Maxillary Pneumosinus Dilatans Presenting With Proptosis: A Case Report and Review of the Literature

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ABSTRACT

BACKGROUND: Pneumosinus dilatans (PSD) is a rare pathological paranasal sinus expansion. This condition is usually symptomatic or cosmetically apparent, requiring surgical intervention. Multiple hypotheses have been postulated as to the cause of this condition; however, the precise cause and pathogenesis remain obscure.

CASE REPORT AND METHODS: An 11-year-old boy presented with right eye bulging and was subsequently found to have PSD of the maxillary sinus. A search was conducted of the PubMed electronic database, using the keywords "pneumosinus dilatans," "pneum(oco)ele," "pneum(oc)ele," "pneum(atoco)ele," and "air cyst." Articles published in English were reviewed.

RESULTS: The literature review identified 29 cases of PSD involving the maxillary sinus. The mean age of presentation was 25 years old. Only the right maxillary sinus was affected in 16 cases, followed by the bilateral sinuses in 7 cases and the left sinus in 6 cases. In 5 cases, all paranasal sinuses, along with the maxillary sinus, were expanded. The most common presenting symptom was facial swelling, which was found in 55% of the cases, followed by proptosis and pain. Computed tomography is the gold standard radiological method for diagnosing PSD.

CONCLUSIONS: Pneumosinus dilatans is a rare condition that is usually symptomatic and requires surgical intervention. The etiology of the disease is attributed to multiple hypotheses, but more studies are needed to explore this condition further.

KEYWORDS: Pneumosinus dilatans, maxillary sinus, pneumocele, paranasal sinuses, proptosis

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Introduction

Abnormal dilatation of the paranasal sinuses is a rare condition that is characterized by hyperpneumatization of 1 or more of the paranasal sinuses. Meyes[1] was first to describe this condition, followed by Benjamins,[2] who named the condition "pneumosinus dilatans" (PSD). Although Urken et al[3] proposed a modern system of classification that is widely used currently, the nomenclature is still controversial.

Pneumosinus dilatans is characterized by a sinus that is abnormally expanded beyond its normal boundaries with normal mucosa and whose bony walls are displaced outwardly to cause facial embossing or intracranial, orbital, or ethmoidal encroachment.[4]

The presentation of this condition varies from asymptomatic patients to nasal obstruction, facial deformities, pain at altitude, or visual changes. Pneumosinus dilatans occurs most frequently in the frontal sinus (63%), followed by the sphenoidal sinus (25%), maxillary sinus (19%), and ethmoidal sinus (18%), and it usually affects a single sinus cavity.[5] The etiology of this condition is poorly understood, and many theories have been hypothesized, including ball-valve mechanism, fibro-osseous dysregulation, gas-forming bacteria, and even genetics.[6,7]

Maxillary involvement was first reported by Noyek and Zizmor[8] as a "pneumocele." To date, 29 cases of maxillary sinus dilatation have been reported in the literature under different terms such as "pneumosinus dilatans," "pneumocele," and "air cyst."

In our article, we present a rare case of maxillary PSD presenting with proptosis and a literature review of maxillary sinus hyperpneumatization.

Case Report

An 11-year-old boy presented to the Rhinology Clinic at King Fahad Medical City with a complaint of right eye bulging for 6 months. In the last few months, he also started to feel a right-sided nasal obstruction and right cheek bulging. He denied any other associated symptoms.

His otolaryngologic examination showed slight swelling of the right cheek in comparison with the left side with right eye proptosis. His nasal endoscopic examination revealed a deviation of the right lateral nasal wall medially toward the septum with a narrow nasal airway and normal mucosa. The patient was referred to the Ophthalmology Clinic for assessment, which confirmed the right eye proptosis with visible right sclera above the superior corneal limbus, normal visual acuity, normal extraocular muscle motion, and normal funduscopic examination findings.

A CT (computed tomography) scan of the paranasal sinuses revealed hyperpneumatization of the right maxillary sinus with

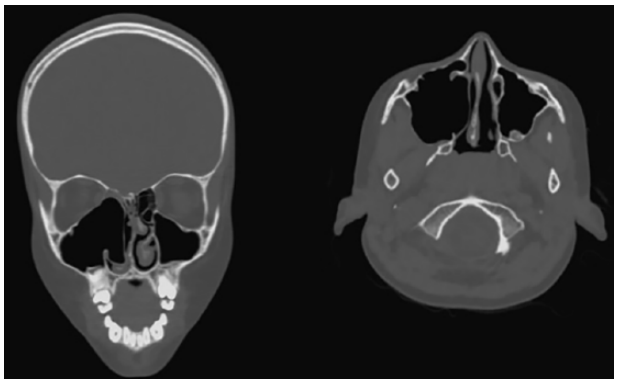

Figure 1. Preoperative frontal (left) and axial (right) CT scan showing hyperpneumatization of the right maxillary sinus with medial expansion causing total occlusion of the left nasal cavity with tapering of the left ethmoid sinuses. The remaining paranasal sinuses and mastoid air cells were well aerated.

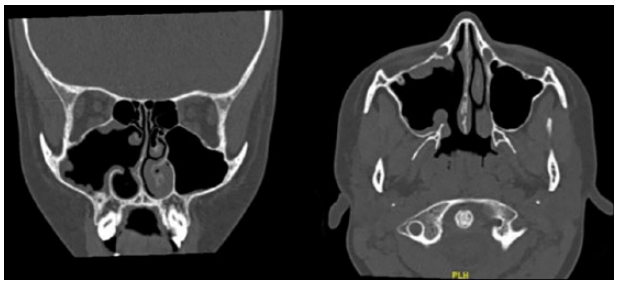

Figure 2. Postoperative frontal and axial CT scans.

medial expansion causing significant narrowing of the nasal airway. No bony erosions or intraorbital pathology were noted (Figures 1 and 2).

The diagnosis of right maxillary PSD was made, and it was decided that the patient should be managed surgically. The patient underwent right functional endoscopic sinus surgery under general anesthesia. The procedure included an uncinectomy, a wide maxillary antrostomy, an anterior ethmoidectomy, and an inferior turbinate turbinoplasty. The postoperative period was uneventful, and no complications were observed.

Follow-up visits after 6 months and 2 years showed significant improvement in the right cheek swelling and right nasal obstruction. Endoscopic examination revealed a patent nasal airway with healthy mucosa.

Discussion

Anatomy and embryology

The maxillary sinus is the largest paranasal sinus with an adult volume of 15 mL. It is the first sinus to develop in utero and undergoes a biphasic pattern of rapid growth: first, from birth to 3 years of life, and then between 7 and 18 years. At birth, the maxillary sinus measures 7 mm in anteroposterior depth, 4 mm in height, and 2.7 mm in width. The maxillary sinus continues pneumatization rapidly between the first and eighth year of age. At 16 years of age, the maxillary sinus usually reaches its adult size, measuring 39 mm in depth, 36 mm in height, and 27 mm in width. The maxillary sinus has a pyramidal shape with an anterior wall corresponding to the facial surface of the maxilla. Its posterior bony wall separates it medially from the pterygomaxillary fossa and laterally from the infratemporal fossa. Its medial wall is formed by the middle meatal mucosa, a layer of connective tissue and the sinus mucosa. The floor of the maxillary sinus is formed by the alveolar process of the maxillary bone and hard palate. The roof of the maxillary sinus corresponds to the floor of the orbit. The maxillary sinus is supplied by the branches of the internal maxillary artery, which include the alveolar, infraorbital, greater palatine, and sphenopalatine arteries. It is innervated by branches of the second division of the trigeminal nerve, the infraorbital nerve, and the greater palatine nerve.

Pathogenesis

Pneumosinus dilatans is a rare condition characterized by benign expansion (pathologic hyperaeration) of 1 or more of the paranasal sinuses beyond its normal margins. As the expansion progresses, destruction of the overlying bone and surrounding structure occurs, leading to varying signs and symptoms. Although the first description of PSD in the literature was by Meyes,[1] the precise etiology and pathogenesis of this condition remain unclear. Several theories have been proposed, including a 1-way valve mechanism, gas-forming microorganisms, mucocele drainage, osteogenic theory, hormonal dysregulation, and genetic predisposition.[9]

The most commonly proposed mechanism and widely accepted theory is a 1-way valve mechanism.[5] In this hypothesis, an obstructive lesion mimics a valve operating at the sinus ostium, leading to the long-term trapping of air inside the affected sinus. The ultimate effect is high intranasal pressure resulting in a bony deformity. In support of this theory is the fact that many patients have reported an increase in symptoms while ascending on an airplane.

A new bone remodeling theory was suggested by Jankowski et al[4] who investigated whether bone remodeling plays a role in PSD. Using fluorine 18-labeled sodium fluoride positron emission tomography-CT (18F-NaF PET-CT) and bone pathological examinations, they found significant 18F-NaF uptake on PET-CT images by the walls affected by PSD, and these changes were correlated pathologically with intense and diffuse bone remodeling, observing that 80% of normal trabecular mineralized bone was replaced by osteoid. This hypothesis also proposes that changes in nitric oxide concentrations after surgical opening for PSD, which has an effect on bone metabolism, might play a role in stopping further sinus expansion. These findings could change our understanding of this condition.

Nomenclature

Abnormal expansion of the paranasal sinuses has been described in the literature using many confusing and poorly defined terms

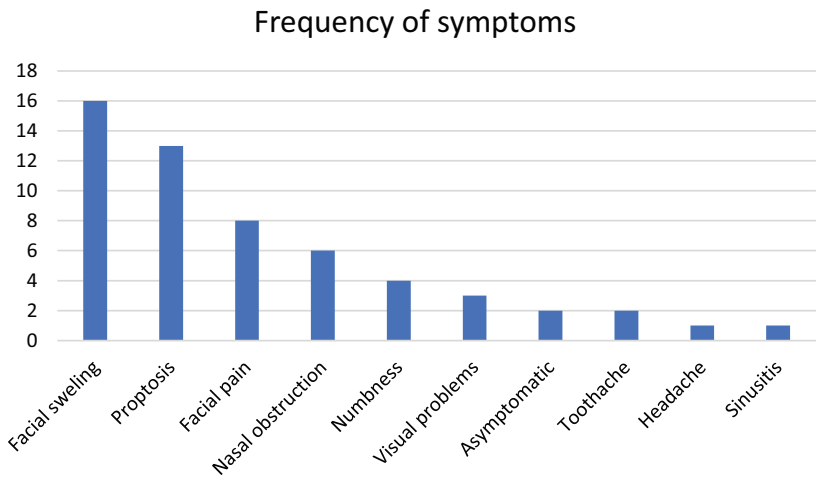

Figure 3. Frequency of different complaints.

(pneumocele, pneumatocele, PSD, and air cyst, among others). However, in 1987, Urken et al[3] adapted the most widely accepted nomenclature for hyperaeration of the paranasal sinuses by performing a review of the literature and comparing his own experiences with the normal anatomy of the sinus. He classified sinus hyperaeration into 3 distinct categories based on the size of the affected sinus and the bony wall integrity:

1. Hypersinus, an aerated sinus that extends beyond the upper limit of the normal anatomic boundaries of the sinus but within the normal range of the affected bone and with normal sinus walls; these patients are clinically asymptomatic.
2. Pneumosinus dilatans, an aerated sinus that extends beyond the normal anatomic boundaries of the sinus and affected bone, with displaced sinus walls and normal bony thickness; these patients may present clinically with some local pressure symptoms.
3. Pneumocele, an aerated sinus that extends beyond the normal anatomic boundaries of the sinus, with displaced sinus walls and focal or generalized thinning of the bony sinus wall; these patients may present clinically with symptoms similar to PSD.

In addition, PSD affecting all paranasal sinuses as well as the mastoid cells has been described as PSD multiplex.[7,10,11]

In our literature review, 29 cases involving the maxillary sinus were identified, 19 cases of which were described as PSD, 7 as pneumocele, 2 as air cyst, and 1 as PSD multiplex (Table 1).

Clinical presentation

The mean age of presentation was 25 years old (range, 9-62 years). Males were affected more commonly, with 18 male patients and 11 female patients reported in the literature. The right sinus was more commonly affected (16 cases), followed by bilateral involvement (7 cases) and left sinus only (6 cases). In 22 of the included

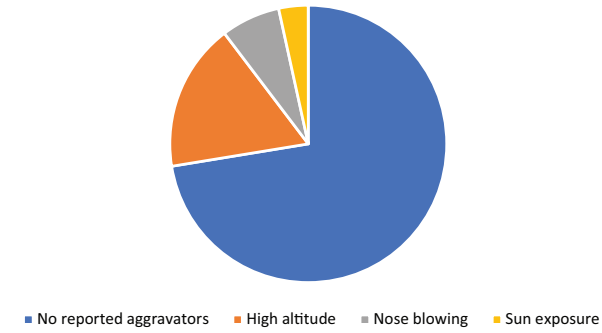

Figure 4. Symptom aggravators.

cases in our review, the maxillary sinus was the only sinus affected, while all paranasal sinuses were affected in 5 cases.

The most common presenting symptom was facial swelling/masses/deformities (Figure 3), which was found in 55% of the cases, followed by proptosis in 45% of the cases and facial pain in 28% of the cases. Only 7% of the patients were asymptomatic and found incidentally during visits for other reasons.

Five patients reported symptoms associated with changes in altitude or during air flights (Figure 4), which might support the 1-way valve mechanism theory as a pathological cause of this condition.

Radiological features

Computed tomography is the main radiological modality required for diagnosing this condition. The main feature of PSD is expansion of the sinus beyond the normal anatomical limits with or without associated cortical bone thinning. In our literature review, the expanded sinus was associated with bony wall thinning in 39% of the cases. Bony wall erosions were seen in some of the severe cases. Magnetic resonance imaging (MRI) can be used to exclude some other differential diagnoses or associated conditions. Based on Jankowski et al's findings, 18F-NaF PET-CT might be useful in cases where the diagnosis is challenging.

Table 1. Review of the studies reporting maxillary PSD.

STUDY	AGE SEX	REPORTED AS	LOCATION	ASSOCIATED SINUSES	PRESENTATION	RADIOLOGICAL FINDINGS	MANAGEMENT	ASSOCIATED CONDITION
Al-Essa et al[12]	47 F	PSD	R	No	Proptosis	CT: Superior bowing of the right orbital floor	No	No
Jankowski et al[4]	47 M	PSD	R	No	Toothache (Atm P)	CT: Large R Max sinus/ walls displaced	FESS	No
Doucette-Preville et al[13]	17 M	Air cyst	L	No	Nasal obstruction/facial pressure/facial protrusion/ eye deviation	CT: Large L Max sinus/bony thinning	FESS	No
Ushas et al[7]	15 M	PSD multiplex	Bi	All + mastoid	Asymptomatic	CT: Osteolysis and large all + air cells	No	No
Hyun et al[14]	13 M	PSD	R	No	Facial deformity	CT: Large R Max sinus/ displacement	FESS	No
Teh et al[15]	18 M	PSD	R	No	Facial pain/periorbital swelling/nasal obstruction	CT: Large R Max sinus/ bony thinning + erosion	FESS	No
Choi et al[16]	19 F	PSD	L	No	Cheek swelling with proptosis	CT: Large Max sinus	Intraoral approach (antral wall turnover)	No
	20 M	PSD	R	No	Cheek swelling with proptosis	CT: Large Max sinus	Subciliary approach (antral wall turnover)	No
	22 F	PSD	R	No	Cheek swelling	CT: Large Max sinus/bony thinning	Intraoral approach (antral wall turnover)	No
	20 M	PSD	Bi	No	Cheek swelling	CT: Large Max sinus	Intraoral approach (greenstick downward fracture)	No
Finsterer et al[17]	43 M	PSD	Bi	All	Asymptomatic	CT: Large all sinuses/bony thinning	No	MD1
Braverman[18]	14 F	Pneumocele	R	No	Facial pressure/rhinitis/ numbness/headache/nasal obstruction/exophthalmos (Atm P)	CT: Large R Max sinus/ bony thinning	FESS	No
Vlckova and White[19]	33 M	PSD	L	No	Cheek paresthesia/ toothache/facial asymmetry/ nasal obstruction/ exophthalmos	CT: Large L Max sinus/bony erosion	FESS	No
Viehweg and Hudson[20]	27 F	PSD	Bi	Sph	Cheek swelling	CT: Large Bi Max sinuses/ cyst in L Max sinus/bony thinning	FESS	No
Sanjari et al[21]	13 F	PSD	Bi	All	Diminished vision	MRI: Large all sinuses	No	Sickle cell trait

Table 1. (Continued)

STUDY	AGE SEX	REPORTED AS	LOCATION	ASSOCIATED SINUSES	PRESENTATION	RADIOLOGICAL FINDINGS	MANAGEMENT	ASSOCIATED CONDITION
Knapp and Klenzner[22]	49 F	Pneumocele	R	No	Facial numbness/cheek pressure (Atm P)	CT: Large R Max sinus/ bony thinning + defect	Microscopic endonasal surgery	No
Karlidağ et al[23]	33 M	PSD	Bi	No	Facial mass/facial pain (Atm P)	CT: Large Bi Max sinuses	Intraoral approach	No
Juhl et al[24]	40 M	PSD	R	No	Exophthalmos/cheek swelling/facial pain/ paresthesia (nose blowing + Atm P)	CT/MRI: Large R Max sinus/ bony thinning	FESS	No
Mauri et al[25]	17 M	PSD	L	No	Facial swelling/nasal obstruction/pain + mass (sun exposure)	CT: Large L Max sinus/ displaced walls	FESS	No
Dillard and Sillers[26]	26 M	Pneumocele	R	No	Orbital displacement	CT: Large R Max sinus/ displaced walls/zygoma thinning	FESS	No
Flanary and Flanary[27]	9 M	Pneumocele	L	No	Exophthalmos	CT: Large Max sinus/bony erosion	FESS	No
Breidahl et al[28]	12 F	PSD	R	No	Cheek swelling	CT: Large R Max sinus/ bony thinning	Intraoral approach (antral wall turnover)	No
	42 M	PSD	R	All	Cheek mass	CT: Large all sinuses	Intraoral approach with lateral maxillary bone graft harvest	No
Stretch and Poole[29]	17 M	PSD	Bi	All	Diplopia/diminished vision/ angiomatous nevus	CT: Large all sinuses	No	Melnick-Needles Syndrome
Tovi et al[30]	20 M	Air cyst	L	No	Cheek mass	CT: Large L Max sinus/bony thinning	Caldwell-Luc procedure	No
Dhillon and Williams[31]	16 F	PSD	R	No	Exophthalmos/diminished vision	X-ray: Large Max sinus/ bone formation	Caldwell-Luc procedure	Fibro-osseous lesion and meningiomata
Wolfensberger and Herrmann[32]	15 M	Pneumocele	R	Ethm	Exophthalmos/cheek pain + pressure (nose blowing)	X-ray and CT: Large R Max sinus/bony thinning	FESS	No
Vines et al[33]	62 F	Pneumocele	R	No	Proptosis	X-ray: Large R Max sinus/ bony erosion	Caldwell-Luc procedure	No
Zizmor et al[34]	13 F	Pneumocele	R	No	Facial swelling/exophthalmos/nasal obstruction	X-ray: Large R Max sinus/ bony thinning + erosion	Caldwell-Luc procedure	No

Abbreviations: Atm P, atmospheric pressure; Bi, bilateral; CT, computed tomography; Ethm, ethmoid; F, female; FESS, functional endoscopic sinus surgery; Front, frontal; L, left; M, male; Max, maxillary; MD1, myotonic dystrophy type 1; MRI, magnetic resonance imaging; PSD, pneumosinus dilatans; R, right; Sph, sphenoid.

Management

The aim of treating PSD is to relieve the symptoms and correct the facial deformities. Symptomatic patients require surgical intervention. The Caldwell-Luc approach and the creation of a nasoantral window were commonly employed because they were the standard techniques used for maxillary sinus pathologies at the time of the earlier reports. Currently, maxillary localization can be achieved with minimally invasive techniques, showing less morbidity and a shortened hospitalization by creating a maxillary antrostomy via endoscopic techniques. However, although a nasoantral window may relieve symptoms (if present), deformities may still persist.[14,20,32] Hyun et al[14] reported good cosmetic results after reduction osteoplasty. Antral wall turnover, greenstick downward fracture, and electrical burring were proposed by Choi et al[16] to correct facial deformities based on the thickness of the antral wall and the extension of the expanded area.

Conclusions

Pneumosinus dilatans is a rare condition that is usually symptomatic and requires surgical intervention, primarily for cosmetic reasons. The etiology of the disease is attributed to multiple hypotheses, but more studies are needed to explore this condition further.

Author Contributions

Wrote first draft of manuscript: AA, YA, FA. Principal investigator: AA. Organized the references: AA, MA. Contributed to writing and reviewing manuscript: AA, YA, MA, FA. Made the reviewers' changes: AA, YA. All authors reviewed and approved of the final manuscript.

Informed Consent

Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

REFERENCES

1. Meyes J. Pneumosinus dilatans. *Nederlands Tijdschr Geneeskd*. 1898;11:143–148.
2. Benjamins C. Pneumo-sinus frontalis dilatans. *Acta Oto-Laryngol*. 1918;1:412–422.
3. Urken ML, Som PM, Lawson W, Edelstein D, Weber AL, Biller HF. Abnormally large frontal sinus. II. Nomenclature, pathology, and symptoms. *Laryngoscope*. 1987;97:606–611.
4. Jankowski R, Kuntzler S, Boulanger N, et al. Is pneumosinus dilatans an osteogenic disease that mimics the formation of a paranasal sinus? *Surg Radiol Anat*. 2014;36:429–437.
5. Ricci JA. Pneumosinus dilatans: over 100 years without an etiology. *J Oral Maxillofac Surg*. 2017;75:1519–1526.
6. Skolnick CA, Mafee MF, Goodwin JA. Pneumosinus dilatans of the sphenoid sinus presenting with visual loss. *J Neuroophthalmol*. 2000;20:259–263.
7. Ushas P, Ravi V, Painatt JM, Nair PP. Pneumosinus dilatans multiplex associated with hormonal imbalance. *BMJ Case Rep*. 2013;2013:010345.
8. Noyek AM, Zizmor J. Pneumocele of the maxillary sinus. *Arch Otolaryngol*. 1974;100:155–156.
9. Bouguila J, Ben Rejeb M, Omezzine M, Mani R, Khochtali H. Pneumosinus dilatans: rare cause of slowly changing frontal contours. *Aesthet Surg J*. 2015;35:NP47–NP53.
10. Hwang K, Lee DK, Lee CJ, Lee SI. Pneumosinus dilatans multiplex, mental retardation, and facial deformity. *J Craniofac Surg*. 2000;11:487–490.
11. Kiroglu Y, Karabulut N, Sabir NA, Yagci AB, Gakmak V, Ozguler U. Pneumosinus dilatans and multiplex: report of three rare cases and review of the literature. *Dentomaxillofac Radiol*. 2007;36:298–303.
12. Al-Essa RS, Alsaleh SA, Alsuhaibani AH. Non-axial proptosis secondary to pneumosinus dilatans of the maxillary sinus. *Saudi J Ophthalmol*. 2018;32:238–240.
13. Doucette-Preville S, Tamm A, Khetani J, Wright E, Emery D. Maxillary air cyst. *J Radiol Case Rep*. 2013;7:10–15.
14. Hyun SM, Min JY, Jang YJ. Reduction osteoplasty for treating pneumosinus dilatans of the maxillary sinus. *J Laryngol Otol*. 2013;127:207–210.
15. Teh BM, Hall C, Chan SW. Pneumosinus dilatans, pneumocoele or air cyst? A case report and literature review. *J Laryngol Otol*. 2012;126:88–93.
16. Choi EC, Shin HS, Nam SM, Park ES, Kim YB. Surgical correction of pneumosinus dilatans of maxillary sinus. *J Craniofac Surg*. 2011;22:978–981.
17. Finsterer J, Stollberger C, Molzer G, Prager E. Pneumosinus dilatans and hypercalcification of the falx and ligamentum petroclinoideum in myotonic dystrophy 1. *Neurologist*. 2010;16:125–128.
18. Braverman I. Pneumocele of the maxillary sinus with orbital and trigeminal nerve involvement: case report and review of the literature. *J Otolaryngol Head Neck Surg*. 2009;38:E35–E38.
19. Vlckova I, White PS. Rapidly expanding maxillary pneumosinus dilatans. *Rhinology*. 2007;45:93–95.
20. Viehweg TL, Hudson JW. Pneumosinus dilitans of the maxillary sinuses, bilaterally: a case report. *J Oral Maxillofac Surg*. 2006;64:726–730.
21. Sanjari MS, Modarreszadeh M, Tarassoly K. Pneumosinus dilatans in a 13 year old female. *Br J Ophthalmol*. 2005;89:1537–1538.
22. Knapp FB, Klenzner T. Pneumocele as a rare differential diagnosis in trigeminal irritation. *Am J Otolaryngol*. 2003;24:236–238.
23. Karlidağ T, Yalcin S, Kaygusuz I, Demirbag E. Bilateral pneumosinus dilatans of the maxillary sinuses. *Br J Oral Maxillofac Surg*. 2003;41:122–123.
24. Juhl HJ, Buchwald C, Bollinger B. An extensive maxillary pneumosinus dilatans. *Rhinology*. 2001;39:236–238.
25. Mauri M, de Oliveira CO, Franche G. Pneumosinus dilatans of the maxillary sinus. Case report. *Ann Otol Rhinol Laryngol*. 2000;109:278–280.
26. Dillard ML, Sillers MJ. Maxillary sinus pneumocele causing orbital displacement. *Am J Otolaryngol*. 1999;20:250–251.
27. Flanary CJ, Flanary VA. Maxillary sinus pneumocele. *Otolaryngol Head Neck Surg*. 1998;119:518–520.
28. Breidahl AF, Szwajkun P, Chen YR. Pneumosinus dilatans of the maxillary sinus: a report of two cases. *Br J Plast Surg*. 1997;50:33–39.
29. Stretch JR, Poole MD. Pneumosinus dilatans as the aetiology of progressive bilateral blindness. *Br J Plast Surg*. 1992;45:469–473.
30. Tovi F, Gatot A, Fliss DM. Air cyst of the maxillary sinus (pneumosinus dilatans, pneumocoele). *J Laryngol Otol*. 1991;105:673–675.
31. Dhillon RS, Williams DC. Pneumosinus dilatans. *J Laryngol Otol*. 1987;101:828–832.
32. Wolfensberger M, Herrmann P. The pathogenesis of maxillary sinus pneumoceles. *Arch Otolaryngol Head Neck Surg*. 1987;113:184–186.
33. Vines FS, Bonstelle CT, Floyd HL. Proptosis secondary to pneumocele of the maxillary sinus. *Neuroradiology*. 1976;11:57–59.
34. Zizmor J, Bryce M, Schaffer SL, Noyek AM. Pneumocele of the maxillary sinus. A second case report. *Arch Otolaryngol*. 1975;101:387–388.