Gaze-evoked Amaurosis

A Report of Five Cases

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Objective: To highlight the various causes of gaze-evoked amaurosis.

Design: Retrospective noncomparative interventional case series.

Participants: Five patients treated at our facility over the past 6 years.

Methods: Clinical presentation, radiologic studies, surgical management, and postsurgical results are presented.

Main Outcome Measures: Visual acuity, clinical findings of gaze-evoked amaurosis.

Results: Only two patients had classic intraorbital etiologies, one with an intraconal cavernous hemangioma and one with an intraconal foreign body. Three patients had extraorbital processes, two with orbital fractures and one with a sinus tumor. Only two of our patients initially were aware of the gaze-evoked amaurosis at presentation. Appropriate surgery was curative in all cases.

Conclusions: Gaze-evoked amaurosis is a rare condition, classically implicating intraconal orbital pathology. In one of the largest case series published to date, we found extraorbital etiologies are also capable of producing gaze-evoked vision loss. Gaze-evoked amaurosis should be suspected and tested for in any orbital condition. *Ophthalmology 2003;110:322–326 © 2003 by the American Academy of Ophthalmology.*

Gaze-evoked amaurosis (GEA) is a rare condition in which transient visual obscurations occur only during eccentric positions of gaze. It is caused by extrinsic position-dependent occlusion of the retinal or optic nerve circulation.[1,2] The condition classically implicates intraconal orbital pathology, although other uncommon extraconal etiologies have also been reported.[3–7] We present five patients, only two of whom manifested classic intraorbital etiologies; one an intraconal cavernous hemangioma and the other an intraconal foreign body. Of the remaining three patients, two had bony fractures with extrinsic optic nerve compression, and one had a juvenile nasopharyngeal angiofibroma with orbital, but extraperiosteal, extension. Only two of our patients were aware of gaze-evoked visual disturbance at presentation. Surgical intervention was curative in all cases. The fact that most patients are unaware of positional blindness, and the ability to resolve this condition with appropriate surgical therapy, suggests that all patients with an orbital disease process should be screened for gaze-evoked amaurosis.

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Case 1: Juvenile Nasopharyngeal Angiofibroma

A 16-year-old male was seen with a 6-month history of progressive left-sided facial swelling, nasal obstruction, and anosmia unresponsive to antibiotics, and more recent onset of left-sided facial pain, paresthesia, epistaxis, and blurred vision. He denied GEA. Imaging revealed an enhancing, heterogeneous, vascular nasopharyngeal mass in the left nasal cavity and maxillary sinus, extending into the anterior and middle cranial fossae, the left infratemporal fossa, and extraperiosteally within the orbit close to the inferior rectus (Fig 1). Visual acuity in neutral gaze was 20/20 in the right eye and 20/50 in the left eye, with reactive pupils and a 1+

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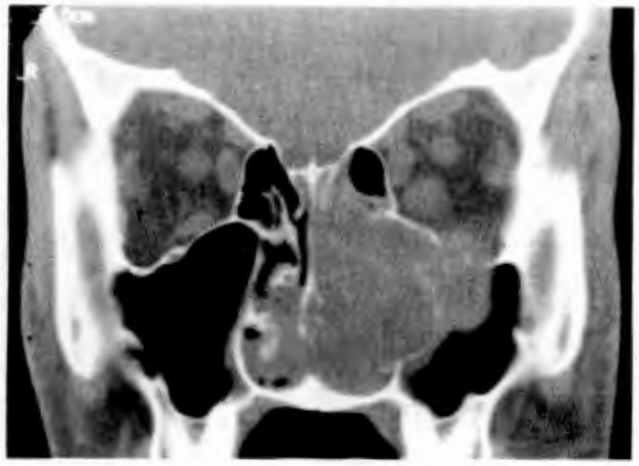

Figure 1. Coronal computed tomogram showing nasopharyngeal mass eroding through the left orbital floor. Pathology was consistent with juvenile nasopharyngeal angiofibroma.

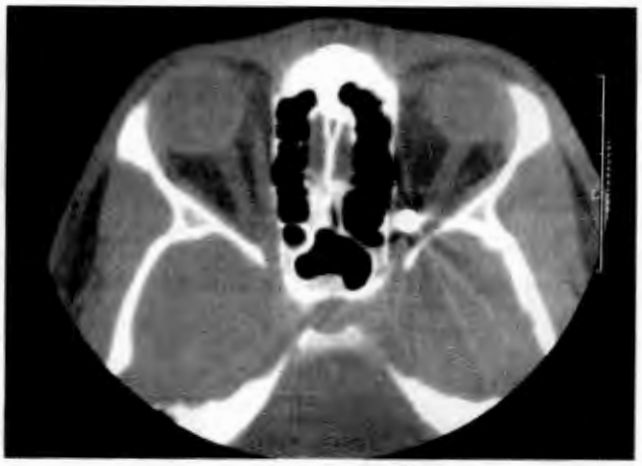

Figure 2. Axial computed tomogram of the orbits showing 6-mm BB at left orbital apex between the medial rectus and the optic nerve.

afferent pupillary defect (APD) in the left eye. There was mild resistance to retropulsion of the left globe, but ductions were full without diplopia. Fundus examination was normal. In left gaze, visual acuity in the left eye dropped abruptly to 20/800, with marked dilation of the pupil. Resumption of primary gaze resulted in immediate return to baseline visual acuity and pupillary reaction. The mass was entirely removed by means of endoscopic surgery after embolization of its vascular supply. Subperiosteal displacement of the left inferotemporal periorbita was confirmed at surgery, without invasion of the orbital soft tissues by the mass. Histopathology revealed juvenile nasopharyngeal angiofibroma. Postoperatively, vision returned to 20/20 with resolution of GEA.

Case 2: Retained Intraorbital Ball Bearing

A 23-year-old male complained of diplopia of 4 months' duration. Best-corrected visual acuity was 20/20 in the right eye and 20/20 in the left eye with reactive pupils. Examination of the right eye was normal, but abnormalities of the left eye included an area of hyperplastic conjunctiva medially, a 3+ APD in primary gaze, optic nerve pallor, and evidence of chronic retinitis sclopetaria, as previously reported.[8] Despite denial of previous ocular trauma or surgery, radiologic studies revealed a spherical 6-mm metallic foreign body at the left orbital apex between the left medial rectus and optic nerve (Fig 2). When presented with these findings, the patient vaguely recalled being hit in the left eye by a BB approximately 12 years earlier but denied previous medical intervention. Given the location of the BB within the orbit, we strongly suspected that GEA would be uncovered during clinical examination. However, despite detailed questioning regarding symptoms of prior amaurotic episodes and repeated testing for the presence of GEA on multiple occasions, no evidence for the existence of this condition could be elicited. As part of this examination, the patient was counseled at great length about symptoms to be expected if GEA were to become manifest. With a stable ophthalmic examination, aside from his persistent diplopia, the patient underwent successful surgical correction of his strabismus. The patient's postoperative course was uncomplicated until 1½ months after surgery, when he reported two brief episodes of nongaze-dependent amaurosis in the left eye. These episodes progressed over the next 7 months to GEA in the left eye that could be consistently elicited during sustained right gaze. Examination verified decreased visual acuity from 20/20 to 20/800 in the left eye during adduction, with persistence of a 3+ APD. Serial visual fields also demonstrated progressive central visual loss, at which point the patient elected to undergo craniotomy and superior orbitotomy for removal of the BB. GEA completely resolved after surgery, with visual acuity of 20/20 in both eyes and improved visual fields that remain stable 1½ years postoperatively.

Case 3: Medially Displaced Zygomaticomaxillary Complex Fracture

A 55-year-old male was the unrestrained driver in a rollover motor vehicle accident, with resulting closed head trauma but no loss of consciousness. Examination at presentation was significant for a markedly displaced left zygomaticomaxillary complex fracture. Visual acuity was 20/20 in both eyes in primary gaze, with reactive pupils and a 1+ APD in the left eye, but with horizontal diplopia from limited abduction of the left eye. The patient noted desaturation to both brightness and color in the left eye, without evidence of optic nerve inflammation or swelling. Although he was initially unaware of GEA, on sustained left gaze the patient's visual acuity in the left eye dropped abruptly to counting fingers accompanied by a gaze-evoked 3+ APD. Visual acuity and pupillary reaction returned to baseline within seconds of resuming primary gaze. Computed tomography demonstrated a lateral orbital wall fragment close to the lateral aspect of the left optic nerve (Fig 3). Surgical reduction and repair of the zygomaticomaxillary complex

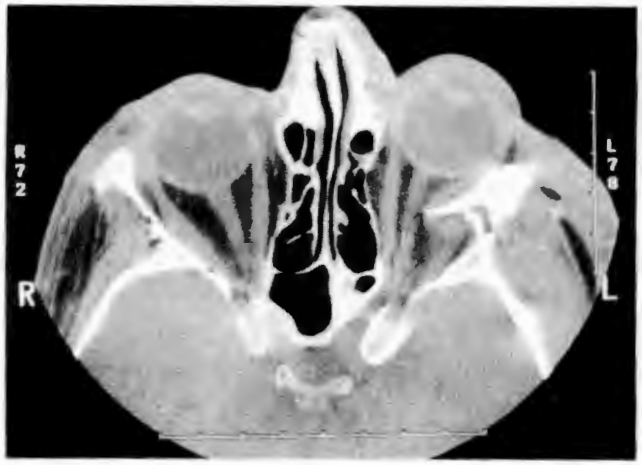

Figure 3. Axial computed tomogram of the orbits showing medially displaced zygomaticomaxillary complex fracture and impingement of the optic nerve.

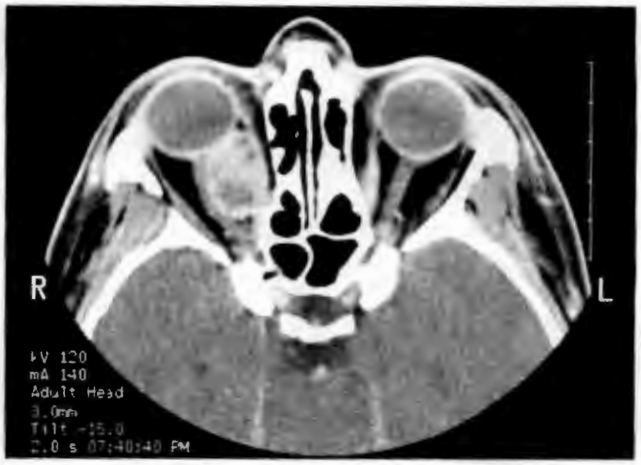

Figure 5. Axial computed tomogram of the orbits showing a right intraconal cavernous hemangioma, abutting and displacing the optic nerve.

and lateral orbital wall fractures resulted in complete resolution of GEA.

Case 4: Medial Wall Fracture

A 37-year-old male fell 15 feet from the roof of his house and sustained multiple head injuries. At presentation he complained of diplopia and "graying out" of vision in the right eye during left gaze. Visual acuity was 20/20 in both eyes with reactive pupils, a trace APD in the right eye in primary gaze, and normal retropulsion, brightness, and color saturation in both eyes. Dilated examination was normal in both eyes. With sustained adduction of the right eye, vision quickly decreased to counting fingers with worsening of the APD to 3+. Vision and pupillary reaction returned to baseline within seconds of resuming primary gaze. Computed tomography revealed numerous bony defects, including a right zygomaticomaxillary complex fracture, a nondisplaced fracture through the right optic canal, and a comminuted fracture of the right medial orbital wall, with a fragment close to the right optic nerve (Fig 4). Surgical intervention included repair of the medial orbital wall and zygomaticomaxillary complex fractures, but repair of the optic canal fracture was not attempted because of the greater likelihood of the right medial wall fracture being responsible for the observed gaze-dependent optic neuropathy. At 3 months follow-up, ocular motility and visual acuity had returned to baseline, and there was no residual APD or recurrence of GEA.

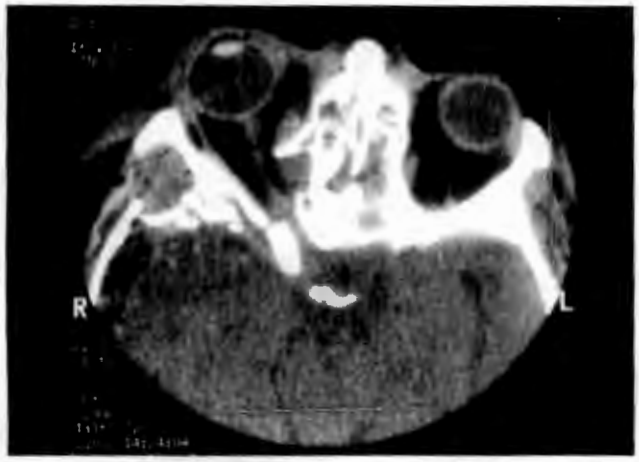

Figure 4. Axial computed tomogram of the orbits showing laterally displaced medial orbital wall fragment and impingement of the optic nerve.

Case 5: Cavernous Hemangioma

A 41-year-old female was seen with a 6-month history of painless periorbital fullness and transient visual loss in the right eye noticed only while shaving her right axilla. Visual acuity in primary gaze was 20/30 in the right eye and 20/20 in the left eye. Pupils were reactive with a 1+ APD in the right eye. There was 3 mm of proptosis in the right eye but normal orbital retropulsion. Funduscopic examination showed a swollen right optic disc with vascular engorgement. At 20° of right gaze, vision in the right eye decreased abruptly to counting fingers at 2 feet, and the APD worsened to 2+. Both of these abnormalities returned to baseline within 10 seconds of resuming primary gaze. Imaging revealed a large intraconal mass in the medial orbit abutting the optic nerve (Fig 5). A 2 × 2 × 3 cm cavernous hemangioma was removed by means of combined lateral and medial orbitotomy. Mild lateral rectus weakness and a peripheral visual field defect in the right eye persisted 7 months after surgery, but visual acuity remained at baseline, and both the APD and GEA completely resolved.

Discussion

GEA is a rare condition, resulting from intraconal pathology in 82% of reported cases. Our review of the 34 previously reported cases of GEA revealed that optic nerve sheath

Table 1. Details of Patients in Current Series

Case	Age/Gender	Pathology	Side	Direction of Gaze-evoked Amaurosis	Visual Acuity in Primary Gaze	Visual Acuity during Gaze-evoked Amaurosis	Location of Lesion Relative to Optic Nerve	Direction of Gaze-evoked Amaurosis Relative to Location of Lesion	Aware of Gaze-evoked Amaurosis at Initial Examination
1	16/M	JNA	OS	Abduction	20/50	20/800	Inferotemporal	Same side	No
2	23/M	Retained FB	OS	Adduction	20/20	20/800	Medial	Same side	No
3	55/M	Lateral wall Fxr	OS	Abduction	20/20	CF	Lateral	Same side	No
4	37/M	Medial wall Fxr	OD	Adduction	20/20	CF	Medial	Same side	Yes
5	41/F	Cavernous hemangioma	OD	Abduction	20/30	CF	Medial	Opposite side	Yes

CF = counting fingers; FB = foreign body; Fxr = fracture; JNA = juvenile nasopharyngeal angiofibroma; OD = right eye; OS = left eye.

meningiomas[1,5,9–12] and cavernous hemangiomas[1,13–15] are the two most common causes of this condition, accounting for 35% and 18% of all cases, respectively. Other intraconal disease processes include traumatic hemorrhagic cyst,[2] optic nerve glioma,[10,11] granular cell myoblastoma,[11] orbital metastasis from renal cell carcinoma,[16] retained metallic foreign body,[17] angiomyoma,[18] thyroid orbitopathy,[19] and neurofibromatosis type II.[20] Rare causes of GEA from extraconal pathology such as pseudotumor cerebri,[3,4] orbital wall fracture,[5] vitritis,[6] and osteoma[7] have also been reported. Of the five cases in our series, only two demonstrated intraconal pathology, with only one of these being a cavernous hemangioma. Our case 1 (juvenile nasopharyngeal angiofibroma) shows that extraorbital masses can also produce the same phenomenon. Despite the varied nature of the offending disease processes, the common etiology in all cases seems to be compromise of the retinal or optic nerve circulation, either by means of compression of the central retinal artery or disruption of the optic nerve microvasculature.[21] Using Doppler ultrasonographic imaging, Knapp et al[2] demonstrated complete disappearance of the arterial waveform in the central retinal artery during GEA that was causing a visual decrease to no light perception. Fluorescein angiography has also been used to document a marked decrease in perfusion to the optic nerve head and surrounding choroid and retinal arterioles during amaurotic gaze.[9,11,12] Pseudotumor cerebri, although lacking a localized compressive lesion, may precipitate amaurosis when eccentric eye movements cause pressures to increase within an already tensely dilated optic nerve sheath.[3,4]

It has been reported that, in most cases of GEA, the compressing mass is located on the side of the optic nerve that is opposite the direction of amaurotic gaze, such that a medial lesion would cause amaurosis in lateral gaze and vice versa. This was not the case in four of our five patients: the juvenile nasopharyngeal angiofibroma, the two orbital fractures, and the one retained intraorbital BB (Table 1). These variations suggest that compression of the nerve's circulation depends not only on the location of the mass within the orbit but also on its proximity to the optic nerve and that the direction of amaurotic gaze may not always be helpful in localization of the offending lesion.

Based on review of the literature, it is evident that GEA can be caused by a variety of pathologic conditions. Although most often the result of intraconal pathology, GEA can also be caused by any extraorbital process that exerts pressure on the orbit and optic nerve. Because many patients are unaware of visual obscurations in extreme directions of gaze, and because suspicion for this phenomenon is low for conditions that are other than intraconal, we recommend routine testing for GEA with any orbital disease process. Testing for GEA is easily performed by having the patient look in all directions of gaze, having the patient hold each eccentric position of gaze for at least 5 seconds, and noting any changes in visual acuity or pupil reactivity. Changes in vision from primary to eccentric gaze can be evaluated either by using a near visual acuity card or by noting changes in the patient's subjective response.

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Historical Image

Rimless pince-nez with case, 1892. This gold, rimless pince-nez features nosepieces with either plastic or tortoise shell inlay. Pince-nez cases were not popular since the eyewear was taken off and put back on so frequently. An engraved sterling silver case was certainly the exception.

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