

Visual loss in the setting of allergic fungal sinusitis: pathophysiology and outcome

A K GUPTA, S BANSAL, A GUPTA, N MATHUR

Abstract

Objective: To hypothesise the probable pathophysiological mechanism responsible for visual loss in allergic fungal sinusitis, other than direct compression.

Design: Retrospective, non-randomised case series. Out of 274 cases of allergic fungal sinusitis, four cases with sudden visual loss were enrolled into the study. The fourth case had visual loss on the contralateral side to bony erosion of the lateral wall of the sphenoid sinus.

Interventions: All four cases were evaluated with fungal smear, immunoglobulin (Ig) E titres, visual evoked potentials, non-contrast computed tomography and magnetic resonance imaging of the paranasal sinuses, and fundus examination. They then underwent endoscopic sinus debridement followed by intravenous methylprednisolone.

Outcome measures: Improvement in vision.

Results: All four cases experienced an improvement in vision: full recovery in three cases and partial improvement in one case.

Conclusion: In view of the operative, radiological and laboratory findings for case four, with the suggestion of a hyperimmune response to fungal antigens (in the form of raised IgE titre and positive fungal serology), we suggest that a local immunological reaction to fungal antigens might be responsible for the observed visual loss in cases of allergic fungal sinusitis, in addition to mechanical compression of the optic nerve.

Key words: Mycoses; Sinusitis; Hypersensitivity; Blindness

Introduction

Fungal infection of the paranasal sinuses is classified into five variants on the basis of histopathology. Allergic fungal sinusitis is the variant most commonly encountered by the rhinologists, with nasal obstruction being the most common presentation. However, this condition can present with ocular manifestations varying from proptosis and diplopia to impaired vision and, rarely, sudden visual loss.¹ Ophthalmic findings are said to occur in as many as 18.3 per cent of cases, probably due to the close proximity of the optic nerve to the paranasal sinuses. However, presentation as sudden visual loss is unusual.² There are isolated case reports of sudden loss of vision due to allergic fungal sinusitis, and the mechanism for this is not well defined.^{3–5}

We here report a series of four cases of allergic fungal sinusitis which presented with sudden loss of vision. We also describe a probable pathophysiological mechanism for visual loss due to the allergic variant of fungal sinusitis.

Patients and methods

This was a retrospective study conducted in the department of otolaryngology, head and neck surgery at the Postgraduate Institute of Medical Education & Research, Chandigarh. We analysed records of all the cases admitted with a final diagnosis of allergic fungal sinusitis, from 1 January 2002 to 30 November 2005. We retrieved records for 274 cases of allergic fungal sinusitis, of which four cases presented with sudden visual loss. The records were analysed in terms of clinical profile, investigative profile (i.e. fundus findings, visual evoked potentials, computed tomography (CT), magnetic resonance imaging (MRI) and immunological analysis), operative findings and post-operative results in terms of visual improvement. All these patients underwent endoscopic clearance of disease, followed by post-operative steroids (i.e. a loading dose of intravenous methylprednisolone 30 mg/kg, followed by a dose of 15 mg/kg for a total of 12 doses every six hours). This was followed by oral steroids (1 mg/kg/day in

From the Department of Otolaryngology, Head and Neck Surgery, Postgraduate Institute of Medical Education and Research, Chandigarh, India.

Accepted for publication: 14 November 2006.

TABLE I
CLINICAL AND INVESTIGATIVE PROFILE OF PATIENTS

	Case 1	Case 2	Case 3	Case 4
<i>Symptoms</i>				
Nasal obstruction	+	+	+	+
Sneezing	+	+	+	+
Rhinorrhoea	+	+	+	+
Postnasal drip	+	+	+	+
Proptosis	-	-	-	+
Headache	-	-	-	+
Visual loss	+	+	+	+
Duration of visual loss (days)	2	4	9	15
<i>Signs</i>				
Nasal polypi	+	+	+	+
Visual acuity	PL -ve (R)	PL -ve (L)	PL -ve (R)	PL -ve (L)
Fundoscopy	Papilloedema (R)	Papilloedema (L)	Papilloedema (R)	Papilloedema (L)
<i>Investigations</i>				
VEP	↓ amplitude & ↑ latency on affected side	↓ amplitude & ↑ latency on affected side	↓ amplitude & ↑ latency on affected side	↓ amplitude & ↑ latency on affected side
IgE levels	Increased	Increased	Increased	Increased
Fungal smear	+	+	+	+
Fungal culture	+	+	+	+

PL = Perception of light; R = right; L = left; VEP = visual evoked potential; Ig = immunoglobulin

a tapering dose over a period of two weeks), then a steroidal nasal spray for six months.

Results

We analysed records for four cases with sudden visual loss secondary to allergic fungal sinusitis, out of a total of 274 cases of allergic fungal sinusitis over a period of 46 months. Patients comprised two men and two women (i.e. male to female ration of 1:1). Patients' ages varied from 21 to 29 years, with a mean of 25 years. The symptoms, signs and investigative profile are detailed in Table I.

The most common symptoms were unilateral nasal obstruction in addition to unilateral visual loss. The most common sign was the presence of polypi in the nasal cavity. The duration of the visual loss varied from 2 to 15 days. The right side was affected in two cases and the left side in the other two cases.

Immunoglobulin (Ig) E levels were raised in all cases. The fungal smear was positive in all cases, and culture grew *Aspergillus flavus* in all cases. Fungal serology was positive for *Aspergillus flavus* in all cases. Visual evoked potentials revealed an increase in the absolute latency and a decrease in the amplitude of the p wave in all cases (Figure 1). Non-contrast CT (Figures 2 and 3) and MRI revealed findings typical of allergic fungal sinusitis (Figures 4 and 5). Fundoscopy revealed papilloedema in all cases.

At surgery, in three cases, there was a small dehiscence of the lateral wall of the sphenoid sinus in the region of the optic canal, and the nerve was found to be oedematous and was hence decompressed. However, in the fourth case, the canal over the optic nerve was dehiscent on the contralateral side to the visual loss; during decompression of the nerve on the affected side, the nerve was found to be oedematous.

Histopathological analysis revealed charcot leyden crystals and allergic mucin, in addition to the fungal hyphae present extra-mucosally.

Post-operatively, vision recovered fully in three cases and partially in case four. None of the cases had any complications related to the surgical procedure. After a minimum follow up of 10 months, all the cases were free of recurrence.

Discussion

Allergic fungal sinusitis is a common but still controversial topic in modern rhinology. Its most common mode of presentation is unilateral nasal obstruction with the presence of polypi. The ophthalmic findings are uniform in the paediatric population, in which, due to the growth and expansion of the craniofacial skeleton, the condition tends to extend intra-orbitally and intra-cranially. We have previously reported ophthalmic findings and compared paediatric and adult cases of allergic fungal sinusitis.⁶

There are isolated case reports of allergic fungal sinusitis presenting as sudden visual loss.³⁻⁵ Out of 274 cases of allergic fungal sinusitis, we found four (1.46 per cent) which presented as sudden visual loss and were admitted for surgery. There was no sex or side predilection. Our series showed the condition to affect young adults, whereas cases reported in the literature have shown the elderly to be more commonly affected.^{3,4}

Immunoglobulin E levels were raised in all our cases, suggesting an allergic response to the fungal antigens. This is in agreement with other reports.³ Fungal serology was positive in all our cases; however, one previously reported case in the literature had negative fungal serology, probably secondary to coexistent oral steroid administration.⁴ The results of visual evoked potentials were in accordance with the literature.⁴ Fundoscopy revealed

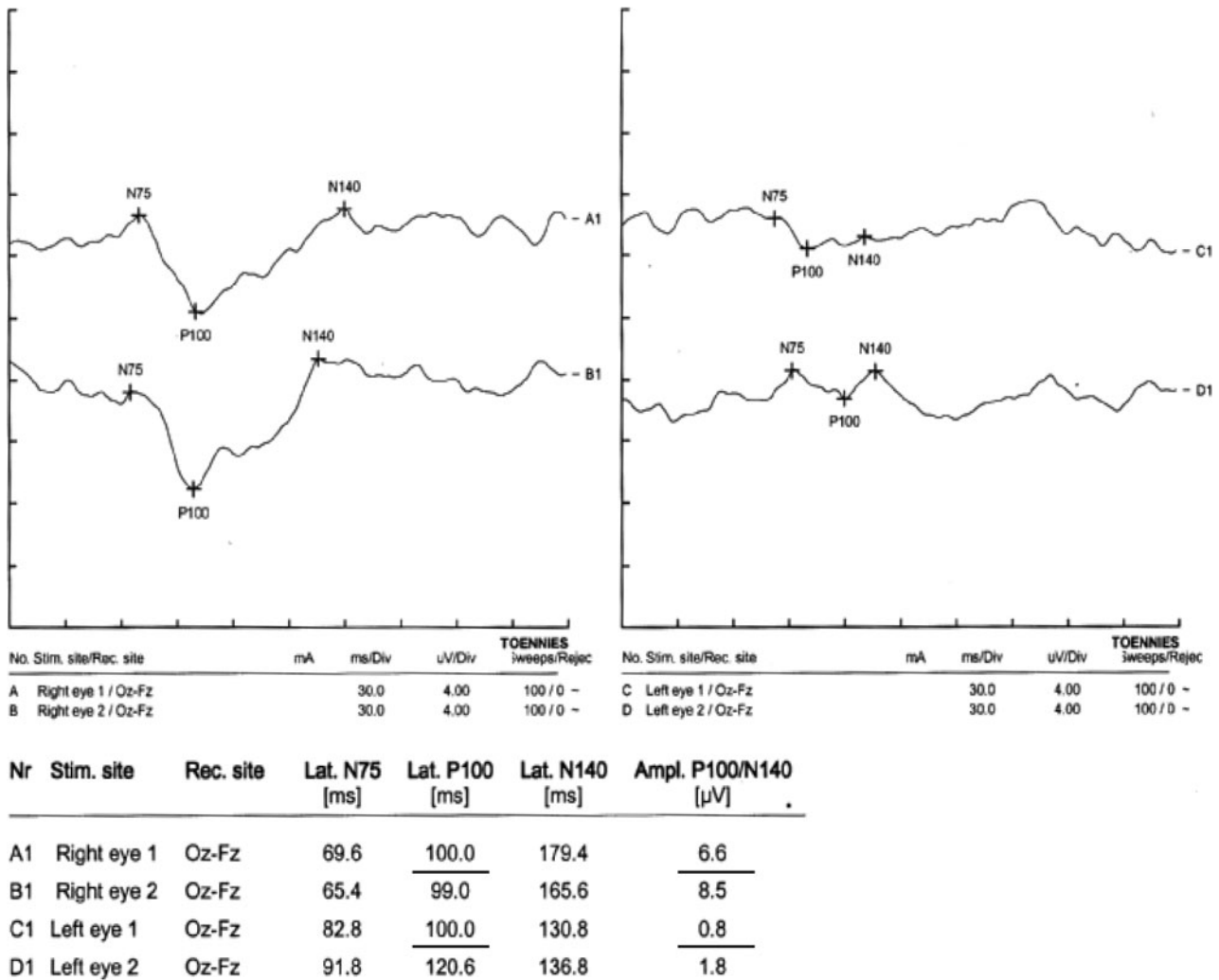


FIG. 1

Visual evoked potentials for case four, showing decreased amplitude and irregular 'p' wave formation in the left eye. Stim = Stimulus; rec = recording; Lat N75 = Latency at the point N75; Lat P100 = Latency at point P100; Lat N140 = Latency at point N140; Ampl = Amplitude; Oz-Fz = Occipital zone-Frontal zone



FIG. 2

Non-contrast computed tomography of case four, axial section, showing erosion of the lateral sphenoid wall on the right side, with bilateral disease in the sphenoid sinuses.

papilloedema in all our cases, in agreement with other reports.³⁻⁵

Non-contrast CT of the paranasal sinuses was suggestive of allergic fungal sinusitis. In three cases, there was evidence of bony erosion of the lateral sphenoid wall in the region of the optic nerve. In the fourth case, the visual loss was on the contralateral side to the bone erosion.

There was an absolute correlation between radiological and surgical findings. After surgical decompression, there was an immediate post-operative (day zero) visual improvement, which improved further with oral prednisolone. Histopathology did not reveal tissue invasion in any case. Other reports in the literature have shown the role of steroids and surgical decompression in such cases.³⁻⁵

In cases of allergic fungal sinusitis, the mechanism of vision loss has thus far been assumed to be secondary to direct compression or direct inflammation of the optic nerve. We did find bony erosion in the region of the lateral sphenoid wall in three cases.



FIG. 3

Non-contrast computed tomography of case four, coronal section, showing erosion of the lateral sphenoid wall on the right side, with bilateral disease in the sphenoid sinuses and erosion of sphenoid roof.

However, in the fourth case, there was erosion on the side with normal vision.

We therefore suggest that a different mechanism was responsible for the visual loss seen in this fourth case. It is possible that aberrant anatomical pathways present in the region of the optic canal could have been responsible for direct inflammation of the nerve in the absence of obvious bony

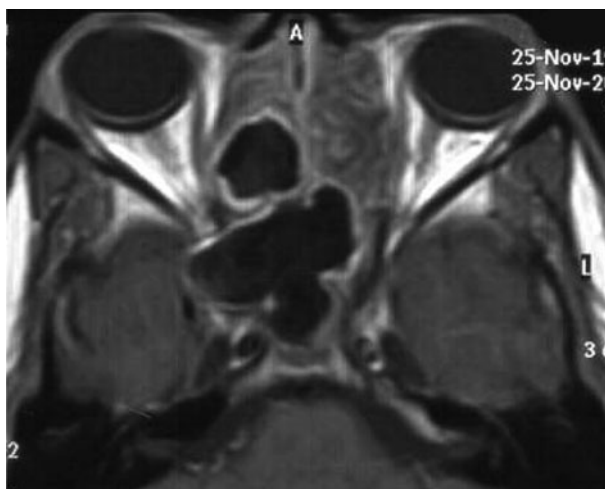


FIG. 4

Magnetic resonance imaging of case four, axial section, showing erosion of the lateral sphenoid wall on the right side, with bilateral disease of the sphenoid sinuses.



FIG. 5

Magnetic resonance imaging of case four, coronal section, showing erosion of the lateral sphenoid wall on the right side, with bilateral disease of the sphenoid sinuses.

erosion. However, in view of the operative, radiological and laboratory findings of the case, with the suggestion of a hyperimmune response to fungal antigens (in the form of raised IgE and positive fungal serology), we suggest that a local immunological reaction to the fungal antigens might be responsible for this patient's visual loss. The debulking surgery (which reduced the antigen load) and steroids (which reduced the immunological response) could therefore have been responsible for restoration of this patient's vision. The role of immunotherapy in allergic fungal sinusitis has been studied, and a reduced reliance on steroids has been suggested.⁷

- **Fungal infection of the paranasal sinuses is classified into five variants on the basis of histopathology. Allergic fungal sinusitis is the most commonly encountered variant, with nasal obstruction being the most common presentation**
- **This paper describes the presentation of four allergic fungal sinusitis cases with sudden loss of vision**
- **The authors suggest that a local immunological reaction to the fungal antigens might be responsible for such visual loss, in addition to mechanical compression of the optic nerve, in cases of allergic fungal sinusitis**

We therefore suggest that, apart from mechanical compression of the optic nerve, a local immunological reaction to fungal antigens might be responsible for the visual loss seen in allergic fungal sinusitis. Henceforth, we suggest that all cases of allergic fungal sinusitis with visual loss should be given the advantage of surgical decompression as well as medical treatment in the form of steroids.

References

- 1 Jay WM, Bradsher RW, LeMay B, Synderman N, Angtuaco EJ. Ocular involvement in mycotic sinusitis caused by bipolaris. *Am J Ophthalmol* 1988;**105**:366–70
- 2 Carter KD, Graham SN, Carpenter KM. Ophthalmic manifestations of allergic fungal sinusitis. *Am J Ophthalmol* 1999;**127**:189–95
- 3 Marple BF, Gibbs SR, Newcomer MT, Mabry RL. Allergic fungal sinusitis induced visual loss. *Am J Rhinol* 1999;**13**:191–5
- 4 Dunlop IS, Billson FA. Visual failure in allergic aspergillus sinusitis: case report. *Br J Ophthalmol* 1988;**72**:127–30
- 5 Graham SM, Carter KD. Response of visual loss in allergic fungal sinusitis to oral corticosteroids. *Ann Otol Rhinol Laryngol* 2005;**114**:247–9
- 6 Gupta AK, Bansal S, Gupta A, Mathur N. Is fungal infestation of paranasal sinuses more aggressive in pediatric population? *Int J Paediatr Otorhinolaryngol* 2006;**70**:603–8
- 7 Mabry RL, Marple BF, Folker RJ, Mabry CS. Immunotherapy for allergic fungal sinusitis: three years' experience. *Otolaryngol Head Neck Surg* 1998;**119**:648–51

Address for correspondence:
Dr Sandeep Bansal,
Senior Research Associate,
Department of Otolaryngology,
PGIMER,
Sector 12, Chandigarh 160012, India.

Fax: 91 172 2744401
E-mail: drsandeepb@rediffmail.com

Dr S Bansal takes responsibility for the integrity of the content of the paper.
Competing interests: None declared
