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LITERATURE REVIEW

Pneumosinus dilatans: Our experience and literature review

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ABSTRACT

Pneumosinus dilatans (PSD) is a rare disorder that causes aberrant paranasal sinus dilation. Pneumosinus dilatans is typically benign and asymptomatic, without any evidence of bone destruction or pathologic changes in the underlying mucosa. Localized discomfort, headaches, facial paraesthesia and deformities, sensory deficits, olfactory loss, and ocular abnormalities are common complaints. The diagnosis of PSD is confirmed by the radiological investigations. The surgical treatment's objective is to return both form and function of the affected sinus to normal.

We briefly present our experience with pneumosinus dilatans and discuss the characteristics of this disease based on the specialised literature.

KEYWORDS: pneumosinus dilatans, sinus dilation, frontal sinus.

INTRODUCTION

Pneumosinus dilatans (PSD) is a rare disorder, with currently 145 cases reported in the literature at this point[1], with poorly understood etiology and pathophysiology that causes aberrant paranasal sinus dilation, characterized by enlargement of one or more paranasal sinuses. Involvement of the frontal sinus is the most common[2], but it can affect all paranasal sinuses. PSD is a disease that is typically benign and asymptomatic.

There is no evidence of bone destruction or pathologic changes in the underlying mucosa, and its etiology is still unknown[3]. There have been many different hypotheses and theories that described the production mechanism of PSD. Some of these hypotheses include the presence of a one-way valve, the spontaneous rupture of a mucocele, the presence of an infection with gas-forming microorganisms, congenital defects, and a hormonal imbalance associated with osteoclastic and osteoblastic bone remodelling, but yet no agreement was established[4]. Meyes[5] first reported the condition in 1898, but he referred to it as a pneumocele. Later, in 1918, Benjamins gave it the term PSD and explained how it differed from a pneumatocele. Urken et al.[6] divided PSD of the paranasal sinuses into three categories: hypersinus, PSD and pneumocele. Localized discomfort, headaches, face paraesthesia and deformities, sensory deficits, olfactory loss, and ocular abnormalities are common complaints. The diagnosis of PSD is confirmed by the radiological investigations. PSD is typically used to refer to an unusually large, aerated sinus; on radiographic examination, the bony walls of the sinus appear to have normal thickness and the mucosa has normal aspect. The hypersinus is observed on the radiologic investigations as a large sinus with solid walls; however, its development stays within the parameters of what is considered normal. Pneumocele appears on radiological images as an aerated sinus with a notable weakening of the sinus wall.

The differential diagnosis of PSD may include fibrous dysplasia, expansive inflammatory processes and mucoceles. Also, hormonal examinations such as the blood level of the growth hormone (GH), the insulin-like growth factor 1 (IGF-1) are required to help differentiate PSD from acromegaly and exostosis. The surgical treatment's objective is to return both form and function of the affected sinus to normal[7,8].

OUR EXPERIENCE

We hereby present our experience with patients diagnosed with pneumosinus dilatans, based on clinical and paraclinical evaluation.

In our cases, we analysed the head and paranasal sinus CT scans and measured the anteroposterior length diameter of each frontal sinus, on the axial and width, and height in coronal sections, using the method described by Tatlisumak et al.[9] (Figure 1). Also, using the sinus computer tomograph scan, the measurements needed to determine the volumes of the

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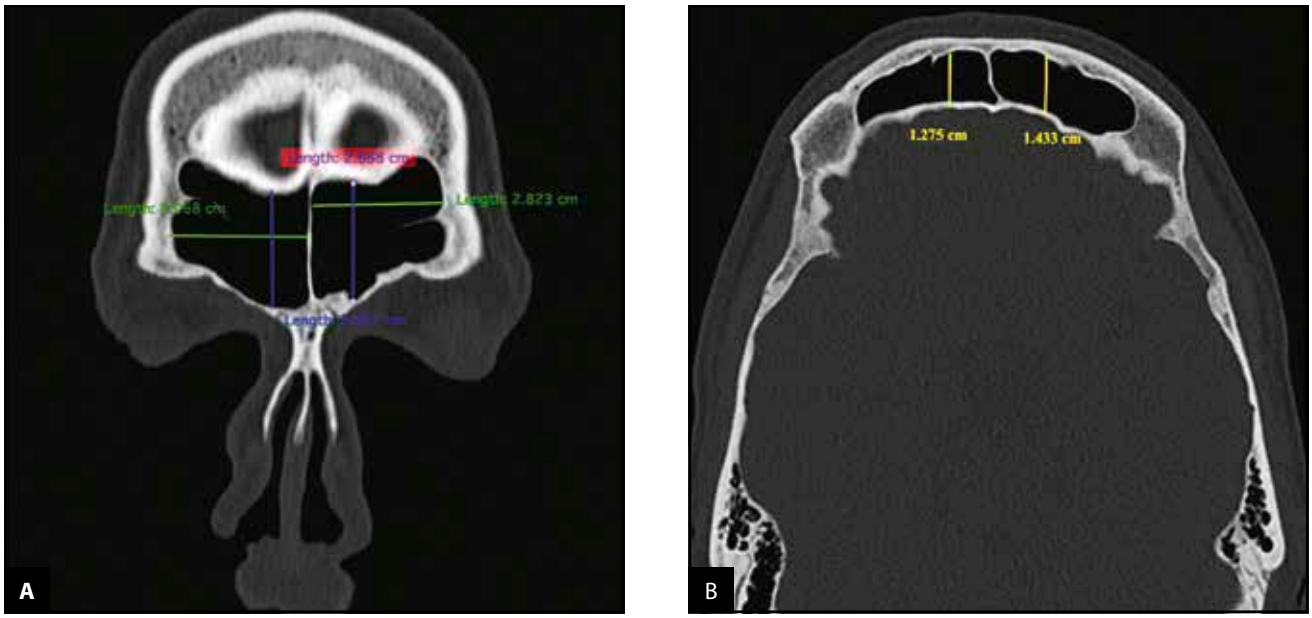

Figure 1. Example of native cranio-facial CT image slice, showing measurement of the frontal sinus: **A.** (coronal view) normal height (blue lines) and width (green lines); **B.** (axial view) anteroposterior length (yellow lines).

Table 1. Linear measurements of the frontal sinus in the total population according to Tatlisumak et al.[9].

	width	*height*	*anteroposterior length*
Left	27.04 +/- 7.79 mm	26.15 +/- 8.75 mm	11.76 +/- 4.74 mm
Right	25.47 +/- 7.82 mm	24.84 +/- 8.61 mm	10.77 +/- 4.14 mm

Table 2. Volumes of the paranasal sinuses according to Karakas and Kavakli[10].

Maxillary sinus	left 11.82 +/- 5.38 cm3	right 11.54 +/- 5.10 cm3
Sphenoid sinus	6.43 +/- 3.41 cm3	
Frontal sinus	4.97 +/- 4.31 cm3	

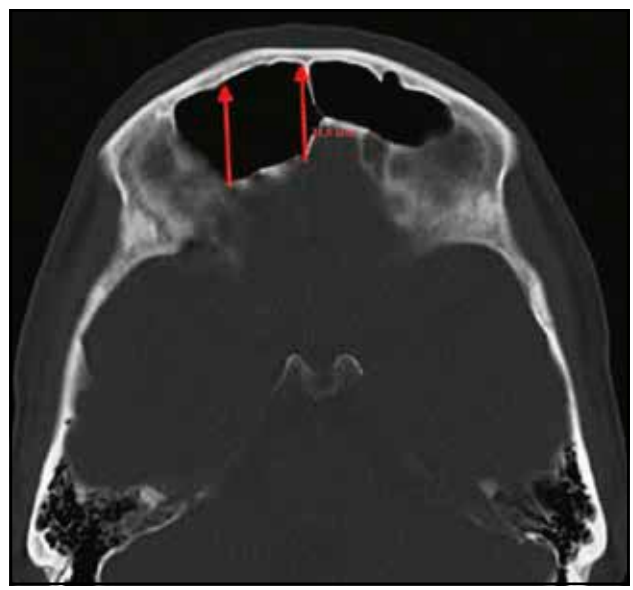

Figure 2. Cranio-facial native CT scan – axial section, revealing right frontal sinus enlargement, beyond the maximum limits (anteroposterior length – 21.5 mm), without any modifications of the bone or the mucosa.

paranasal sinuses – maxillary, sphenoid and frontal, using the method described by Karakas and Kavakli, were performed[10]. First, it was determined the area of each section of the sinus on all the sections, and then that area was multiplied by the thickness of the corresponding section. Next, the volumes of each section were summed up, and then that total was divided by the number of sections to get the accurate volume of the sinus. In all 4 cases presented, all the parameters – width, height, anteroposterior length and volume – of the frontal sinus exceeded the maximum limits (Table 1, Table 2) that were outlined in the relevant academic literature.

A 37-year-old woman, without significant pathological antecedents, presented to our clinic for chronic and persistent headache, symptomatology which started one year before. ENT clinical examination and nasal endoscopy were normal, without signs of acute or chronic rhinosinusitis. The native cranio-facial CT scan examination (Figure 2) revealed right frontal sinus enlargement, beyond the maximum limits, with-

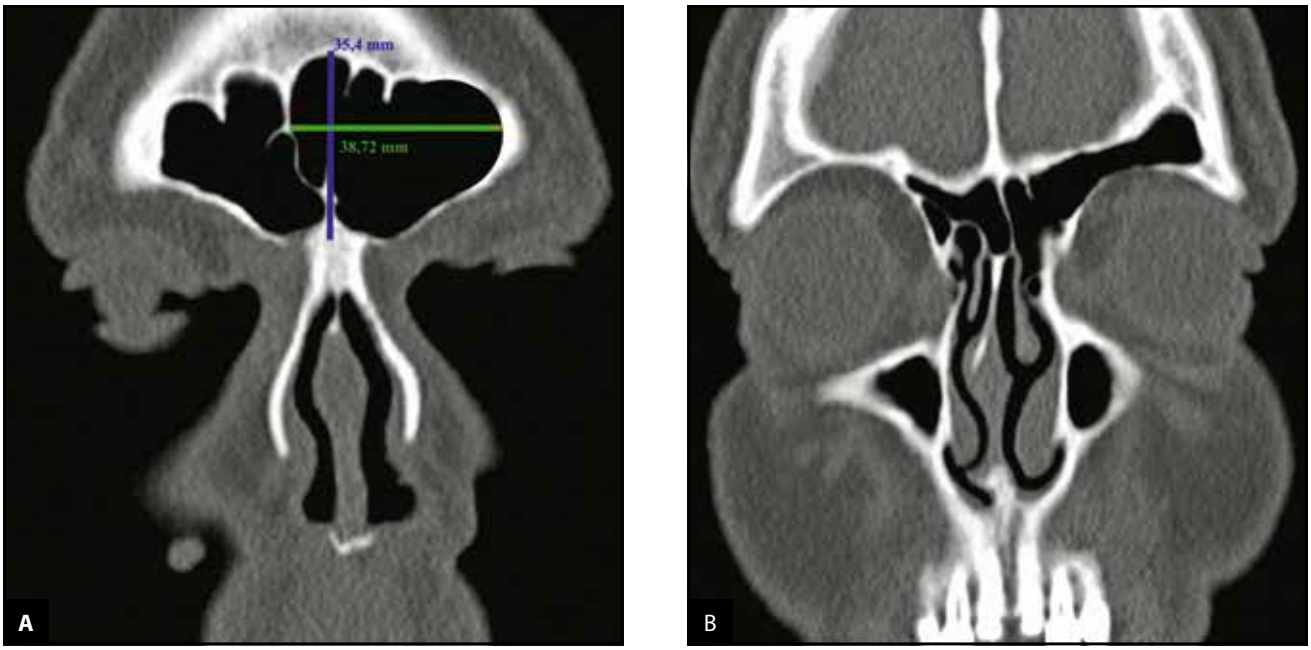

Figure 3. Cranio-facial native CT scan (**A.** and **B.** coronal view) **A.** extensive dilatated left frontal sinus – height 35.4mm (blue line), width 38.7mm (green line), with no signs of mucosal or bony injury and **B.** Coronal view evidencing "deer horn" aspect of the left frontal sinus.

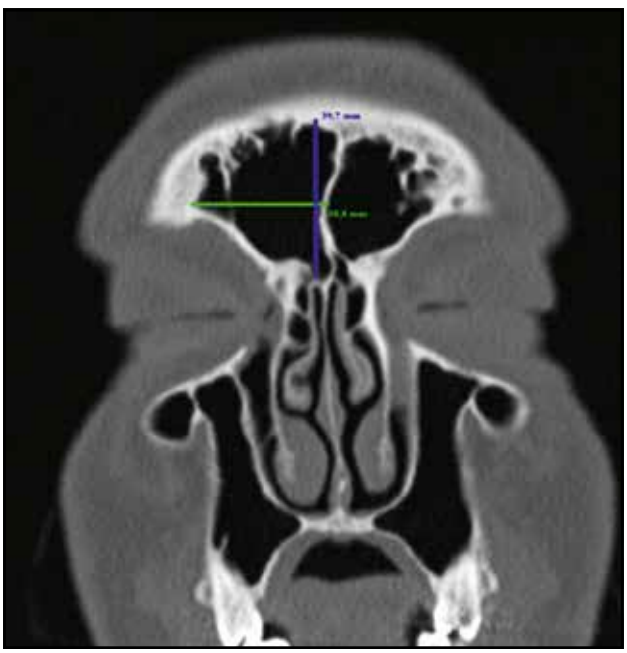

Figure 4. Cranio-facial CT scan, coronal section – enlargement of right frontal sinus – height 39.7mm (blue line), width 38.8mm (green line), with no evident bony erosion and without any abnormalities.

out any modifications of the bone or the mucosa. In this case, no specific treatment was recommended, and the patient is still attending the periodic follow-up.

A 53-year-old woman patient, with no remarkable medical history, presented to the ENT department for episodes of isolated pressure-like headache, specifically over the frontal, anterior parietal, and temporal regions, for approximately 10 years. The ENT clinical examination highlighted painless sinus points, non-detectable on inspection and palpation, abnormality in the frontal region. The nasal mucosa was of normal appearance, without signs of acute or chronic rhinosinusitis, and without nasal polyposis. Complete blood count was within normal limits and ophthalmologic and neurological examination did not reveal any pathological changes. The cranio-facial CT (Figure 3) showed an enlarged right frontal sinus with no evident bony erosion and without any abnormalities. Common analgesics such as non-steroidal anti-inflammatory drugs during the painful episodes represented the treatment in this case.

A 58-year-old man without any considerable medical history, performed a routine cranio-facial CT scan which revealed an abnormal expansion, above the established limits, of the left frontal sinus, with no evidence of bone or mucosal damage (Figure 4). The patient did not present any symptomatology, such as headache, rhinorrhea, nasal obstruction, or swelling of the frontal region. Clinical and endoscopic examination of the nasal cavity were normal, with no pathological signs. In this case, no treatment was required, due to the lack of symptoms or involvement of adjacent tissues.

A 24-year-old woman, with unremarkable medical history, addressed to the ENT Department for chronic nasal obstruction, rhinorrhea, headache, facial pain and hyposmia, symptomatology which started approximately two years before. Nasal endoscopy revealed bilateral nasal polyposis (grade II on the right nasal fossa and grade III on the left nasal fossa). Cranio-facial CT scan showed pansinusitis and an extensive dilatated right frontal sinus, with no signs of mucosal or bony injury (Figure 5). No swelling of the fron-

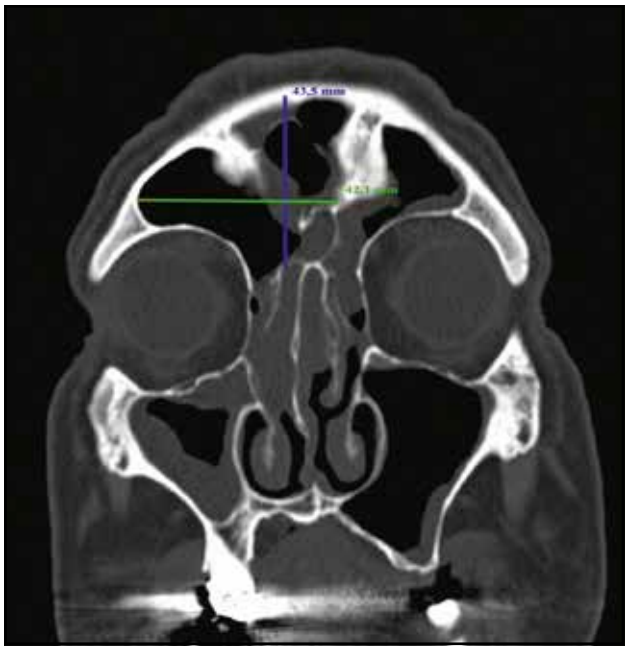

Figure 5. Cranio-facial CT scan A. Coronal view showing pansinusitis and an extensive dilatated right frontal sinus – height 43.5mm (blue line), width 42.1mm (green line), with no signs of mucosal or bony injury.

tal region was observed. In this case, functional endoscopic sinus surgery – nasal polypectomy, left antrostomy, anteroposterior bilateral ethmoidectomy, and bilateral permeabilization of the fronto-nasal duct – was performed to remove the obstruction to ensure adequate drainage of the sinuses. At the 12-month routine follow-up, the patient did not present any recurrence of the symptomatology and no signs of relapse of the polyposis and rhinosinusitis were observed at the nasal endoscopy. Also, the paranasal sinus native CT shows no evidence of inflammation at this level, only the enlargement of the right frontal sinus was maintained just as it was in the previous CT scan.

DISCUSSIONS

Pneumosinus dilatans (PSD) is a rare condition characterized by the abnormal enlargement of one or more paranasal sinuses beyond the normal margins, but without affecting the thickness of the bone and mucosa[11].

The frontal sinus seems to be the most affected. However, the sphenoid sinus, maxillary sinus, and ethmoid cells may also be impacted[3]. The sphenoidal sinus and the posterior ethmoid sinus are the most important for vision loss, this is due to their relationship with the optic nerve through its intimate relationships with the optic nerve canal[12].

According to clinical symptoms, radiographic enlargement and thinning of the sinus walls, Urken et al.[6] divided PSD into three categories: PSD, hypersinus and pneumocele..

An unusually large aerated sinus is typically described as a PSD. According to radiographic examination, the sinus bony walls are of normal thickness and show no signs of bone degradation, hyperostosis, or mucosal thickening. A huge, aerated sinus with intact walls, that doesn't develop beyond the bounds of normal is referred to as a hypersinus. Since hypersinusitis typically has no symptoms, treatment is not necessary. An aerated sinus called a pneumocele has a noticeable weakening of the sinus wall. What distinguishes pneumocele from pneumosinus dilatans is thinning, which can affect specific or generalized sections of the sinus wall[13].

PSD can occur at any age, but more frequently among people between 20 and 40 years old. The average age of presentation is 32 years and men are more susceptible (57%) than women for developing the disease[8].

According to the literature, the condition is quite uncommon among children. This could be a result of both the gradual development of PSD and the age at which the typical paranasal sinus develops[14]. In the presented cases, the average age of the patients was 43 years old of which 3 (75% of the cases) were women and 1 (25% of the cases) man. The values were similar to those present in the specialized literature.

With less than 200 occurrences reported to date, pneumosinus dilatans is considered a rare illness. A previous paper from 2013[2] analysed 123 cases, while a meta-analysis from 2021 included a total of 145 patients from 103 studies[1].

According to the meta-analysis from 2013, 25% of the analysed patients with frontal sinus PSD symptoms also had intracranial pathology (meningioma, orbital tumor, or arachnoid cyst). Patients with ethmoid PSD were 83% more likely to associate exophthalmos, vision loss, or arachnoid cyst, whereas those with sphenoid PSD had an 83% chance of having an associated diagnosis of meningioma, meningitis, or arachnoid cyst[2]. It is notable that the most frequent symptom encountered in the cases of the previously presented patients was headache, without the presence of any deformation of the frontal region or the presence of swelling at this level.

In 1967, Lombardi et al.[15] reported the first series of 51 cases and noted that PSD had a predilection for the frontal sinuses, which were then followed by the sphenoid, maxillary and ethmoid sinuses. Currently, according to the meta-analysis carried out in 2021, the frontal sinus remains the most frequently affected, representing 62% of all cases, while the sphenoid sinus represents 24%, the maxillary sinus 20%, and the ethmoid sinus 19%[1,2]. In 2019, in the literature were found 29 cases of PSD involving the maxillary sinus. The right maxillary sinus was affected in 16 cases, while the involvement of bilateral sinuses was described in 7 cases and the left sinus was implicated in 6 cases, respectively[16]. Regarding the presented cases, it was observed only the implication of one side of the frontal sinus – right frontal sinus in 3 cases (75% of the cases) and left frontal sinus in 1 case (25% of the cases).

In the specialized literature, three cases of PSD multiplex (involvement of all the sinuses and of both mastoid air cells) have been reported[17,18].

Although the etiology and the pathogenesis of PSD have

Table 3. Explained theories regarding the PSD development mechanism.

One-way valve	A massive, aerated sinus results from air pressure entering the sinus but being unable to exit due to the existence of an obstructing polyp, mucosa, or anatomical anomaly[6].
Mucocele spontaneous rupture	An empty, larger sinus can develop from the mucocele's subsequent rupture and drainage[7].
Infection with gas forming microorganisms	A big aerated sinus could be caused by an infection with gas-forming microorganisms, but this theory has not enough data in the literature and needs more research[8].
Hormonal imbalance associated with bone remodelling (osteoclastic/ osteoblastic)	The hormone changes may stimulate osteoblast and osteoclast activity, cause excessive bone formation as well as aberrant sinus pneumatization and expansion[19].
Congenital defects	Among the congenital conditions that may be responsible for PSD's presence are acromegaly, McCune-Albright syndrome, osteogenesis imperfecta, Lawrence-Seip syndrome, gonadal dysgenesis, Klinefelter syndrome, Turner syndrome, and Prader-Willi syndrome[19].

been discussed and researched by various authors, no consensus has been reached[4]. Various theories have been proposed regarding the PSD production mechanism, among which are the presence of one-way valve, mucocele spontaneous rupture, presence of infection with gas-forming microorganisms, congenital defects, and hormonal imbalance associated with bone remodelling (osteoclastic/osteoblastic)[11] (Table 3).

The majority of authors agreed with Dhillon and Williams[20] that one-way valve was caused by either superfluous mucosa or a mild inflammatory condition, which increased sinus pressure. Others think that the hormonal effect of osteoblastic activity leads to the growth and expansion of the sinus, but Smith et al.[21] does not consider the hormonal alterations hypothesis to be valid. Benedikt et al.[22] sustain that the causative agent is the spontaneous drainage of a mucocele.

The diagnostic of PSD is based on clinical examination and confirmed by specific radiological findings. The misplaced structures are often connected to the clinical symptoms. In some cases, PSD can be asymptomatic, being discovered accidentally during radiological investigations[23].

Frontal bossing and prominence of the supraorbital ridge are the most characteristic signs of frontal PSD. The large expansion of the frontal sinus may cause exophthalmia, cranial nerve palsies, and neurological deficits due to the pressure effects[1]. Compressive optic neuropathy of the intracanalicular optic nerve may reduce visual acuity and even vision loss in patients with PSD of the sphenoid or ethmoid sinuses. However, concomitant meningiomas of the optic nerve sheath seem to be a more frequent cause of optic neuropathy in PSD[12]. Headache, sinus pressure, or anosmia are also common symptoms reported by patients with PSD.

Craniofacial CT scanning with or without contrast represents the gold standard method in the diagnosis of PSD. Enlargement of one air cell or the entire sinus beyond the normal boundaries, normal-thick bony walls, and the absence of hyperostosis or mucosal thickening are required for the diagnosis of PSD. Additionally, imagistic investigation (craniofacial CT or MRI) could identify if arachnoid cysts, meningiomas, or cerebral hemiatrophy are associated[24].

Report et al., in 2017, referred to the PSD of the frontal sinus as the "Deer Horn Sinus" because, on coronal computed tomography (CT) images, the condition can resemble a deer horn[25].

Clinical findings, imaging investigations such as craniofacial X-rays, CT and MRI and hormonal cytology are useful in order to establish the differential diagnosis of PSD with other diseases that produce bone deformity – exostosis, acromegaly, fibrous dysplasia, mucoceles and other inflammatory processes[7].

PSD is either referred to in its solitary form or associated with other conditions including fibro-osseous disease, Klippel-Trenaunay-Weber syndrome, meningioma, cerebral hemiatrophy and arachnoid cyst[19].

In order to select the appropriate surgical strategy, the goal of PSD treatment is to return the damaged paranasal sinus to its natural function and structure.

The preferred treatment for PSD patients who do not have facial swelling is the medical treatment with common analgesic or, in the cases of the presence of an obstruction at the nasosinusal level, functional endoscopic sinus surgery (FESS) is the mainstay. This technique involves removing the obstruction at the level of the sinus drainage ostium. In the case of the presence of significant swelling of the frontal region, endoscopic surgery is not taken into account; in such cases, open surgery with osteoplastic flap reconstruction is preferred[16]. Various open approach techniques have been described, including the craniofacial approach using alloplastic material to repair and to remodel the frontal defect, the reduction of the frontal sinus's anterior wall without replacing it, and the excision of the anterior wall of the frontal sinus and replacement with bone fixed using mini plates or with a titanium mesh plate[26].

CONCLUSIONS

PSD is an extremely rare pathology that is defined by an increased extension of one or more paranasal sinus/es beyond the usual limits. The etiology of this condition is yet unknown. The cranio-facial CT is required to be performed in order to establish the diagnosis and to decide on therapeutic options, such as correction surgery, FESS

or medical treatment.

In this article, we present four cases of patients diagnosed with PSD. None of the patients showed any facial swelling or deformities, but the most frequent symptom for which they addressed the ENT Department was headache. Three of the cases did no required any surgical intervention due to the fact that no signs of nasal obstruction or cutaneous swelling were observed.

Conflict of interest: The authors have no conflict of interest to disclose.

Contribution of authors: All the authors have equally contributed to this work.

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