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Pneumosinus dilatans of the frontal sinuses: two cases and a discussion of its aetiology

J. L. Walker, B.Med.Sci., B.M.B.S., N. S. Jones, M.D., F.R.C.S.

Abstract
Pneumosinus dilatans is a rare condition with the dilatation of aerated paranasal sinuses. We present two cases and discuss its aetiology, the radiological classification of enlarged aerated sinuses, and its treatment. One much quoted hypothesis for the cause of this condition has been that it is due to a 'ball valve' effect of mucosal closure at the frontal recess. However, one of our subjects has started, and done, a considerable amount of subaqua diving since presenting with their condition and has had no symptoms on diving, or progression of their frontal swelling. This observation, along with the fact that retained secretions are not seen within the sinuses in this condition, raises doubt about the theory that a one-way valve is responsible.

Key words: Paranasal Sinuses; Pneumosinus Dilatans; Radiology; Aetiology

Introduction

Pneumosinus dilatans is a rare condition affecting the paranasal sinuses. It is characterized by the benign expansion of an aerated sinus beyond the normal margin of the frontal bone.[1] The expansion may involve either all, or part of, the sinus. The precise aetiology and pathogenesis of pneumosinus dilatans is unknown although a large number of mechanisms have been proposed.[2–8] These are discussed together with the radiological classification of aerated sinuses, and methods of treatment. Two cases of pneumosinus dilatans of the frontal sinuses are presented and discussed.

Case reports

Case 1

A 33-year-old lady first notied a change in her appearance following comments by her family whilst looking through the family photo album. They noticed she had developed a more prominent forehead. She was otherwise asymptomatic. On examination she had a mild diffuse bilateral bossing of her forehead (Figure 1). Endoscopic examination of her nose was normal. Her frontal recess looked healthy with no evidence of prominent agger-nasi air cells. A computed tomography (CT) scan showed a diffuse enlargement of both frontal sinuses that were clear (Figure 2a and b). She has been followed up for over six years with no discernible change in her appearance and remains asymptomatic.

Case 2

The second case is a 19-year-old male who gradually noticed a bony swelling above his left eye that had become more prominent over the preceding year. At presentation he complained of rhinorrhoea with intermittent nasal obstruction and bouts of sneezing, all of which were eased with topical nasal steroids.

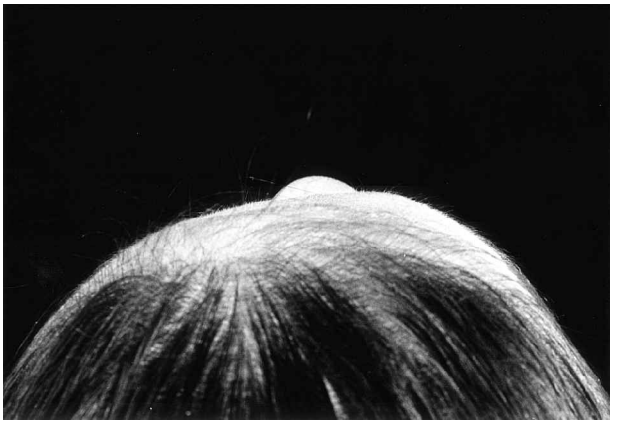

Fig. 1
Aerial view of the frontal sinuses showing bossing.

On examination a smooth bony fullness was noted on his left forehead (Figure 3). Endoscopy revealed bilateral oedematous mucosa and his nasal airway contained clear mucus. Skin prick tests were positive and allergic rhinitis was diagnosed. This was treated medically and a mucocele of the left frontal sinus was suspected. A CT scan showed diffuse enlargement of the left frontal sinus and some minor soft tissue thickening in the maxillary antrum. The frontal and ethmoid sinuses appeared clear with enlargement of the left frontal sinus. A diagnosis of pneuosinus dilatans of the left frontal sinus was made. Follow up at three years revealed no change. Since his original presentation he has since started subaqua diving and done a considerable amount of diving without any symptoms during or after diving.

From the Department of Otorhinolaryngology, Head and Neck Surgery, Queens Medical Centre, University Hospital, Nottingham, UK.
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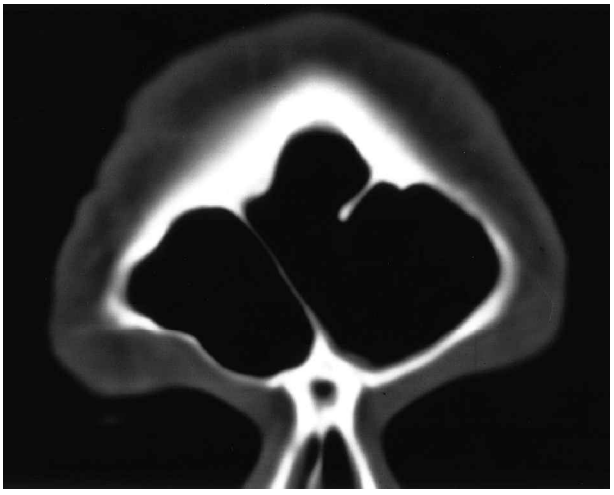
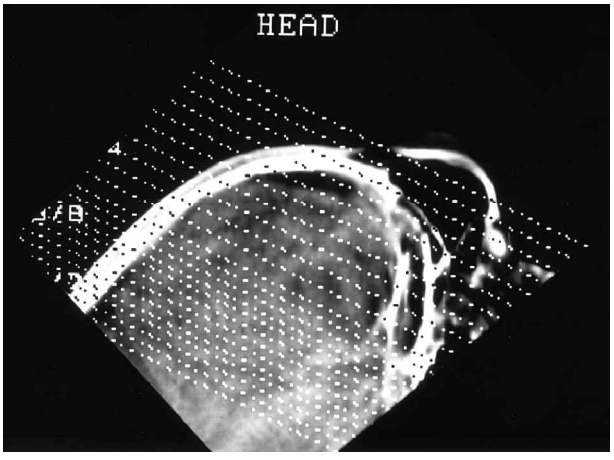


(a)



(b)

FIG. 2

(a) A coronal CT showing an expanded frontal sinus.
(b) A sagittal CT view showing the frontal sinus in the same patient.

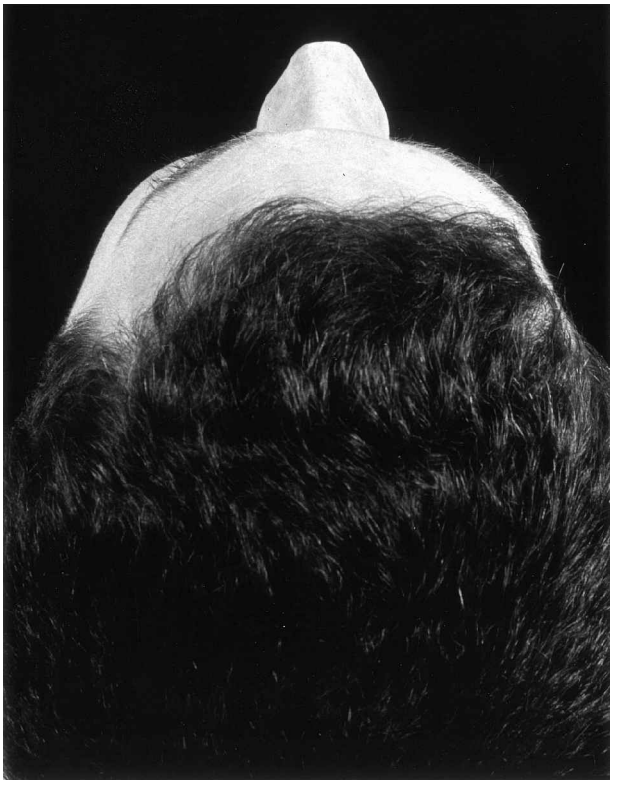


FIG. 3

Aerial view showing expansion of the left frontal sinus.

Discussion

Pneumosinus dilatans is a rare condition. First described by Benjamin[1] it refers to a benign expansion of an aerated paranasal sinus. The sinus walls are always intact and of normal thickness and are outwardly displaced beyond the normal boundaries of the frontal bone.[9] Squamous epithelium lining the sinus appears macroscopically normal and its either histologically normal or shows changes of low-grade inflammation.

The frontal sinuses are the most frequently affected of the paranasal sinuses representing about 65 per cent of reported cases followed in order of decreasing frequency by the sphenoid, maxillary and ethmoid sinuses.[10] Pneumosinus dilatans is often bilateral. Currently there is no consensus about the aetiological factors that influence the development of normal frontal sinuses and that control the normal cessation of sinus growth. Many different factors have been implicated,[11] teeth development, the mechanical stresses of mastication, and growth hormones.[12] Normal variations in frontal sinus development do occur. A degree of hypoplasia is more common but overgrowth of sinuses has been described. It has been proposed that there is a continuum of excessive growth of aerated sinuses defined both clinically and radiologically by CT. These are, in order of progressive growth: a hypersinus, pneumosinus dilatans and pneumocele. A hypersinus is an enlarged sinus with normal walls that do not extend beyond the normal boundaries of the frontal bone.

Pneumosinus dilatans, like a pneumocele, has walls which are expanded either focally or diffusely beyond the margin of the frontal bone. A pneumocele, unlike pneumosinus dilatans however, has walls with either generalized or focal thinning with total or partial loss of its integrity.[5] This classification may be an oversimplification as many cases of pneuosinus dilatans and pneumocele are focal[14] and there are no documented cases in the literature of pneumosinus dilatans progressing to a pneumocele. Most writers use the term pneumosinus dilatans and pneumocele interchangeably as the conditions are difficult to differentiate radiographically.[5]

A review of the literature reveals that the age of presentation of pneumosinus dilatans varies from puberty to the elderly, with most presenting between the ages of 20–40 with an overwhelming preference for males.[10,13] There are no recorded cases of frontal sinus pneumosinus dilatans in children because the frontal sinuses grow slowly until puberty after which they rapidly grow to adult size.[14] It is possible that in cases occurring in early puberty this disorder may represent an idiopathic developmental anomaly.[15] The deformity has a fluctuating course: it may be slowly progressive[16] or may develop suddenly following a refractory period.[13] Most cases of pneumosinus dilatans are asymptomatic and are only diagnosed when a deformity is noticed such as frontal bossing or when the mass effect due to involvement of surrounding structures leads to symptoms.[13,17] Pneumosinus dilatans of the paranasal sinuses may extend to encroach intracranially or on the ethmoid bone, nose or orbit.[16,18,19]

Depending on the site and rate of expansion and the vulnerability of the surrounding structures this may result in diplopia, reduced visual acuity, headache or other local

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pressure symptoms.[2] The disease may be either diffuse or focal with involvement ranging from a single cell to the whole of the sinus. The lateral recess is the region most commonly expanded in focal frontal disease.[13]

The aetiology of excessive sinus growth remains obscure and many pathological processes have been proposed by different authors. Pneumosinus dilatans has been shown to be associated with pathological conditions such as planum meningiomas and other brain tumours,[20] arachnoid cysts,[7] fibrous dysplasia, prolonged cerebrospinal fluid shunting and cerebral hemiatrophy infantile meningoencephalitis, Von Recklinghausen's disease and acromegaly.[10] It can also be idiopathic as in our two cases and under these circumstances it is considered a separate entity.

Six different hypotheses have been advanced to explain the aetiology of these cases of pneumosinus dilatans. These are: the presence of a gas-forming micro-organism, spontaneous drainage of a mucocele, the presence of a one-way valve, congenital, programming of osteoblastic and osteoclastic activity and hormonal.

Local causes such as an infection caused by a gas-forming organism have been discounted as no such organism has ever been isolated. Other proposed mechanisms include drainage of a spontaneously emptied mucocele through the sinus ostia with the patient presenting with an episode of profuse nasal discharge prior to their diagnosis of pneumosinus dilatans.[2,6] The senior author has seen one example of this, not presented here. The vast majority of patients, however, fail to describe such symptoms and Benjamins[1] thought this an unlikely cause.

It has been proposed that the cause may be multifactorial and include an existing malformation, congenital or hormonal factors and chronic inflammatory changes. Chronic inflammatory changes are occasionally noted histologically[17,20] although the surrounding 'non-affected' paranasal sinuses have rarely been seen.[21] It has been proposed that inflammation of the surrounding sinuses may stimulate the pneumatization power of the subepithelial layers of the sinus mucosa causing pneumosinus dilatans.[5,17] Although many patients report a moderate degree of nasal obstruction[18] other symptoms are rarely reported and it is felt that this is also an unlikely cause.[17]

The most commonly proposed pathogenic mechanism and the one which has acquired the most credibility is a local process involving obstruction of the sinus ostium with a one-way valve mechanism.[8] A similar mechanism has been implicated in the formation of extra-cranial and cranial pneumocele associated with abnormal fistulous connections.[22] The presence of a polyp or soft tissue mass near the affected sinus ostia is not an unusual finding.[23] It has been proposed that this one-way valve results in long-term air trapping in the affected sinus with air entering under positive pressure after events such as nose blowing which is subsequently prevented from escaping by the valve.[3,18] Several authors have been unable to demonstrate a patent ostium[18] and puncture of a sinus in one affected patient by Benjamins led to air escaping forcibly.[1]

The best experimental evidence for a one-way valve mechanism comes from antral pressure studied[16] showing the absence of patent ostia together with loss of normal antral pressure variation with the respiratory cycle and increased sinus pressure for several minutes after Valsalva's manoeuvre. Other evidence for a one-way valve comes from the pain experienced in the affected sinus by some patients during changes in atmospheric pressure such as flying or diving or by nose blowing[24] although this did not occur in our second patient when diving. Evidence mitigating against the trap door theory includes the normal drainage of sinus secretions in the affected sinus when one would expect mucus to be trapped by the partial obstruction of the valve. In addition, none of the involved sinuses have notable mucoperisteal thickening or other evidence of infection that would be expected with osteal obstruction. A complete occlusion of the sinus would be expected to lead to negative antral pressures and effusion but not to sinus expansion.[16] Furthermore the highly vascularized nature of the mucosal lining would be expected to reduce the pressure of the gases by diffusion in a relatively short time preventing any long-term effects and in fact the pressures demonstrated after Valsalva's manoeuvre have been shown to be normal in some affected patients[16] and to return to normal within 10 minutes. For there to be a trophic effect on bone one would suspect that a sustained pressure would be required. This theory would not explain the focal cases of pneumosinus dilatans.

The congenital conditions associated with pneumosinus dilatans such as the recent finding of an association with polyosteotic fibrous dysplasia (McCune Albright syndrome) may help to support the theory of a genetically altered sinus becoming enlarged after it is acted upon by a programme alteration in the balance between osteoclastic and osteoblastic activity, hence the association of pneumosinus dilatans with conditions such as acromegaly. This theory is as yet unsubstantiated. Another is that it is an endocrine disorder but the reason for its localization cannot readily be explained.[23]

A CT scan is the investigation of choice to make a diagnosis of pneumosinus dilatans and differentiate the swelling from a variety of other causes.

A variety of surgical options have been proposed, based on a presumed aetiology of occlusion of the maxillary sinus ostium by a one-way valve. These include direct sinus needle puncture, creation of a naso-antral window through a Caldwell-Luc exploration and creation of a naso-antral window endoscopically. In the majority of cases this treatment has been shown to arrest expansion of the sinus. It is this treatment which provides the best evidence for the one-way valve mechanism theory.[23] Although growth of the sinus may be arrested by these procedures the deformity will persist. In patients with a functioning ostia the cosmetic deformity is of primary importance. It can be corrected by direct resection and a bone graft from the cranial vault. The reversed bony cyst wall or other donor sites may be used. No recurrences have been documented in patients who have undergone the aforementioned surgery but the follow-up time is short.[23]

Conclusion

The aetiology of the two presented cases of pneumosinus dilatans is obscure. It is of interest that the second patient had started subaqua diving after he had presented with his frontal swelling and that he has had no symptoms during, or after, diving and has had no change in his frontal swelling. This observation, along with the fact that retained secretions are not seen within the sinuses involved in this condition, raise doubt about the theory that a one-way valve is responsible. Neither patient is currently symptomatic and the condition has not progressed.

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Address for correspondence:
Professor N. S. Jones,
Department of Otorhinolaryngology,
Queen's Medical Centre,
University Hospital,
Nottingham NG7 2UH, UK.

E-mail: nick.jones@nottingham.ac.uk

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