Introduction

Abnormal dilatation of the paranasal sinuses is a rare condition that is characterized by hyperpneumatization of 1 or more of the paranasal sinuses. Meyers\(^1\) 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 fundus examination findings.

A CT (computed tomography) scan of the paranasal sinuses revealed hyperpneumatization of the right maxillary sinus with...
Clinical Medicine Insights: Ear, Nose and Throat

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 uncinctomy, 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 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, 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.

The most commonly proposed mechanism and widely accepted theory is a 1-way valve mechanism. 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 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.
(pneumocele, pneumatocele, PSD, and air cyst, among others). However, in 1987, Urken et al. 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. 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 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.
| STUDY                        | AGE | SEX | REPORTED AS | LOCATION | ASSOCIATED SINUSES | PRESENTATION | RADIOLOGICAL FINDINGS | MANAGEMENT | ASSOCIATED CONDITION |
|------------------------------|-----|-----|-------------|----------|--------------------|--------------|----------------------|------------|---------------------|
| Al-Essa et al12              | 47  | F   | PSD         | R        | No                 | Proptosis    | CT: Superior bowing of the right orbital floor | No         | No                  |
| Jankowski et al21            | 47  | M   | PSD         | R        | No                 | Toothache    | CT: Large R Max sinus/walls displaced         | FESS       | No                  |
| Doucette-Preville et al13    | 17  | M   | Air cyst    | L        | No                 | Nasal obstr/ facial pressure/facial protrusion/eye deviation | CT: Large L M sinuses/bony thinning | FESS       | No                  |
| Ushas et al7                 | 15  | M   | PSD multiplex | Bi       | All + mastoid     | Asymptomatic | CT: Osteolysis and large all + air cells | No         | No                  |
| Hyun et al14                 | 13  | M   | PSD         | R        | No                 | Facial deformity | CT: Large R M sinuses/displacement         | FESS       | No                  |
| Teh et al15                  | 18  | M   | PSD         | R        | No                 | Facial pain/periorbital swelling/nasal obstr | CT: Large R M sinuses/bony thinning + erosion | FESS       | No                  |
| Choi et al16                 | 19  | F   | PSD         | L        | No                 | Cheek swelling w/ proptosis | CT: Large Max sinus | Intraoral approach (antral wall turnover) | No         |                     |
|                              | 20  | M   | PSD         | R        | No                 | Cheek swelling w/ 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 al17            | 43  | M   | PSD         | Bi       | All                | Asymptomatic | CT: Large all sinuses/bony thinning | No         | MD1                 |
| Braverman18                  | 14  | F   | Pneumocele   | R        | No                 | Facial pressure/rhinitis/numbness/headache/nasal obstr/exophthalmos (Atm P) | CT: Large R M sinuses/bony thinning | FESS       | No                  |
| Vlckova and White19          | 33  | M   | PSD         | L        | No                 | Cheek paraesthesia/facial asymmetry/nasal obstruction/exophthalmos | CT: Large L Max sinuses/bony erosion | FESS       | No                  |
| Viehweg and Hudson20         | 27  | F   | PSD         | Bi       | Sph                | Cheek swelling | CT: Large Bi Max sinuses/oyst in L Max sinus/bony thinning | FESS       | No                  |
| Sanjari et al21              | 13  | F   | PSD         | Bi       | All                | Diminished vision | MRI: Large all sinuses | No         | Sickle cell trait  |
### Table 1. (Continued)

| STUDY | AGE | SEX | REPORTED AS | ASSOCIATED CONDITION | LOCATION | RADIODLOGICAL FINDINGS | MANAGEMENT |
|-------|-----|-----|-------------|----------------------|----------|------------------------|------------|
| Alatar et al | 15 | M | Pneumocele | FESS | CT: Large R Max sinus/ bone thinning | No |
| Knapp and Klenzner | 16 | M | Pneumocele | FESS | CT: Large R Max sinus/ bone thinning | No |
| Karlida et al | 17 | M | Pneumocele | FESS | CT: Large R Max sinus/ bone thinning | No |
| Juhl et al | 18 | M | Pneumocele | FESS | CT: Large R Max sinus/ bone thinning | No |
| Mauri et al | 19 | M | Pneumocele | FESS | CT: Large R Max sinus/ bone thinning | No |
| Dillard and Sillers | 20 | M | Pneumocele | FESS | CT: Large R Max sinus/ bone thinning | No |
| Flanary and Flanary | 21 | M | Pneumocele | FESS | CT: Large R Max sinus/ bone thinning | No |
| Breidahl et al | 22 | M | Pneumocele | FESS | CT: Large R Max sinus/ bone thinning | No |
| Stretch and Poole | 23 | M | Pneumocele | FESS | CT: Large R Max sinus/ bone thinning | No |
| Vines et al | 24 | M | Pneumocele | FESS | CT: Large R Max sinus/ bone thinning | No |
| Zitzer et al | 25 | M | Pneumocele | FESS | CT: Large R Max sinus/ bone thinning | No |

### Abbreviations:
- Atm P: atmospheric pressure
- Bi: bilateral
- CT: computed tomography
- Ethm: ethmoid
- 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
- Sun exposure: sun exposure
- Nose blowing +: nose blowing +
- X-ray: x-ray
- X-ray and CT: x-ray and computed tomography
- X-ray and CT and FESS: x-ray, computed tomography, and functional endoscopic sinus surgery
- X-ray, CT, and FESS: x-ray, computed tomography, and functional endoscopic sinus surgery
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. Hyun et al reported good cosmetic results after reduction osteoplasty. Antral 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.

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