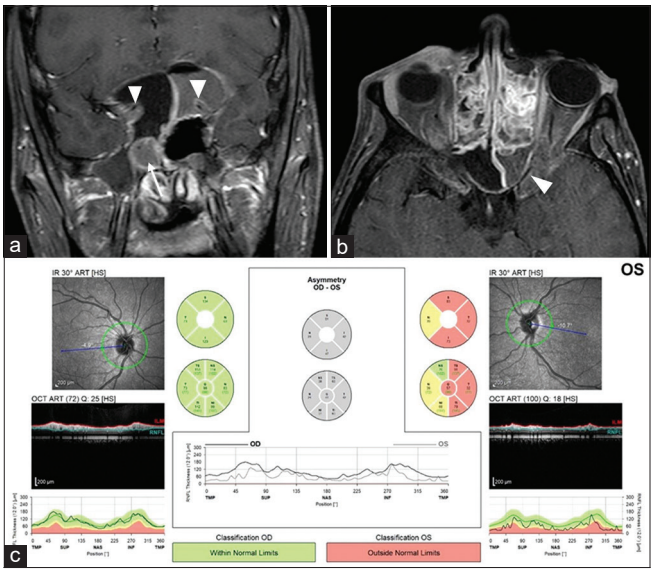

Figure 2: Optic nerve study. (a) Coronal contrast-enhanced T1-weighted magnetic resonance (MR) image shows the expansion of the sphenoid sinus and displacement of intracranial structures. The optic nerves are visible as they are passing through the sphenoid sinus (arrowheads). Polypoidal lesions are visible in both right (white arrow) and left sphenoid sinuses (surrounding the optic nerve). (b) Axial contrast-enhanced T1-weighted MR image demonstrates the fullness of the sphenoid sinus on the left side with direct contact of the optic nerve with mucocele material. Note the expansion of ethmoidal sinuses that are packed with contrast-enhanced lesions that are consistent with the diagnosis of intranasal polyps. (c) An optical coherence tomography scan of the peripapillary nerve fiber layer shows a significant loss of retinal nerve fiber layer in the left eye

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 (cecocentral 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.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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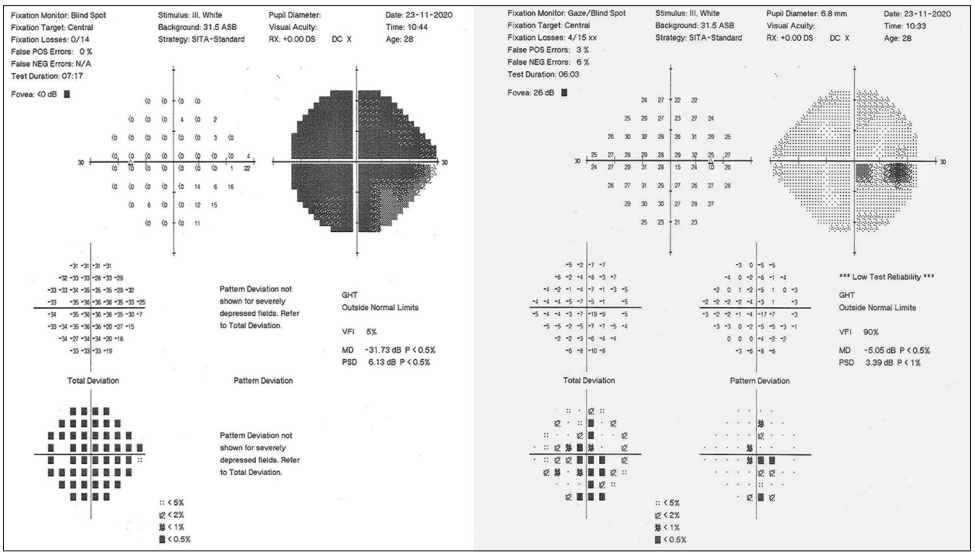
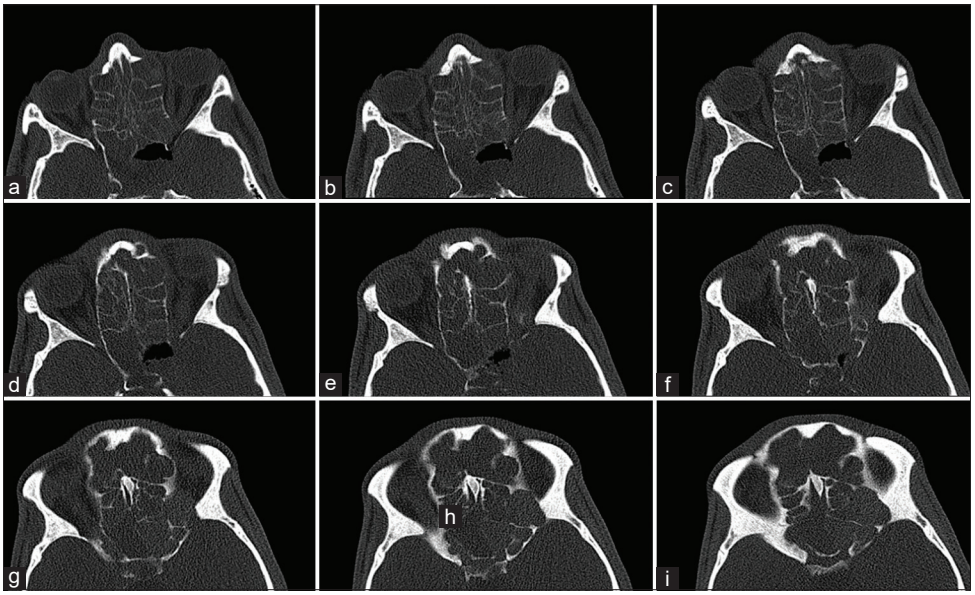


Supplementary Figure 1: Visual field report



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