Pneumosinus Dilatans of the Sphenoid Sinus

Four cases of pneumosinus dilatans of the sphenoid sinus are reported, supplementing the eight cases previously reported in the literature. This rare entity is characterized by expansion of a paranasal sinus that contains only air. In one patient, severe visual loss due to compression of the optic canal by the adjacent enlarged sinus was seen. Galactorrhea occurred in one patient, and three of the four patients reported headaches. There was dehiscence of the sinus roof in two cases, which apparently resulted in a cerebrospinal fluid fistula in one. Previous reports of this entity are reviewed, and the radiographic findings and clinical presentations are discussed. It is proposed that the term “pneumosinus dilatans” be used to describe all dilated, air-filled sinuses with outwardly bulging walls when the primary cause is uncertain.

Pneumosinus dilatans is a rare disorder of unknown origin characterized by expansion of a paranasal sinus that contains only air. Involvement may range from a single cell to an entire sinus. Since the first report in 1918 by Benjamin [1], 60 cases have appeared in the literature [2–11], only eight of which have involved the sphenoid sinus [2–5]. From 1980 to 1985, we diagnosed four cases of pneumosinus dilatans of the sphenoid sinus. Unilateral blindness occurred in one case secondary to compression of the optic nerve by an enlarged sphenoid air cell. Galactorrhea occurred in one case.

Case Reports

Case 1

A 14-year-old boy noted a gradual painless decrease in visual acuity of the right eye 2 years before presentation. Physical examination disclosed mild right proptosis and profound right optic nerve atrophy. Visual acuity was correctable to 20/15 in the left eye, but the right eye was able to detect only hand motions at 1 ft. There was no history of trauma, surgery, or sinusitis, and the patient was otherwise healthy. Plain radiographs revealed severe stenosis of the right optic canal and an enlarged sphenoid air cell. CT and MRI showed an enlarged, ballooned, right sphenoid air cell obliterating the optic canal (Fig. 1). The MR images also showed the thinned but present optic nerve between the orbital apex and optic chiasm.

Case 2

A 45-year-old man had a history of intermittent sinusitis and headaches. Plain sinus radiographs revealed no evidence of inflammatory disease, but the sinuses were highly aerated, and the sphenoid sinus in particular was markedly enlarged and ballooned (Fig. 2). There was no history of acromegaly or other medical problems.

Case 3

A 26-year-old man had a history of head trauma and excision of a frontal subdural hematoma 9 years before. No skull fracture was noted at the time of the injury. His history
Fig. 1.—Case 1. Enlarged right sphenoid sinus obliterating optic foramen. A, No right optic foramen is visible on oblique view of orbit (arrow). B, Normal left optic foramen (arrow). C, Axial CT shows enlarged right sphenoid sinus (arrow) narrowing superior orbital fissure. D, Coronal CT shows widened sinus obliterating right orbital apex (closed arrow), while left orbital apex is normal (open arrow). E, MR also shows effects of enlarged sinus and thinned but present optic nerve (arrow).

was also remarkable for the presence of chronic sinus headaches and allergic rhinitis. Ophthalmologic examination, including visual-field testing, was normal. Plain sinus radiographs and sinus tomography showed enlargement and ballooning of the sphenoid sinus, as well as a defect in the bony roof (Fig. 3). CT showed the sphenoid sinus enlargement but was otherwise negative.

Case 4

A 31-year-old woman had headaches and galactorrhea. CT and MRI revealed marked enlargement and ballooning of the left sphenoid sinus, expanding into the left infratemporal fossa region. Coronal CT showed a defect in the roof of the left sphenoid sinus (Fig. 4). At surgery, the enlarged sinus was found to contain cerebrospinal fluid. Pathologic examination of the sinus wall disclosed only mild acute and chronic inflammation.

Discussion

The paranasal sinuses are air-filled spaces formed by evagination of the mucous membranes of the nasal cavity into the adjacent facial and skull bones. The walls of the sinuses are composed of compact bone, sinus growth having displaced cancellous bone. The sinuses grow slowly until puberty, after which they grow rapidly to their adult size. In old age, resorption of the diploe leads to further enlargement [12].

The sphenoid sinuses lie side by side within the body of the sphenoid bone, separated by a bony septum. Sphenoid pneumatization may extend to the dorsum sella, the posterior
Pneumosinus dilatans may be diagnosed on standard radiographs, CT, or MR. The diagnostic criteria include: (1) the enlargement of an air cell or an entire sinus, (2) the presence of only air in the abnormal space, and (3) the ballooning outward of the walls of the sinus, which may be thinned and/or demineralized.

Of the 60 previously reported cases of pneumosinus dilatans, the frontal sinuses were involved in 39, the sphenoid sinuses in eight, the maxillary sinuses in seven, and the ethmoid sinuses in six. In his review of the literature in 1967, Lombardi found 51 of these cases, 48 of which occurred in men with ages ranging from 20–40 years [2]. Of the nine cases reported since 1967, the maxillary sinuses were involved in five [7–11], the sphenoid sinuses in three [3–5], and the ethmoid sinuses in one [6], with patients ranging in age from 13–76 years. Five of the nine patients were men.

Impaired visual acuity, bitemporal hemianopsia, and/or optic atrophy occurred in each of the eight previously reported cases of sphenoid pneumosinus dilatans, but severe visual loss had been reported only twice before. One of these cases was a 73-year-old man with a dilated sphenoid sinus who had bitemporal visual field deficits and left optic atrophy (10/70 vision, left eye) [4]. The other was a 76-year-old woman with an enlarged sphenoid sinus that caused left optic atrophy and impaired right visual acuity [3]. In neither case was the optic canal obliterated, and visual loss was believed to be related to optic chiasm compression. The only other symptom associated with sphenoid pneumosinus dilatans was mild hypopituitarism, reported by Lombardi in a single case [2].

In our series of dilated sphenoid sinuses, severe visual loss was found in one patient, secondary to compression of the optic canal. Three of four patients reported headaches. Galactorrhea occurred in one patient.

Case 4 of this series differed from the others because the enlarged sphenoid sinuses contained cerebrospinal fluid rather than air. A dehiscence was found in the roof of the enlarged sinus, and this is believed to be the pathway of the cerebrospinal fluid into the sinus. Lombardi did not describe frank dehiscences of sinus walls in his paper, but dehiscences were found in six of the nine cases of enlarged sinuses reported since 1967. In addition, Kaufman et al. [13], who described five cases of cerebrospinal-fluid fistulas through dehiscences of the floor of the middle-cranial fossa, emphasized that pneumatization of the middle-fossa floor must be present in order to allow fistulas to develop. In none of the cases reported by Kaufman et al., however, was there sinus
dilatation. Besides the dehiscence found in case 4, a smaller defect of the sphenoid-sinus roof was noted in case 3 of this series. Other explanations for the presence of cerebrospinal fluid within the sphenoid sinus seem unlikely. The location is unusual for a meningocele and the patient’s age is atypical.

There is some confusion in the literature regarding the nomenclature of abnormally enlarged sinuses. Several reports have appeared in which abnormally dilated maxillary sinuses have been referred to as pneumoceles [5, 7–11]. In most of these reports, the relationship of this entity to pneumosinus dilatans is not mentioned. Som, however, claimed that thinning or destruction of the sinus walls characterizes a pneumocele as a separate entity from pneumosinus dilatans [5]. This is contradicted by Lombardi, who reported the largest series of pneumosinus dilatans, including many cases in which the sinus walls were ballooned outward, thinned, or demineralized. A cause has not been firmly established for either pneumosinus dilatans or pneumocele, although authors who use the term pneumocele tend to favor a ball-valvelike air-trapping origin. Because reported cases using both terms appear to be radiographically indistinguishable, because symptoms may be identical, and because the origin for neither entity is firmly established, we suggest that the cases formerly described under both names represent variations of the same entity. A single term to describe all dilated, air-filled sinuses of uncertain origin with outwardly bulging walls seems sufficient. We favor the term pneumosinus dilatans over pneumocele because the former is more descriptive and specific.

The term pneumosinus dilatans has also been applied to those cases of thickening and upward bowing of the roof of the sphenoid sinus associated with planum sphenoidal meningiomas [14], to the generalized sinus enlargement seen in acromegaly, and to the sinus enlargement that accompanies long-standing loss of brain tissue as in cerebral hemiatrophy. The last three examples should not be considered cases of pneumosinus dilatans.

The origin of pneumosinus dilatans remains poorly understood; proposed causes include developmental, inflammatory, and obstructive mechanisms. Several authors, who have used the term pneumocele, favor the explanation of a ball-valvelike air-trapping mechanism in which failure of the air pressure within the sinus to rapidly equilibrate with the environment leads to sinus expansion [7–11]. Supporting this theory are two cases in which patients became symptomatic only during a change in altitude, the first suffering pain from a dilated maxillary sinus [8], and the second experiencing blurred vision in the left eye only during airplane flights because of an enlarged sphenoid air cell compressing the optic nerve [5]. This disorder has been successfully treated in some cases by surgically creating a wide communication between the sinus and the nasal cavity [5, 7–11]. This air-trapping theory, however, does not explain why these patients do not accumulate fluid within the involved sinus. Furthermore, in only one reported case has stenosis of a sinus ostium been documented [11]. Chronic inflammation may incite expansile aeration of the involved sinus. This is supported by the findings of chronic inflammatory changes in the few sinuses that have been histologically examined [5, 7]. In some cases, the abnormally dilated sinus may simply represent an idiopathic developmental anomaly. This seems most likely in those cases presenting in early puberty, when the sinuses may normally undergo rapid growth.

In summary, we present four cases of sphenoid pneumosinus dilatans. Unilateral blindness secondary to obliteration of the optic canal, not previously reported, was seen in one case. In one case, cerebrospinal fluid filled the dilated sphenoid sinus, apparently through a dehiscence in the roof of the sinus.

Addendum

Recently, we have diagnosed another case of pneumosinus dilatans of the sphenoid sinus in a 41-year-old man who 10 years earlier had lost all vision in his left eye and has had gradual deterioration of vision in his right eye. Over the past 2 months, his right-eye visual acuity deteriorated from 20/60 to 20/80. Standard radiographs of the orbits, CT, and MR revealed enlarged aerated paranasal sinuses, with extension of the sphenoid sinuses into the lesser wings of the sphenoid bone, resulting in bilateral narrowing of the optic canals. MR showed atrophy of the optic nerves and chiasm.

REFERENCES