Isolated Pneumosinus Dilatans of Maxillary Sinus Combined with Generalized Hypersinus: An Atypical Cause of Acute Facial Pain

VERDEJA, Raul, et al.

Abstract

Pneumosinus dilatans is a rare disease of unknown etiology characterized by expansion of 1 or more paranasal sinuses. The lesion includes an abnormal enlargement of the sinus cavity, but no evidence of bony destruction or pathological changes of the underlying mucosa. In some reports the symptoms involve compression of the optical nerve and associations with meningiomas or arachnoidal cysts. Osteoplasty or decompression surgery seem to be the only effective treatments.

Reference


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Background:
Pneumosinus dilatans is a rare disease of unknown etiology characterized by expansion of 1 or more paranasal sinuses. The lesion includes an abnormal enlargement of the sinus cavity, but no evidence of bony destruction or pathological changes of the underlying mucosa. In some reports the symptoms involve compression of the optical nerve and associations with meningiomas or arachnoidal cysts. Osteoplasty or decompression surgery seem to be the only effective treatments.

Case Report:
We describe the case of a 28-year-old man presenting an aggressive pneumosinus dilatans. All the paranasal sinuses showed expansion, although symptoms presented only on the left maxillary sinus. Previous radiographs made 5 years before for dental treatment allowed us to evaluate the evolution of the lesion. Due to the pansinusal dilatation, multidisciplinary investigations were performed. The patient initially consulted his dentist for an acute painful swelling of the left facial side. Since no dental cause could explain the maxillary “swelling”, he was referred to a maxillo-facial surgeon. A CT scan revealed dilatation of others sinuses, and an intraoral anthrotomy of the left maxillary sinus was performed. In the same procedure, a biopsy of the mucosa was taken, revealing normal findings. Definitive drainage was performed by an ENT specialist using endoscopic meatotomy. Evolution was satisfactory without any complaints, although signs of optical nerve edema were noticed during ophthalmological examination. The 5-month follow-up showed that the disease was stabilized with no need for further treatment. The patient was then lost to follow-up.

Conclusions:
A rare disease of the paranasal sinuses, pneumosinus dilatans should be part of the differential diagnosis for recurrent facial pain.

MeSH Keywords:
Causality • Facial Pain • Paranasal Sinus Diseases

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Background

First described by Meyes (1898) as “pneumatocele” [1], pneumosinus dilatans (PSD) is a rare disease in which 1 or more paranasal sinuses expand, or dilate, abnormally. The characteristic of this disease is the abnormal dilatation of air-filled sinuses lined by a normal mucosa. The main difference from pneumocele, described in 1918 by Benjamin [2], is that PSD provokes a thinning or destruction of the sinusal bony structure. Nowadays, most authors follow the classification proposed by Urken et al. [3] distinguishing hypersinus, pneumosinus dilatans, and pneumocele.

Hypersinus describes a sinus larger than usual that does not extend beyond its normal boundaries and does not demonstrate thinning or erosion of the bony wall [4]. A PSD affecting all the sinuses has been reported as pneumosinus dilatans multiplex [5]. Trauma or iatrogenic pneumatocele can lead to a subperiosteal collection of air external to the sinus [4].

The etiology of PSD is still unknown. Various causes have been proposed, including: a one-way valve effect, gas-forming organisms, spontaneously discharged mucocele, and dysregulation of sexual or growth hormones producing an osteoblastic or osteoclastic activity leading to abnormal expansion [4]. The main symptoms are local pain, headache, ocular alteration, anosmia, and cosmetic disturbance. The preferred treatment is endoscopic surgery to deflate the sinus. To the best of our knowledge, there have been 123 cases worldwide reported in the literature. Most of the patients are males age 20–40 years old. PSD involves only 1 paranasal sinus in 84% of all cases, and in about 63% of cases the frontal sinus is affected [6].

Case Report

A 28-year-old man was referred by his dentist with an acute pain in the left side of the maxilla. Medical history of this otherwise healthy young man was not relevant, except for a facial trauma 6 years before involving only the teeth, without any bony fracture.

The clinical examination showed a painful swelling of the left middle face. Intraorally, a painful hard tumefaction of the

Figure 1. Preoperative CT scan: axial views showing asymetrical expansion of the left maxillary sinus and normal thickness of the bony walls and mucosa.
vestibular region was present. Orthopantomography showed maxillary sinus enlargement without signs of a bony lesion. The facial CT scan revealed a pansinusal dilatation with a very discrete swelling of the lining mucosa (Figures 1–3). An intraoral anthrotomy and a biopsy of the maxillary sinus under local anesthesia were performed as emergency surgical treatment (Figure 4). Four days later, the patient was asymptomatic, and the histopathological examination revealed normal bony and mucosal structures. The evolution of the lesion was then assessed on previous radiographs made 5 years before for dental treatment (Figure 5). Due to the pansinusial dilatation, multidisciplinary investigations were performed by maxillo-facial, ENT, and ophthalmology specialists. Ophthalmological investigation showed signs of ophthalmic nerve compression, but without any clinical relevance. Further examinations were planned. Definitive treatment by drainage of the maxillary sinus was made through nasal approach.

At 5-month follow-up, the patient was asymptomatic. He was then lost to follow-up.

Discussion

Worldwide, 123 cases of PSD have been reported in the literature [6], but only 25 of them involved the maxillary sinus [6]. The term PSD was introduced by Benjamins in 1918, although the first description of these singular pathology was by Meyes in 1898. The sphenoid and frontal sinuses are mostly involved, but the other paranasal cavities can also be affected. The mean age of patients is 20–40 years. Currently associated pathological conditions are multiple cerebral meningioma, spontaneous pneumoencephalos, exophthalmus such as orbital meningioma, and neurofibromatosis [7–11].

The etiology of pneumosinus dilatans is still uncertain. Five mechanisms have been proposed; a spontaneous draining mucocele, the presence of gas-forming microorganisms, the presence of a one-way valve, congenital abnormality, and hormonal change [3,4,9,12].
Diagnosis results from medical history, clinical examination, and radiological investigations using CT or MRI exams [6,9,12–14]. As the pathogenesis of the pain is still unknown, the treatment involves surgical decompression by opening the affected sinus [4,6,9,15]. The accompanying pathologies have to be treated separately.

The present case, involving all paranasal sinuses (sphenoidal, ethmoidal, frontal, and maxillary) was accompanied by maximal anatomical modification and symptoms on the left side of the maxilla, leading to the discovery of the disease.

**Conclusions**

PSD is a very rare disease of the paranasal sinuses, mostly affecting the frontal sinus of young male patients. The correlation with intracranial lesions, as well as compression of neighboring
anatomical structures, suggests a radiological and multidisciplinary approach to reach the correct diagnosis and achieve effective treatment. The literature shows a high frequency of misdiagnosis caused by confusion with other causes of sinusal dilatation. As the etiology is still unknown, the only reliable treatment is surgical decompression. Indications for treatment are pain or local compression and esthetic disturbances. PSD should be part of the differential diagnosis for recurrent facial pain.

References: