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Loss of Vision Outcome for Allergic Fungal Sinusitis: Case Report and Literature Review

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Abstract

Visual loss is a rare manifestation of allergic fungal rhinosinusitis (AFRS). We report a case of an adult male who was diagnosed with AFRS and who presented during the COVID-19 pandemic lockdown with sudden-onset complete vision loss and a lack of recovery after surgical and medical management. We reviewed the literature on reported cases of AFRS complicated by visual loss to identify factors associated with visual outcomes. We found 50 patients who were diagnosed with acute visual loss due to AFRS, with an average age of 28 ± 14 years. Complete and partial recovery after surgical intervention were reported in 17 and 10 cases, respectively. However, the absence of vision improvement was reported in 14 of the cases. Early diagnosis and prompt intervention can return vision back to normal. However, delayed presentation, complete loss of vision, and acute onset of visual loss are associated with worse outcomes.

Keywords: allergic fungal sinusitis, orbital complications, blindness, loss of vision

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Introduction

Sinonasal pathologies associated with fungal presence were classified in the recent European Position Paper on Rhinosinusitis and Nasal Polyps (EPOS) 2020 into invasive fungal rhinosinusitis, fungal ball, and allergic fungal rhinosinusitis (AFRS). The EPOS 2020 also suggested that the corresponding immune responses are immune suppression, immune competence, and immune hypersensitivity, respectively.[1]

AFRS is considered the most common type of non-invasive form of fungal rhinosinusitis. Although the presence of fungus plays a fundamental role in the pathophysiology of AFRS, it does not behave as an infectious agent. AFRS is believed to be a complex immune reaction involving humoral and cellular responses as part of type 1 and type 3 hypersensitivity reaction characterized by the presence of eosinophilic mucin and Charcot–Leyden crystals with non-invasive fungal hyphae within the nasal cavity.[1–3]

Inflammation in AFRS produces a thick nasal discharge that may obstruct paranasal sinus drainage, and mucocele-like expansion may occur, which can affect adjacent structures, including the visual pathway. This process occurs due to compression of the neural structure or its blood supply rather than invasion of the dura.[4] Even so, AFRS has a favorable outcome if managed promptly with surgical removal of fungal debris and corticosteroid therapy.[1]

Unlike AFRS, invasive fungal rhinosinusitis is characterized by the presence of fungal hyphae within the sinonasal mucosal tissue, demonstrating a classic invasion pattern that results in thrombosis and dural or bone invasion. This process, which happens almost exclusively in immunocompromised patients, requires aggressive surgical removal of the infected tissues and the application of medical antifungal agents.[1] Even with such aggressive management, patients usually experience catastrophic outcomes, such as blindness, stroke, and death.[4]

In this paper, we report a case that presented with sudden visual loss as a complication of AFRS. We also conducted a review of the literature to identify the outcomes of such cases after surgical and medical management.

Case

Clinical Presentation and Diagnosis

During the lookdowns for the coronavirus disease of 2019 (COVID-19), a 46-year-old man presented to the emergency department complaining of sudden-onset left-sided visual loss after waking up from sleep, with associated drooping of the upper lid on the same side. The patient had been diagnosed with AFRS according to Bent and Kuhn criteria after functional endoscopic sinus surgery (FESS) seven years prior.[5] Three days before the presentation, he had started to feel a pressure-like sensation in his left eye, nasal congestion, anosmia, and facial pressure. He also experienced left facial numbness over the cheek and mouth. The patient had a history of bronchial asthma; otherwise, he was immune competent.

Upon examination, the nasal cavity revealed polyps extending bilaterally to the floor of the nose. Visual acuity was 20/20 in the right eye and no light perception in the left. There was full mobility of the external ocular muscles, with mild discomfort on the affected side. In addition, there was an afferent pupillary defect in the left eye. The reset of the ophthalmological assessment and other cranial nerves were unremarkable.

The patient was started on prednisolone 60 mg daily, intranasal rinses, and intravenous amoxicillin/clavulanic acid empirically. Computed tomography (CT) of the paranasal sinuses showed that all were opacified by a hyperdense lesion, with the left sphenoid sinus affected by bone remodeling and destruction of its boundaries, which resulted in the extension of the lesion to the brain parenchyma, the posterior part of the left rectus gyrus, the left cavernous sinus, the left internal carotid artery, and the left part of the sellar and suprasellar regions (Figure 1). Magnetic resonance imaging (MRI) of the brain and paranasal sinuses reported erosion of the sphenoid sinus wall, with further extension into the anterior cranial fossa, causing dural thickening and enhancement (Figure 1). Minimal extension into the left optic canal and superior orbital fissure was noted, with close proximity to the supraclinoid left internal carotid artery.

Surgery

The patient was taken for revision FESS, which showed nasal polyps reaching the nasal floor, adhesions all over the nasal cavity with scar bands over the left maxillary ostium, thick allergic mucin in the maxillary, and ethmoid and sphenoid sinuses bilaterally. In the left sphenoid sinus, there was dehiscence of the left internal carotid artery and exposure of the dura. Excised tissues and mucins were sent for histopathological and microbiological studies.

Post-Operative Management and Outcomes

Antibiotics and prednisolone were resumed post-operatively, and nasal irrigation was ceased. After the surgery, the patient's headache, sense of smell, nasal obstruction, and left cheek numbness improved. However, his vision was the same as upon presentation. MRI venography performed after four days after surgery showed that the venous sinuses were well opacified, with no filling defects to suggest thrombosis. An MRI angiogram showed persistence of the minimal extension into the left optic canal and superior orbital fissure, likely due to residual disease.

The patient was then discharged with oral voriconazole and prednisolone. Histopathological examination revealed an absence of invasive patterns in the tissue and fungal cultures were negative. The patient's vision had not improved for 2 years from initial presentation, with a continuing absence of light perception.

Discussion

Orbital involvement is a rare manifestation in AFRS cases, the prevalence of which ranges between 18.3% and 34% of all cases, with severity ranging from proptosis and diplopia to vision impairment.[6,7] In the literature, two mechanisms have been mentioned as responsible for the effect of AFRS on vision. These are bony expansion and remolding of the sinuses, which result in direct compression over the optic nerve or optic chiasma and irritation and inflammation of the optic nerve triggered by a hyperimmune response to fungal antigens.[8,9]

Compression over the optic nerve may cause dysfunction in different mechanisms, including conduction blockage, ischemic injury, demyelination, and retrograde or anterograde degeneration. Overlapping between these mechanisms may occur, and activated apoptotic pathways may follow, resulting in permanent nerve damage.[10] Gupta et al identified a hyperimmune response to a fungal antigen that affects the

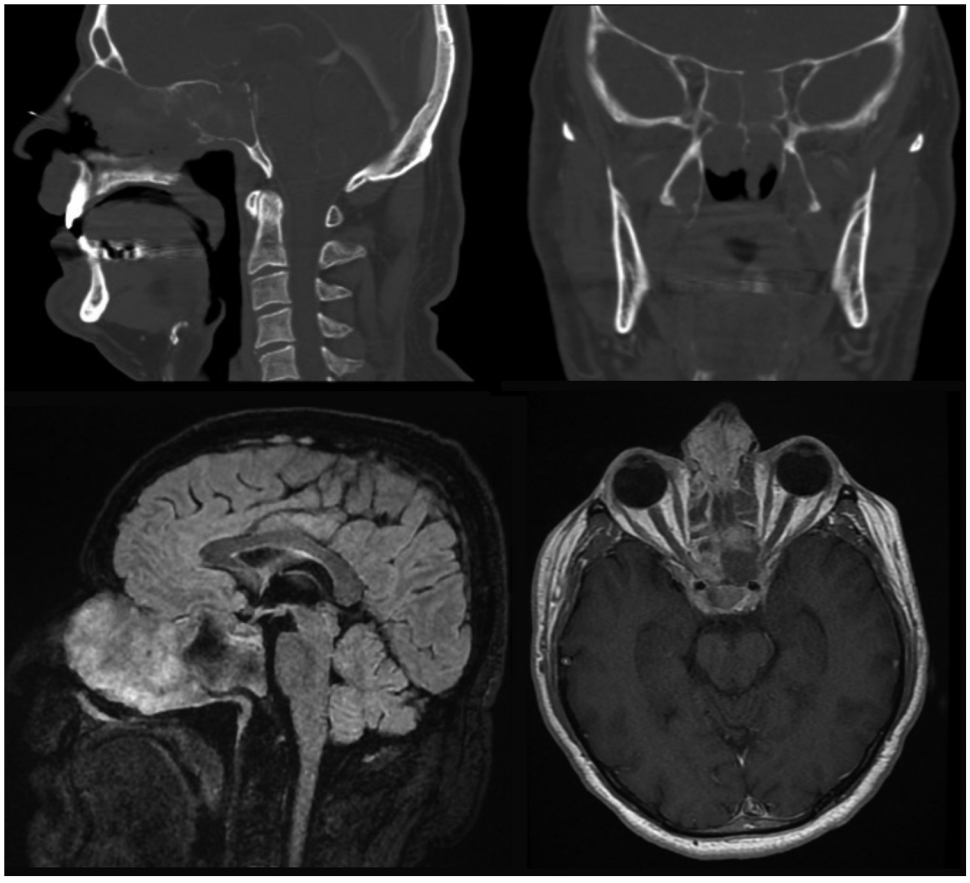

Figure 1. Computed tomography (upper) showing left sphenoid sinus affected by bone remodeling and destruction of its boundaries, which resulted in possible extension of the brain. Magnetic resonance imaging (lower) shows erosion of the sphenoid sinus wall with further extension into the anterior cranial fossa, causing dural thickening and enhancement with intact optic nerve.

optic nerve in a patient who developed visual loss with no sphenoid wall erosion. The optic nerve was found to be edematous after decompression. The hyper-immune response theory supports the improvement of the patient after surgical debulking, which decreases the load of the fungal antigen, and the use of corticosteroids, which decreases the immune reaction.[8] Generally, visual complications in AFRS have had favorable outcomes. Visual improvement in some of the reported cases was observed upon initiation of medical management and before definitive surgical decompression.[11,12]

In the literature, we found that the most important factor associated with favorable visual outcomes is the duration of the decrease in vision (Table 1). Delayed intervention decreases the probability of vision improvement (Table 2). Alaraj et al. suggested that there are better outcomes for patients who are managed within the first 30 days of visual loss onset compared to those with late presentation.[7] This statement is consistent with our findings as a significant difference (p-value = 0.038) was observed between patients who had full, partial and no visual recovery when we compared the mean number of days between experiencing the visual impairment and before intervention (Table 2). In addition, complete recovery was noted to be more frequent in patients with sudden visual loss, and partial or absent recovery was associated with progressive courses.[11] Moreover, complete visual loss was suggested to pose an unfavorable risk to the possibility of reversing vision loss.[19]

Orbital complications in AFRS were found to occur commonly in otherwise healthy patients. The cases were reported in different races and ethnicities, without any specific demographical patterns. One study suggested that neuro-ophthalmic complications could be more common in humid environments.[20]

Table 1. Previous Mentioned Cases of Vision Loss due to AFRS.

Study	Case number	Age & gender	Visual loss duration (days)	Pre-VA	Post-VA	Visual recovery†
Dunlop 1988[13]	1	69, F	5	6/24	6/9	Full
Brown 1994[14]	2	30, F	90	6/60 (R) 6/36 (L)	NR	NR
Marple 1999[6]	1	30, M	90	6/18	6/6	Full
	2	43, M	Few days	6/15	6/6	Full
	3	16, M	21	6/7.5	6/7.5	No
Attallah 1999[15]	1	15, M	180	No LP	6/6	Full
Graham 2005[16]	1	65, F	NR	LP	6/7.5	Full
Herrmann 2006[17]	1	11, F	42	6/12 (R) No LP (L)	6/6 (R) No LP (L)	Full No
Gupta2007[8]	1	NR	2	No LP (R)	Complete recovery	Full
	2		4	No LP (L)		Full
	3		9	No LP (R)		Full
	4		15	No LP (L)	Partial recovery	Partial
Aakalu 2009[18]	1	30, M	4	HM (R)	6/6-1	Full
Thakar 2011[19]	1	22, F	20 (R) 360 (L)	HM (R) No LP (L)	6/9 (R) No LP (L)	Full No
	2	30, M	30	6/18 (R)	6/9	Full
	3	18, M	180	6/12 (R) 6/60 (L)	6/12 (R) 6/60 (L)	No No
	4	17, M	90	6/24 (R)	6/24 (R)	No
	5	21, F	90	6/60 (L)	6/60 (L)	No
	6	22, M	30	6/18 (L)	6/12 (L)	Partial
	7	24, M	7	6/36 (L)	6/12 (L)	Partial
	8	19, F	20 (R) 10 (L)	1/60 (R) 3/60 (L)	6/12 (R) 6/9 (L)	Partial Full
	9	50, F	7	HM (L)	6/9 (L)	Full
	10	30, M	4	No LP (L)	No LP (L)	No

(Continued)

Table 1. Continued.

Study	Case number	Age & gender	Visual loss duration (days)	Pre-VA	Post-VA	Visual recovery†
Al-Radadi 2011[9]	1	35, F	60	6/6*	Complete recovery	Full
Sridhar 2012[20]	1	25, F	300	5/200 (L)	NR	NR
	3	64, F	90	No LP (R)	No LP (R)	No
Vashishth 2014[11]	1	20, M	150	6/24 (L)	NR	NR
	2	22, F	2	6/24 (L)		
	6	22, M	150	No LP (L)		
	10	30, F	240	No LP (L)		
Tong 2015[21]	1	32, M	Several months	FC (R)	6/36 (R)	Partial
Bobart 2017[22]	1	30, M	365	No LP (R) 20/80 (L)	Improved	Partial
Alaraj 2018[7]	1	NR	NR	NR	2 had permanent visual loss	NR
	2					NR
	3					No
	4					No
Cyriac 2019[12]	1	22,M	3	3/60 (R)	6/10 (R)	Full
Walter 2020[4]	1	23, F	NR	FC (L)	FC (L)	No
Singh 2021[23]	1	22, F	120 (R)	No LP (R)	No LP (R)	No
			20 (L)	HM (L)	6/20 (L)	Partial
	2	20, F	180 (R)	HM (R)	HM (R)	No
			30 (L)	FC (L)	6/9 (L)	Full
	3	16, M	45 (R)	HM (R)	2/60 (R)	Partial
	4	57, M	5 (L)	LP (L)	6/18 (L)	Partial

VA: Visual acuity; *NR:* Not reported; *LP:* Light perception; *HM:* Hand-motion; *FC:* Finger counting; *(R):* Right eye; *(L):* Left eye.
† Full visual recovery was considered if improvement was 6/9 or more.
* Visual field loss: Right temporal hemianiopia.

In addition, at the time of COVID-19, logistics delays in delivering diagnoses and management caused the progression of AFRS to give rise to more complications at the time of presentation.[23]

Management of AFRS consists mainly of decompression of the sinus and relief of the expansion pressure over the orbit, brain, and/or optic nerve. This step is achieved by corticosteroids and surgical removal of fungal elements. Then, sinus drainage restoration must be achieved to allow for topical medication delivery for better disease control. Afterward, adjuvant medical management includes oral and topical steroids, which help to reduce the recurrence of AFRS.[21] All reported cases were managed with surgical intervention, except for two patients. The first patient refused the surgery and she received intravenous antifungal medication

Table 2. Characteristics of the Cases Based on Vision Outcome.

	Complete recovery	Partial recovery	No recovery	*p*-value†	No report for vision outcome
Number of cases	17 (34%)	10 (20%)	14 (28%)	-	9 (18%)
Average age	32.9 ± 17.9	28.1 ± 12.1	23.5 ± 13.6	0.827	25.6 ± 4.4
Gender					
Male	43%	78%	42%	0.239	29%
Female	57%	22%	58%		71%
Average days of visual loss	33.1 days	69.7 days	123.4 days	0.038*	-

† *p*-values < 0.05 were considered statistically significant.
† Chi square and One-way ANOVA tests were used.
* Statistically significant.

(micafungin and voriconazole) for more than 10 months. Although the headache and extraocular movement improved, her affected eye still had no light perception.[20] The second patient did not undergo surgical intervention due to her general health condition. She received steroid therapy and ampicillin-sulbactam and her vision improved, which supported the immune response hypothesis.[16] As the recurrence rate in AFRS is relatively high, continued surveillance is recommended to detect the disease and its complications early, hence treating it promptly to avoid further complications.[22]

Conclusion

Vision loss in AFRS is a rare manifestation that can occur in otherwise healthy patients. Early diagnosis and prompt intervention can return vision to normal. However, delayed presentation, complete loss of vision, and acute onset of visual loss are associated with worse outcomes.

Declarations

Ethical Approval

Ethical approval is not applicable because this article does not contain any studies with human or animal subjects.

Author Contribution(s)

Ahmed Alhussien: Formal analysis; Investigation; Writing – original draft.

Abdulrahman Alghulikah: Conceptualization; Investigation; Methodology; Writing – review & editing.

Hussain Albaharna: Conceptualization; Data curation; Investigation; Supervision; Writing – review & editing.

Abdulrahman Alserhani: Formal analysis; Formal analysis; Writing – review & editing.

Saud Alromaih: Formal analysis; Methodology; Writing – review & editing.

Mohammad Aloulah: Conceptualization; Supervision; Writing – review & editing.

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Competing interests

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Availability of Data and Materials

The authors confirm that the data supporting the findings of this study are available within the article.

Statement of Informed Consent

Written informed consent to publish the information and radiology imaging in this article was obtained from the patient.

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