CASE REPORT

Pneumosinus dilatans associated with infantile cerebral palsy and nasal polyposis: Case report

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ABSTRACT

Pneumosinus dilatans is a rare disease characterized by air-filled expansion of a paranasal sinus. Approximately 134 cases are reported in the literature, but not only one associated with infantile cerebral palsy (ICP) and nasal polyposis. We herein present this case report aimed to further characterize this uncommon condition: a 28-year-old female diagnosed with infantile cerebral palsy and nasal polyposis, in whom the cranio-facial CT scan revealed the association of a pneumosinus dilatans involving the frontal sinus. The patient is currently undergoing a protocol for endoscopic surgery of the nose and paranasal sinuses.

KEYWORDS: pneumosinus dilatans, polyposis, infantile cerebral palsy.

INTRODUCTION

Pneumosinus dilatans (PD) is a rare disease characterized by pathologic hyperaeration of one or more paranasal air sinuses[1-3]. The sinus appears filled with air, without radiological evidence of localized bone destruction, mucous membrane thickening or hyperostosis. Facial deformity can be a sign for this pathology. The first who described the disease was Meyes WP in 1898 under the name of Pneumatocoele[4]. Then, in 1918, Benjamin CE assigned the name of Pneumosinus Dilatans[5].

In most cases the frontal sinus is involved[6]. However, the maxillary sinus, ethmoidal cells, and the sphenoid sinus can also be affected[7].

Pneumosinus dilatans has been usually found when diagnosing another cranio-facial or cerebral pathology. Although associations with nasal polyposis, intracranial tumors, unusual bony hyperostosis of the cranio-facial skeleton and hypovitaminosis D have been reported[8-10], a case of PD associated with infantile cerebral palsy and nasal polyposis has never been published in the medical literature.

CASE REPORT

This is a 28-year-old patient with a history of infantile cerebral palsy due to neonatal hypoxia, who was referred to the Otolaryngology Service of our hospital for chronic rhinosinusitis with nasal polyps with more than 4 years of evolution. On physical examination, we found an uncooperative

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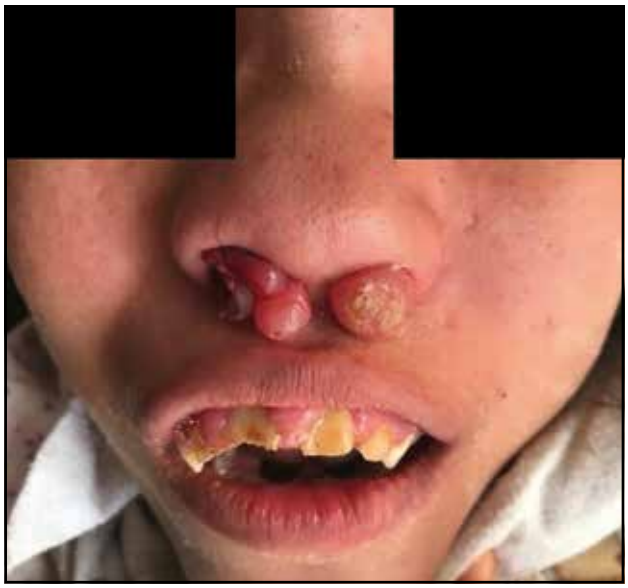

Figure 1. Clinical examination of our patient – pale pink tumors protruding from both nostrils.

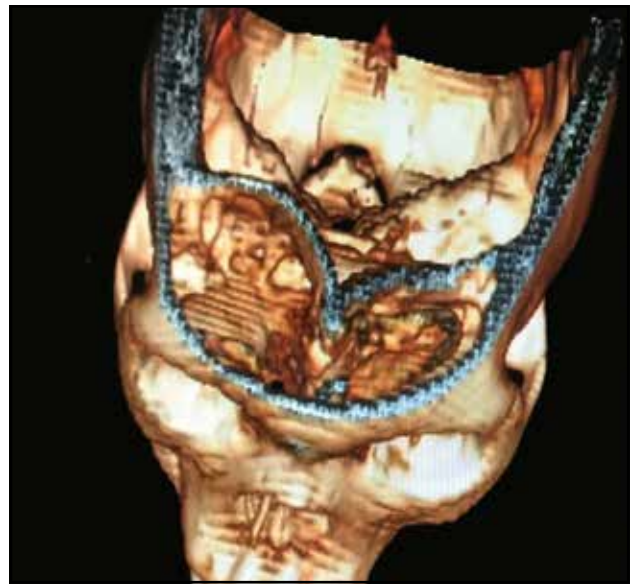

Figure 3. 3D reconstruction CT scan, showing the expansion of the sinus.

bedridden patient with pale pink tumors that protruded from both nostrils (Figure 1).

The paranasal sinus tomography identified an opacification of all the nasal and paranasal cavities due to soft tissue density lesions (Figure 2), and a widely dilated frontal sinus with soft tissues density content A three-dimensional reconstruction tomography (3D reconstruction CT scan) was also performed, and the image showed a widely dilated frontal sinus (Figure 3).

The imagistic evaluation was completed with a cranio-facial MRI. The examination confirmed the soft tissue lesions at the level of nasal cavities and paranasal sinus. The T2 sequence excluded the intracranial extension of the pathologic content (Figure 4).

We decided to take a biopsy of the intranasal tumor. The histopathologic examination identi-

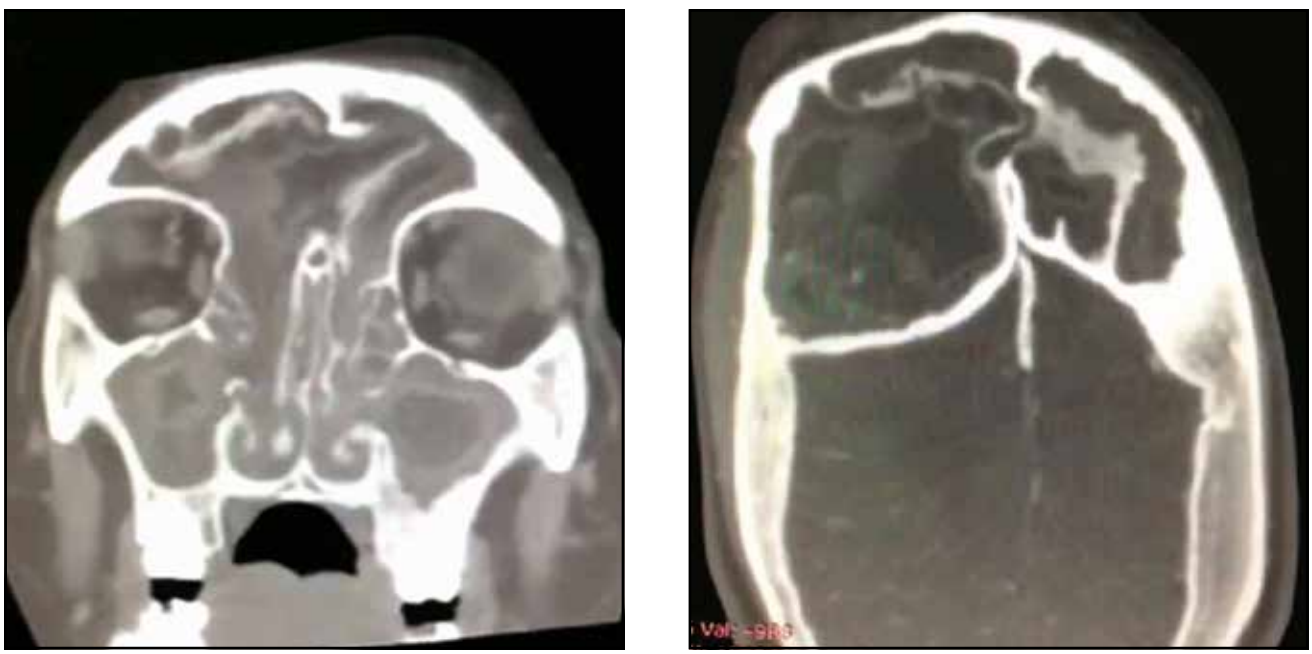

Figure 2. Cranio-facial CT scan: coronal slice - opacification of all nasal and paranasal cavities due to soft tissue density lesions; axial slice - markedly expanded frontal sinus, filled with mucous membrane thickening and secretions due to nasal polyposis.

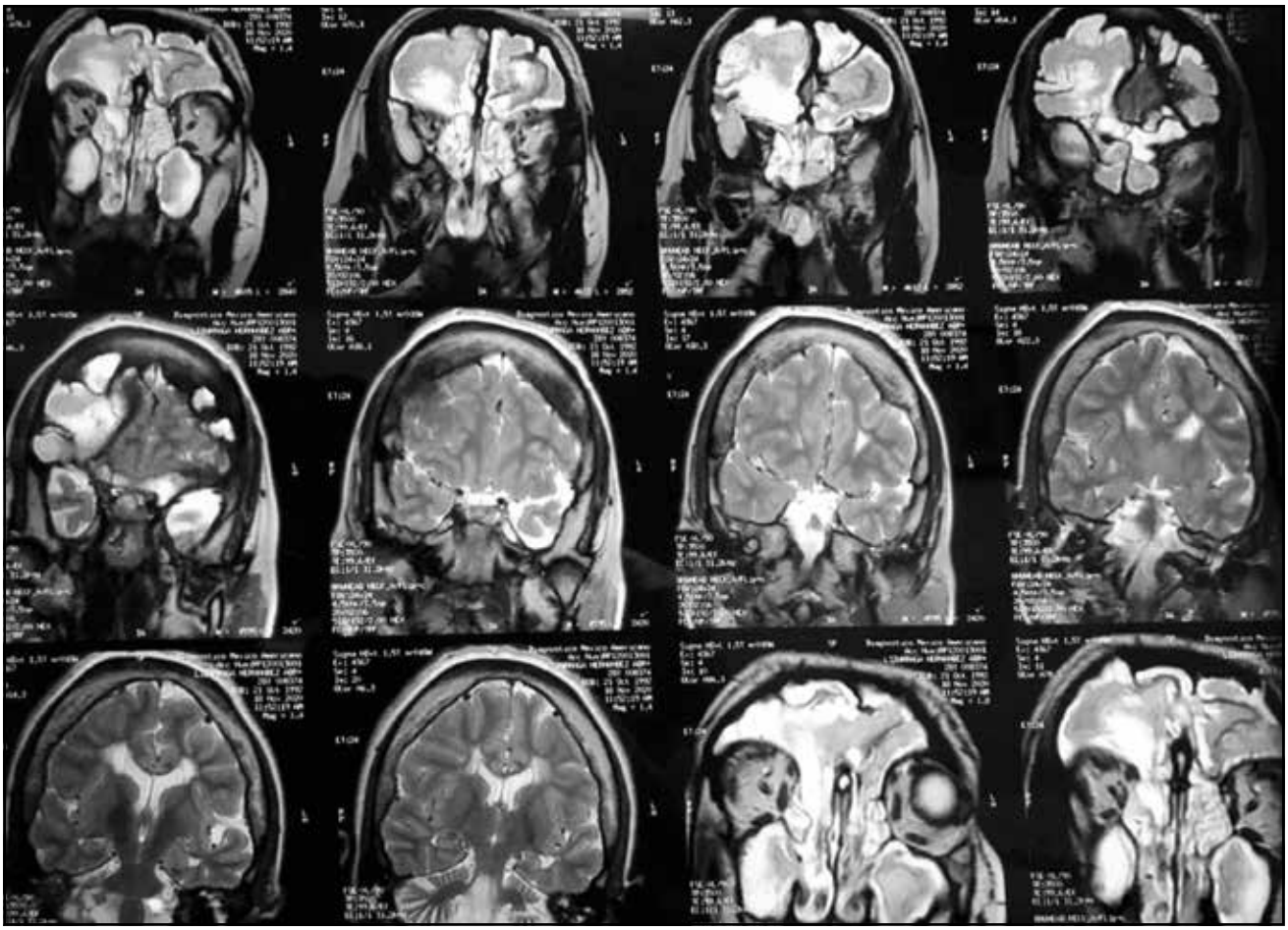

Figure 4. The nuclear magnetic resonance image in T2 sequence shows the absence of intracranial involvement.

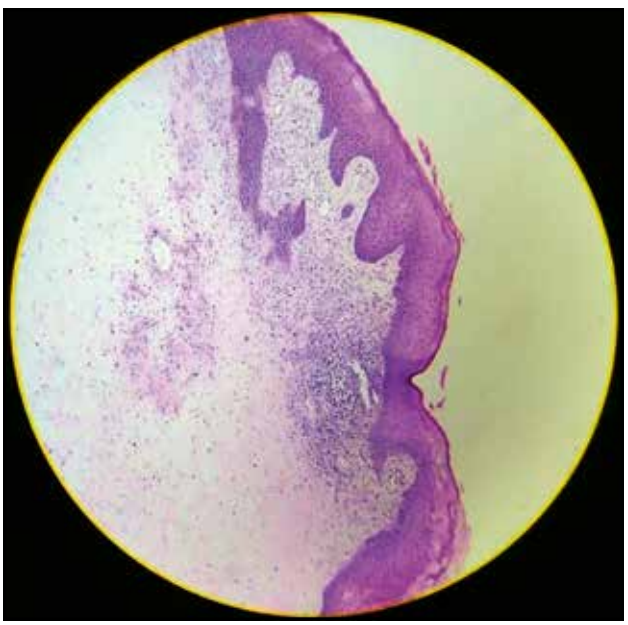

Figure 5. Histological section of the nasal polyp, showing the typical pattern of the polyp cells, inflammatory polyp with intense infiltrate of inflammatory cells in the stroma.

fied a typical pattern of respiratory epithelium, edematous and lax stroma with hyperplastic mucous glands, inflammatory infiltrate with some eosinophils, neutrophils and mast cells (Figure 5), specific for inflammatory nasal polyposis.

Given the special conditions of the patient, we decided to perform an endoscopic surgery procedure in order to permeate the nasal cavity and improve nasal breathing. A bilateral nasal polypectomy was performed with the microdebrider with the permeabilization of both nasal fossae. This procedure allowed us to open the right frontonasal recess with the exit of thick white secretions (Figure 6). The polypoid formations from the left frontonasal recess were ablated with the aspiration of some mucous, viscous secretions from the right frontal sinus. Due to an important bleeding, we decided to interrupt the procedure and to complete it in a second stage, which was performed 6 months after this last intervention. The postoperative evolution and follow-up were within normal limits.

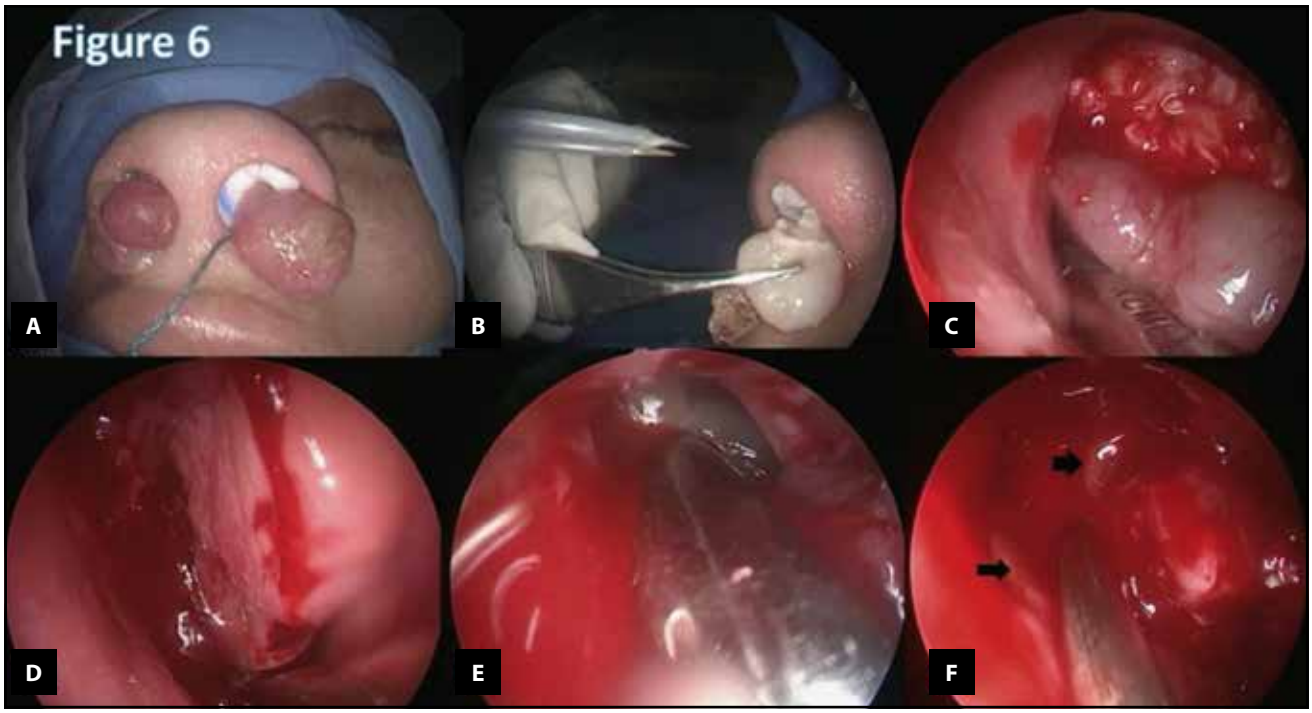

Figure 6. Series of images during surgery: **A** and **B**. the preoperative external appearance; **C**. resection with a microdebrider in the nasal cavity; **D**. permeabilized nasal cavity; **E** and **F**. permeabilization of the recess of the right frontal sinus and the exit of thick white material is evident, indicated by the black arrow.

DISCUSSIONS

This entity is believed to be a rare disorder, but the true incidence of pneumosinus dilatans remains largely unknown[11]. According to Dr. Joseph A. Ricci[8] as of 2017, there have been 134 cases reported since the original case description in 1898.

The etiology of PD is still unknown. Various researchers have proposed various theories[8,12-15], but none have substantial evidence to support their validity and the available data have never been comprehensively summarized in one place. Several theories have been presented in the literature. A frequently involved mechanism seems to be the "ball-valve" or one-way valve mechanism which implies the existence of an obstruction of the frontonasal duct leading to sinus air trapping or a disruption of the cellular milieu due to a fibro-osseous process[8,9,13]. A spontaneous drainage of a frontal mucocele, trauma, hormonal dysregulation, a gas-forming microorganism, the osteoblastic and osteoclastic activity, or congenital defects have also been proposed[12-15]. Unfortunately, the hormonal involvement is a less substantiated theory, while there is no data to support the bacterial process.

As a result of the scarcity of scientific evidence for the different theories proposed on the etiology, patients with pneumosinus dilatans have not been evaluated or treated in a standardized way. Also, the presence of pneumosinus dilatans and concomitant pathologies is scarcely published. The few cases found in the literature show an association of PD with meningioma, nasal polyposis, meningocele, fibro-osseous disease, Klippel-Trenaunay-Weber syndrome, Melnick-Needles syndrome, brain atrophy[6,13,16-20].

In our case, pseudosinus dilatans was associated with infantile cerebral palsy and chronic rhinosinusitis with nasal polyposis, being the first case reported in the literature. The finding of infantile cerebral palsy with frontal lobe cortical atrophy could suppose the hypothesis that the great dilation of the frontal sinus was favoured by the low intracranial resistance in this particular patient. However, this is only a presumption with no robust scientific basis.

The diagnosis of pseudosinus dilatans is usually made on clinical symptoms and is confirmed by the imaging characteristics. The clinical symptoms are directly related to the displacement of the surrounding structures: prominence of the supraorbital ridge, headache, sinus pressure, diplopia, anosmia. According to Thimmasettaiah et al., from the clinical point of view, there can be described two types of PD: one form with early signs and symptoms related to the increase in the sinus pressure, and one form in which the first symptom is frontal deformity, the other symptoms being absent or with late onset[21].

The imaging modality of choice is the craniofacial computerized topography (CT scan), imaging clinching the diagnosis[15].

Correct diagnosis is of great importance in choosing the best surgical approach. The goal of PD treat-

ment is to restore the normal function and shape of the affected paranasal sinus. Different surgical approaches are described in the literature: craniofacial approach, with remodelling and repair of the frontal bulge with alloplastic material that can lead to a high incidence of local complications; reduction of the anterior wall of the frontal sinus without obliterating the cavity with unsatisfactory aesthetic results; removal of the anterior wall of the frontal sinus and its replacement by bone fixed using miniplates or with a titanium mesh plate[13,21-25].

CONCLUSIONS

Our case represents a very unique situation and possibly like this, very few reported in the literature. The challenges in such cases can be various. Starting from the values of the hemodynamic function and going up to the anatomic and surgical variants discovered intraoperatively, all involve the risk of skull base damage. For this reason, we believe that excellent communication between the members of the operating team is essential.

In the present patient, it is probable that a series of incidents that characterize the case as extremely rare have been combined. Finding a case with infantile cerebral palsy, nasal polyposis and pneumosinus dilatans is an extremely strange association and not reported in the national and international literature up to the moment.

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