# PNEUMOSINUS DILATANS – A RARE DISORDER OF PARANASAL SINUSES

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#### **ABSTRACT**

Pneumosinus dilatans is a very rare disorder of the paranasal sinuses, most commonly affecting the frontal sinuses (Pneumosinus dilatans frontalis). So far only 134 cases are reported in the literature. The exact etiology of this disorder is unknown. The pathology causes abnormal dilatation of paranasal sinuses. Most patients are asymptomatic while others can have clinical features of frontal bossing, head ache, visual defects etc. The diagnosis is by computed tomographic imaging. We here report a case of pneumosinus dilatans frontalis in a 22 year old male patient presented with nasal obstruction.

KEY WORDS: Pneumosinus, Frontal Sinus, Paranasal Sinuses, Frontal Bossing, Paranasal Sinus Imaging.

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#### **INTRODUCTION**

Pneumosinus dilatans is a rare disorder of paranasal sinuses with unknown etiology. The disease is characterized by pathological expansion of paranasal sinuses which contains only air and lined with normal sinus mucosa. This can involve only a single sinus or multiple sinuses. Meyes first described about this disease in 1898 and labelled it pneumatocoele, but it was Benjamin who assigned the name Pneumosinus dilatans for this disorder in 1918 [1,2]. The disease most frequently affects the frontal sinuses, followed by sphenoid sinuses, maxillary sinuses and ethmoidal cells [2-4].

Previously, various terms have been used for this

pathology, like pneumosinus frontalis, pneumocoele, pneumatocoele, aerocoele, sinus hypertrophy, hyper-pneumatisation, sinus ectasia and sinus blistering etc. In 1987, Urken et al suggested a classification system based on radiological enlargement, wall erosion and clinical symptoms [5]. He classified them into hypersinus, pneumosinus dilatans and pneumocoele. Hypersinus is an asymptomatic aerated paranasal sinus with normal walls confined to the normal boundaries. Pneumosinus dilatans is symptomatic aerated sinus which expands beyond the normal boundaries with normal and intact walls. Pneuomecoele is an aerated sinus with thinning off or a defect in the wall of sinus

and/or loss of integrity of bone. When the dilatation involves multiple sinuses and mastoid air cells also, the condition is termed pneumosinus dilatans multiplex [1].

As mentioned earlier, frontal is the most commonly involved sinus. A total of 134 cases of pneumosinus frontalis has been described in literature in 117 articles[6]. Here we are presenting a case of pneumosinus dilatans frontalis in a 22-year-old male to add some contribution to literature because of the rarity of this condition.

#### **CASE REPORT**

A 22-year-old male patient presented to department of ENT with right side nasal obstruction for 2 years. The obstruction was insidious in onset and gradually progressive in nature, usually persisting throughout the day. The obstruction aggravates following episodes of cold or upper respiratory tract infections. He also gives history of intermittent nasal discharge which was non-foul smelling and non-blood tinged. There was no history of head ache, fever or facial pain. On anterior rhinoscopic examination, there was right side nasal septal deviation and left inferior turbinate hypertrophy. Diagnostic nasal endoscopy was performed which showed a deviated nasal septum with sharp septal spur on right side at the nasal valve area. Computed tomography (CT) of paranasal sinuses confirmed the diagnosis and also revealed abnormally dilated bilateral frontal sinuses almost extending up to parietal bones (Figure -1). Bilateral extensive pneumatization of sphenoid sinus was also noted involving both pterygoid bones (lateral recess of sphenoid). There were no signs of pathologies inside any sinuses.

After necessary workups, septoplasty under general anesthesia was done for the patient. The surgery and post-operative periods were uneventful and the patient was discharged on 2<sup>nd</sup> post op day with oral antibiotics, analgesics, antihistamines for 5 days. Follow up at 1 week, 3 months and 6 months were successful and the patient was symptom free.

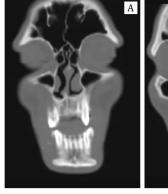
#### **DISCUSSION**

Pneumosinus dilatans is often an incidental radiologic finding. Most of the time the patients are asymptomatic. Symptomatic presentation includes cosmetic deformity, facial asymmetry, head ache, ocular and visual problems etc [6,7]. The most common symptomatic presentation is frontal bossing and prominence of supraorbital ridge [7].

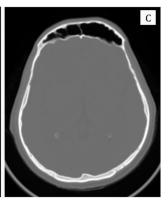
The etiopathogenesis of pneumosinus dilatans is unknown. In general, they are classified into primary (idiopathic) and secondary types. Various hypothesis has been postulated for etiology of primary pneumosinus dilatans. These includes hormonal imbalance, infection with gas forming organisms, spontaneously discharging mucocele etc. The most accepted theory is a one-way (ball) valve effect supported by presence of polypoid mucus in drainage pathway of affected sinuses [1]. Recently in 2014, Jankowsky et al suggested osteogenic diseases as the etiology of primary pneumosinus dilatans. [8].

The secondary types are syndromic, due to compensatory over development of sinuses in association with agenesis of brain (e.g. as in cranio-cerebral hemiatrophy [9]) or due to reduced intracranial pressure [10].

Fig. 1: Radiological Images of Pneumosinus Dilatans – (A) Coronal cuts showing hyperpneumatisation of Frontal sinuses (B) –Coronal cut showing pneumatisation extending laterally above the orbits involving the parietal bones (C) – Axial cut showing bulging left frontal sinus.







Few cases are reported in association with Melnick Needles Syndrome [11], Klippel-Trenaunay-Weber syndrome [12], mental retardation, facial deformity and arachnoid cysts [1,13].

Pneumosinus dilatans is diagnosed with standard radiography or CT Scan of nose and paranasal sinuses. The CT findings suggestive of pneumosinus dilatans are - enlarged sinus, thinning of the sinus wall, hyperaeration, hyperlucency, bony dehiscence, expansion of recesses, effacement of the ethmomaxillary angle and displacement of the middle meatus[1]. Afroze et al in their report labelled the characteristic radiological finding as as a deer horn pattern when the pneumatization extends to the parietal bones [14]. Magnetic resonance imaging (MRI) and Nasal endoscopic examination ate needed to exclude concomitant intracranial lesions like meningocele, meningoencephalocele, CSF rhinorrhea and chronic rhinosinusitis.

No treatment is needed in asymptomatic patients of pneumosinus. In symptomatic patients of frontal pneumosinus dilatans, surgical correction with a craniofacial repair of malformation is recommended [1,13,15]. Several authors have published different surgical techniques and approaches. The most commonly performed procedure is creation of a nasoantral window, to equilibrate the intrasinus pressure [7]. Literature has no reports on recurrence in treated lesions.

# **CONCLUSION**

Pneumosinus dilatans is a rare disorder of paranasal sinuses which every radiologist, oto-rhino-laryngologists, maxillofacial and plastic surgeon needs to be aware of. The etiology of pneumosinus dilatans remains unclear. Imaging studies are needed for the diagnosis and for differentiating between pneumosinus dilatans and pneumocoeles. Management of symptomatic patients is by surgical exploration of the sinus and recontouring of the anterior table of the frontal sinus.

# Conflicts of Interests: None

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