Journal of Neuro-Ophthalmology 20(4): 259–263, 2000

Pneumosinus Dilatans of the Sphenoid Sinus Presenting With Visual Loss

Craig A. Skolnick, MD, Mahmood F. Mafee, MD, and James A. Goodwin, MD

Objective: To report 3 cases of pneumosinus dilatans of the sphenoid sinus associated with visual loss.
Materials and Methods: Retrospective case series describing history of visual loss, visual examination, visual field deficits, and radiologic imaging.
Results: Three patients developed visual loss associated with pneumosinus dilatans of the sphenoid sinus.
Conclusions: Pneumosinus dilatans of the sphenoid sinus is a rare disorder that should be considered in patients presenting with unexplained visual loss.
Key Words: Optic neuropathy—Pneumosinus dilatans—Sphenoid sinus.

Pneumosinus dilatans is a rare condition in which dilated paranasal sinuses lined by normal mucosa are filled with air. Classically, there is no overlying osseous hypertrophy or destruction of bone. The frontal sinus is the most commonly affected, but the sphenoid sinus is the most important for visual loss because of its intimate relation with the optic nerve in the optic canal. This report concerns three patients with visual symptoms and sphenoidal pneumosinus dilatans.

CASE REPORTS

Case 1

A 48-year-old African-American woman had gradual painless loss of vision OD over 6 months. She has had diffuse, nonlocalizing headaches for 3 years, occasionally associated with nausea. There is no history of trauma. She has euthyroid thyroidomegaly, hyperparathyroidism, early menopause, and Zollinger-Ellison syndrome with severe peptic ulcer disease. She has had yearly prolactin levels and thyroid function tests, the results of which have been normal. Her visual acuity was hand motions OD and 20/20 OS, with a relative afferent pupillary defect OD. Goldmann visual field testing OD showed a large absolute nasal and central defect. The visual field was normal OS. Fundus examination showed temporal pallor and increased cupping of the right optic disc and normal left optic disc. Computed tomography (CT) showed dehiscence of both optic canals (right greater than left), with the intracanalicular portion of the right optic nerve seen in close contact with the air (Fig. 1A). Magnetic resonance imaging (MRI) with gadolinium contrast showed extensive pneumatization of the sphenoid bone extending toward the right optic canal with an area of air intensity adjacent to the right optic nerve (Fig. 1B).

Case 2

A 26-year-old Hispanic woman with rhinitis had immediate loss of the upper field of vision OD after blowing her nose. There was some recovery of visual field after an hour, leaving a permanent nasal scotoma that respected the horizontal midline and extended nearly to the physiologic blind spot (Fig. 2). The visual field was normal OS. She experienced some transient flashing pink lights in the part of the upper field that recovered. She denies trauma or headaches. Visual acuity was 20/25 OD and 20/15 OS, with no relative afferent pupillary defect. Examination 2 years after presentation showed sectoral pallor of the inferotemporal optic disc with corresponding loss of nerve fiber layer striations. Examination 1 year after onset had not shown these findings. MRI with gadolinium contrast showed pneumatization of the sphenoid sinus and posterior ethmoid cells adjacent to the optic canal. CT of the orbits showed an area of bony dehiscence between the right posterior ethmoid air cell and the floor of the optic canal. This bony wall was intact on the left side (Fig. 3).

Case 3

A 25-year-old white man had eight episodes of transient visual loss OS over a 6-month period. A typical episode began with a patchy central and peripheral darkness that progressed over 2 minutes, leaving a small central island, which then closed completely, at which time the eye had no light perception. This lasted approximately 15 minutes, followed by gradual opening in a

Manuscript received February 17, 2000; accepted June 27, 2000.
From the Department of Ophthalmology and Visual Sciences, University of Illinois at Chicago College of Medicine, Chicago, Illinois; and the Department of Radiology, University of Illinois at Chicago College of Medicine, Chicago, Illinois.
Address correspondence and reprint requests to James A. Goodwin, M.D., Department of Ophthalmology and Visual Sciences, University of Illinois at Chicago College of Medicine, 1855 West Taylor Street, Chicago IL 60612.

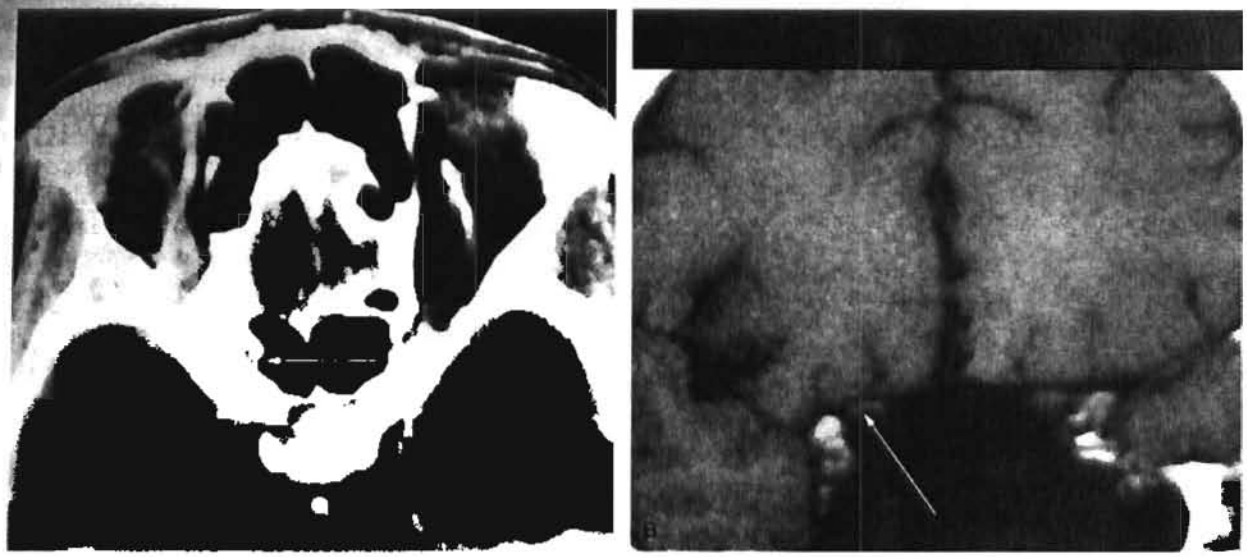

FIG. 1. Case 1—CT (1a) reveals dehiscence of both optic canals (right greater than left) with the intracanalicular portion of the right optic nerve seen in close contact with the air (arrow). MRI with gadolinium contrast (1b) demonstrates extensive pneumatization of the sphenoid bone extending towards the right optic canal with an area of air intensity adjacent to the right optic nerve. Arrow indicates air indenting optic nerve from below. MRI and CT images are enhanced for contrast only.

patchy fashion. He denied any particular activity, event, or posture temporally related to the attacks. There has never been pain or headache associated with the attacks, but occasionally he has had a steady pressure over his temples and behind his eyes. Visual acuity was 20/15 OU without afferent pupillary defect. Results of Goldmann visual fields and fundus examination were normal OU. Work-up for collagen-vascular disease indicated an elevated anti-Smith antibody. CT showed extensive pneumatization of the sphenoid bone with extension into the lesser wing of the sphenoid and anterior clinoid adjacent to the left optic canal (Fig. 4). Part of the optic canal was dehiscent, and the optic nerve appeared to be in close contact with the air space.

DISCUSSION

Frontal pneumosinus dilatans was first fully described by Benjamin in 1918 (1). Lombardi et al. (2) reviewed the literature in 1967 and found 51 cases, with 39 involving the frontal sinus and only five involving the spheno-ethmoidal sinus. Most patients were male (48 of 51), and most were between 20 and 40 years of age. The presentation depends on the sinus affected. Frontal sinus involvement may produce localized tenderness or variable proptosis secondary to orbital communication. Complaints may include headache and visual disturbances such as decreased visual acuity, bitemporal hemianopia, and diplopia secondary to motility disorder. The

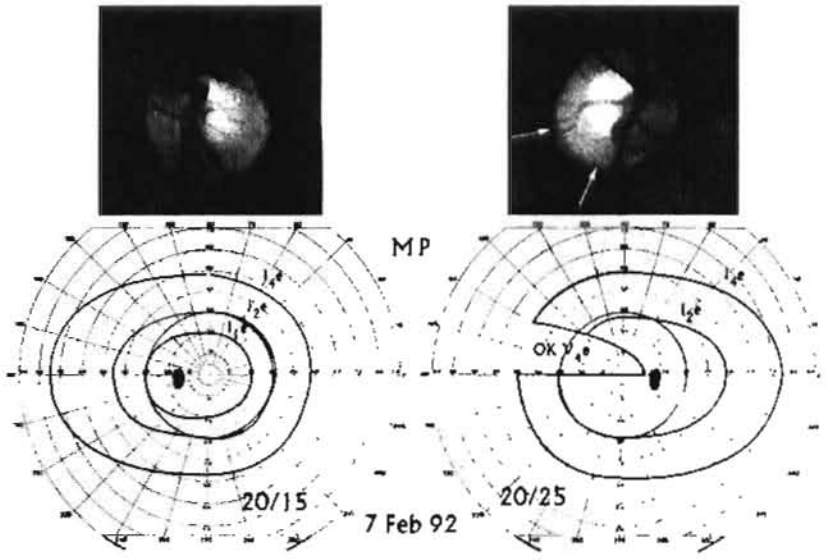

FIG. 2. Case 2—Sectoral pallor of the right inferotemporal optic disc (between arrows) with loss of nerve fiber layer striations. Corresponding OD visual field depicts nasal scotoma that respects the horizontal midline and extends nearly to the physiologic blind spot. Normal left optic disc and visual field. Both disc photos are equally enhanced for color and contrast.

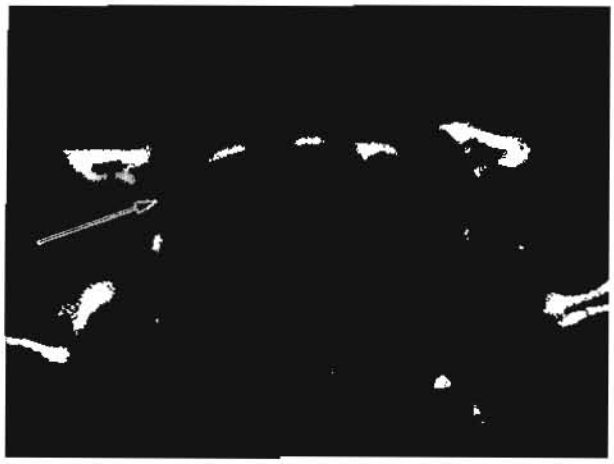

FIG. 3. Case 2—CT shows an area of bony dehiscence between the right posterior ethmoid air cell and the floor of the optic canal (arrow). This bony wall is intact on the left side. CT images are enhanced for contrast only.

differential diagnosis includes mucocele, acromegaly, pneumatocele, fibrous dysplasia, and Sturge-Weber syndrome. There is a predilection for certain sites: the lateral recesses of the frontal sinuses, the superior recess of the maxillary sinus, the sellar region in the spheno-ethmoidal sinuses, and the orbit with anterior ethmoidal involvement.

During development, ethmoid sinus cells expand within the ethmoid box, leading to compression and displacement of cancellous bone, forming compact bone. The stimulus for pneumatization of the paranasal sinuses is thought to be growth of the mucosal lining into bone. Sinuses grow slowly until puberty, after which they grow rapidly to their adult size. There is a wide variability in "normal pneumatization" of the paranasal sinuses, with the sphenoid sinus showing the most variability. Pneumatization of the sphenoid sinus may occur in the dorsum sella, the clivus, the clinoid processes, pterygoid plates, and the greater and lesser wings of the sphenoid bone. Simple aeration of the anterior clinoid without enlargement or bulging is a normal variant, occurring in 13% of the population (3). Explanation for sinus dilatation remains speculative. Suggestions have included congenital abnormality, inflammation (4), and a valve-like obstructive mechanism of the sinus (5). Pneumosinus dilatans may be static for some time, and then overgrowth may rapidly occur.

Pneumosinus dilatans has been associated with meningioma and fibro-osseous disease. Lloyd (6) described six cases presenting with proptosis, three of which had meningioma associated with fronto-ethmoidal sinus dilatation. Two patients with fibrous dysplasia had maxillary antral dilatation, and one patient with an ossifying fibroma of the posterior orbit had fronto-ethmoidal sinus dilatation. Wiggli and Oberson (7) reported seven patients with anterior chiasmatic angle meningiomas associated with spheno-ethmoid dilatation with variable degrees of hyperostosis. This same relationship has been reported without overlying hyperostosis (8). Hirst et al. (4) have reported three cases of sphenoidal pneumosinus dilatans associated with intracanalicular meningiomas of the optic nerve sheath without adjacent hyperostosis. Two of these cases had bilateral meningiomas. Spoor et al. (9) described a case of pneumosinus dilatans of the frontal and sphenoid sinuses in combination with Klippel-Trenaunay-Weber syndrome (port-wine hemangiomas, deep venous abnormalities, and soft tissue and bony hypertrophy) and familial Adie's pupil who developed bilateral optic nerve sheath meningiomas. Patients with sphenoidal pneumosinus dilatans and unexplained progressive visual loss may have occult meningiomas of the optic nerve sheath missed with standard neuroimaging.

Pneumosinus dilatans without an associated pathologic process rarely causes visual loss. A total of 11 cases have been previously reported in the literature (2,5,10–16). Two patients underwent craniotomy, and two underwent sphenoid sinus decompression. Sugita et al. (5) described a young man with multiple episodes of transient bilateral complete blindness, occurring while driving up a mountain or after take-off in an airplane. His visual acuity at presentation was counting fingers OD and 20/30 OS, and tomograms suggested bone defects in the sphenoid sinus around the optic canals. The attack was inducible in a hyperbaric chamber when the atmospheric pressure was lowered. The patient underwent sphenoid sinus decompression via trans-maxillary sinusotomy. Postoperatively, the patient's visual acuity improved to 20/70 OD and 20/20 OS, and he did not have a recurrent episode of transient blindness, even when rechallenged in the hyperbaric chamber. The authors speculated that air inside an abnormally large sphenoid sinus can expand under decreased atmospheric pressure and push the optic nerve through a bony defect, thus causing a disturbance of the regional blood flow.

Approximately 1 mm of optic canal wall separates the optic nerve from the sinus cavity. Excessive pneumatization can lead to thinning and gross dehiscence of the canal wall. The optic nerve is bound within the optic canal by the dural sheath and is relatively immobile and susceptible to local forces. Radiographic (CT) studies of patients with inflammatory sinus disease and suspected optic nerve disease have shown up to 3% of optic nerves contact or protrude slightly into the posterior ethmoid air cells (17,18), and the optic canal traverses or bulges into the sphenoid sinus in 6% to 8% of patients (18,19). The frequency of dehiscent bone between the sphenoid sinus and the optic nerve is 4% in two independent cadaveric studies (20,21). CT studies have shown up to a 24% incidence of optic canal bony dehiscence as defined by absence of bone density along the medial wall of the optic canal (18). Differences in frequency may be attributable to inability to visually discern bone thickness less than 0.5 mm on a CT examination. The presence of anterior clinoid pneumatization increases the likelihood of optic nerve exposure and canal dehiscence (18).

The mechanism leading to optic neuropathy is uncertain. With direct communication between the sinus and the optic canal, one could postulate a direct compressive effect by mucosa or air leading to ischemic damage.

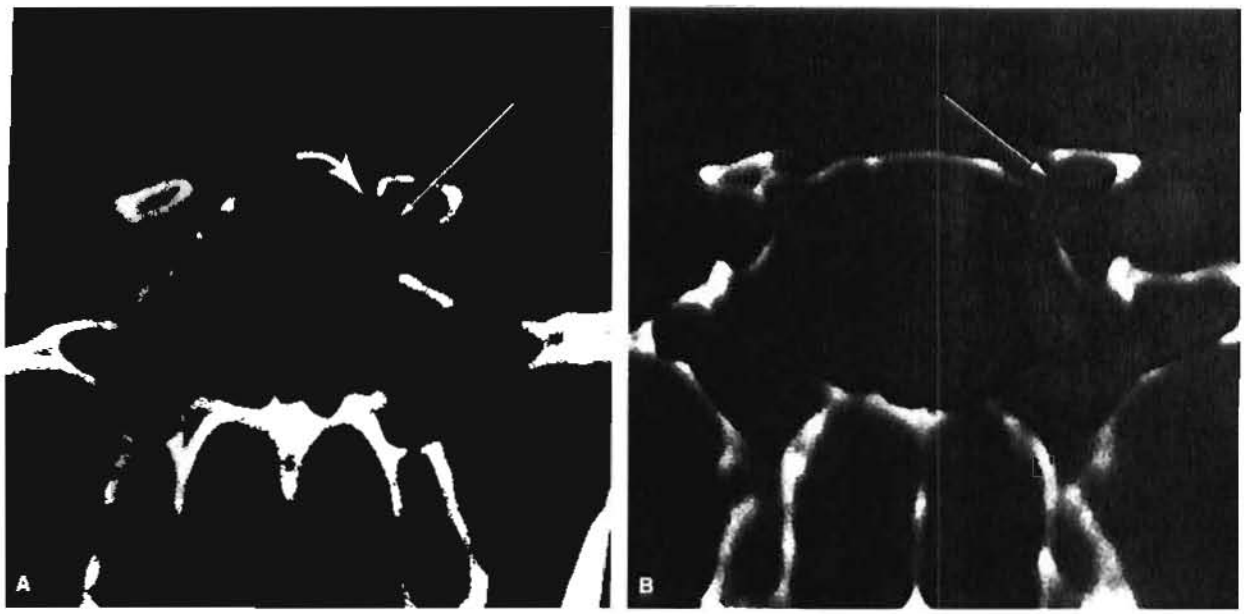

FIG. 4. Case 3—CT demonstrates extensive pneumatization of the sphenoid bone with extension into the lesser wing of the sphenoid and anterior clinoid (4a, 4b straight arrow) adjacent to the left optic canal (4a curved arrow). Part of the optic canal is dehiscent and the optic nerve appears to be in close contact with the air space (4a, 4b straight arrow). CT images are enhanced for contrast only.

Upward displacement of both the pituitary fossa and planum sphenoidale with distortion of the tuberculum sellae may cause direct compression of the optic chiasm or interfere with chiasmal circulation in patients who present with bitemporal visual field defects (22). Three of 11 previously reported cases had this radiologic finding (2,13,15).

None of the three cases presented here underwent surgical decompression. It is possible that an occult meningioma of the optic nerve sheath exists in these patients. Patients with unexplained optic atrophy and normal neuroimaging should be followed-up as meningioma suspects. Patients should receive periodic CT and MRI, with special attention to the optic canals and sellar region.

These cases represent the association of a specific radiographic finding with three different clinical presentations. Because of the small number of cases and varying subjective and objective data, it is difficult to clinically characterize sphenoidal pneumosinus dilatans. Although cases 1 and 2 displayed optic neuropathy, one cannot definitely determine the cause to be pneumosinus dilatans. Case 2 did not have a detectable relative afferent pupillary defect on examination. It is possible that the disparity in optic nerve function was not detectable, because the total area of affected visual field was small. Videopupillography was not available when this patient initially presented. Case 3 lacks objective evidence of optic nerve disease. This patient may in fact be having a variant of visual migraine; however, the association of his exclusively left eye symptoms and pneumosinus dilatans on the same side is compelling.

Because this is a rare disorder, it is not clear how to manage these patients. It has been suggested that sudden elevation of the intrasinus pressure as with sneezing or with altitude change may cause direct damage to an exposed optic nerve. Our second patient (case 2) is a dramatic example of this mechanism, and for her, surgical options aimed at preventing future attacks or progressive optic neuropathy must be considered. These would include creating an outlet for decompression from the sphenoid into the maxillary sinus (transmaxillary sinusotomy), enucleating the sinus to remove the mucosal lining (because it is thought to be the stimulus for pneumatization [12]), or packing the sinus with fat to tamponade a bony dehiscence in the optic canal.

REFERENCES

1. Benjamins CE. Pneumosinus frontalis dilatans. *Acta Otolaryngol* 1918;1:412.
2. Lombardi G, Passerini A, Cecchini A. Pneumosinus dilatans. *Acta Radiol (Diagn) (Stockh)* 1968;7:535–542.
3. Scuderi AJ, Harnsberger HR, Boyer RS. Pneumatization of the paranasal sinuses: Normal features of importance to the accurate interpretation of CT scans and MR images. *Am J Roentgenol* 1993; 160:1101–4.
4. Hirst LW, Miller MR, Hodges FJ, et al. Sphenoid pneumosinus dilatans: a sign of meningioma originating in the optic canal. *Neuroradiology* 1982;22:207–10.
5. Sugita K, Hirota T, Iguchi I, et al. Transient amaurosis under decreased atmosphere pressure with sphenoid sinus dysplasia. *J Neurosurg* 1977;46:111–4.
6. Lloyd GA. Orbital pneumosinus dilatans. *Clin Radiol* 1985;36: 381–6.
7. Wiggli U, Oberson R. Pneumosinus dilatans and hyperostosis: Early signs of meningiomas of the anterior chiasmatic angle. *Neuroradiology* 1975;8:217.
8. Leonardi M, Fabris G. Le pneumosinus dilatans, signal radiologique direct des meningiomas de l'angle antereur du chiasma. *Ann Radiol (Paris)* 1976;19:803.

9. Spoor TC, Kennerdell JS, Maroon JC, et al. Pneumosinus dilatans, Klippel-Trenaunay-Weber syndrome, and progressive visual loss. *Ann Ophthalmol* 1981;13:105–8.
10. Reicher MA, Bentson JR, Halbach VV, et al. Pneumosinus dilatans of the sphenoid sinus. *Am J Neuroradiol* 1986;7:865–8.
11. Stretch JR, Poole MD. Pneumosinus dilatans as the aetiology of progresive bilateral blindness. *Br J Plast Surg* 192:45:469–73.
12. Williams JP, Shawker TH, Lora J. Pneumosinus dilatans of the sphenoid sinus. *Bulletin of the Los Angeles Neurological Societies* 1975;40:45–8.
13. Agati D. Ipertrofie etmoido-sfenoidali a protundenza endocranica. *Radiol Med* 1946;32:151–4.
14. Bendescu I. Beiderseitige Optikusatrophie, verursacht durch Pneumosinus dilatans der rechten Keilbeinhöhle. Z *Augenheilk* 1932;79:41–50.
15. Macialowicz T. A case of dilating pneumosinus of the sphenoid sinus and the posterior ethmoid cells. *Pol Rev Radiol Nucl Med* 1969,33(4):324–30.
16. Hajek M. Zwei verschiedene, bisher nicht beschriebene Tumorarten der Stirnhöhle und des Siebbeinlabyrinthes in ein und demselben Individuum. *Passow-Schäfers Beitr*. 1926;23:465.
17. Bansberg SF, Harner SG, Forbes G. Relationship of the optic nerve to the paranasal sinuses as shown by computed tomography. *Otolaryngol Head Neck Surg* 1987;96 331–5.
18. DeLano MC, Fun FY, Zinreich SJ. Relationship of the optic nerve to the posterior paranasal sinuses: A CT anatomic study. *Am J Neuroradiol* 1996;17:669–675.
19. Dessi P, Moulin G, Castro F, et al. Protrusion of the optic nerve into the ethmoid and sphenoid sinus: prospective study of 150 CT studies. *Neuroradiology* 1994;36:515–6.
20. Fuji K, Chambers SM, Rhoton AL Jr. Neurovascular relationships of the sphenoid sinus. *J Neurosurg* 1979;50:31–9.
21. Renn WH, Rhoton AL Jr. Microsurgical anatomy of the sellar region. *J Neurosurg* 1975;43:288–98.
22. Hirst LW, Miller NR, Allen GS. Sphenoidal pneumosinus dilatans with bilateral optic nerve meningiomas. *J Neurosurg* 1979;51: 402–7.