

Bilateral Visual Loss Caused by Pneumosinus Dilatans: Idiopathic Cases are not Always Reversible

Kaveh Abri Aghdam, Ali Aghajani, Mostafa Soltan Sanjari

Eye Research Center, The Five Senses Institute, Rassoul Akram Hospital, Iran University of Medical Sciences, Tehran, Iran

Abstract

Purpose: To report a rare case of primary pneumosinus dilatans (PSD) and to specify the cardinal imaging findings associated with this condition.

Methods: A 20-year-old patient presented with bilateral profound visual loss as a result of primary PSD. A detailed review of clinical findings and presumed pathophysiological basis of vision loss was performed.

Results: Other than undiagnosed primary hypothyroidism, no other abnormalities were found. With the diagnosis of PSD, the patient underwent optic nerve decompression through transnasal sphenoidotomy. However, after nine months of follow-up, no improvement in the patient's vision was attained.

Conclusion: Unlike previous reports of favorable visual results after sphenoidotomy and bilateral decompression of the optic nerves, vision recovery was not achieved in this case.

Keywords: Optic atrophy, Paranasal sinus, Pneumosinus dilatans

Address for correspondence: Ali Aghajani, Eye Research Center, The Five Senses Institute, Rassoul Akram Hospital, Sattarkhan-Niayesh St., Tehran 14456-13131, Iran.

E-mail: aliaghajani_y@yahoo.com

Submitted: 24-Jul-2020; **Revised:** 05-Feb-2021; **Accepted:** 10-Mar-2021; **Published:** 05-Jul-2021

INTRODUCTION

Primary pneumosinus dilatans (PSD) is a rare condition that is characterized by dilation of one or more of paranasal sinuses beyond the normal anatomic area without erosion of surrounding bony boundaries. It is a distinct feature from hypersinus (dilated sinus that does not extend beyond the normal anatomic boundaries of that sinus) and pneumatocele (a dilated sinus that extends beyond the normal anatomic boundaries and causes erosion of the surrounding bony walls).¹ The condition affects mostly the frontal sinus followed by sphenoid, maxillary, and ethmoid sinus, respectively.² Dilated sinuses are filled with air and covered with normal mucosa.³ Multiple associated etiologies have been described for this condition, including skull base meningioma and hormonal

imbalance, and even sickle cell anemia.⁴⁻⁶ One of the most important presentations of this disorder is a visual disturbance that is because of the proximity of the optic nerve to paranasal sinuses.⁷ Here, we present a patient with PSD of multiple paranasal sinuses and bilateral visual loss.

CASE REPORT

A 20-year-old female patient, suspected of leber hereditary optic neuropathy, was referred to the neuro-ophthalmology clinic. She reported a gradual decrease in the vision of her right eye for 4 years ago and the left eye for the past year. There was no history of trauma, and the patient also denied any administration and usage of potentially neurotoxic drugs and alcohol. No abnormality (such as Cro-Magnon-

Access this article online

Quick Response Code:



Website:
www.jcurrophthalmol.org

DOI:
10.4103/2452-2325.288940

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: WKHLRPMedknow_reprints@wolterskluwer.com

How to cite this article: Abri Aghdam K, Aghajani A, Soltan Sanjari M. Bilateral visual loss caused by pneumosinus dilatans: Idiopathic cases are not always reversible. *J Curr Ophthalmol* 2021;33:197-200.

like brow) was observed in the facial and external orbital appearance of the patient. Ophthalmic examination revealed a best corrected visual acuity of light perception in the right eye and 20/400 in the left eye. The pupils were 5 mm, reactive to light with a positive relative afferent pupillary defect in the right eye. The color vision was not assessable in the right eye and was severely impaired in the left eye (2 out of 14 Ishihara plates). The intraocular pressure by Goldmann applanation tonometry was 14 mmHg, bilaterally. External examination, extraocular motilities, and slit-lamp examination were normal. The fundus examination showed normal disc size and vasculature in both eyes but with bilateral optic atrophy [Figure 1a and b]. Peripapillary circular optical coherence tomography (OCT) scans (Spectralis HRA + OCT; Heidelberg Engineering, Heidelberg, Germany) revealed severe bilateral retinal nerve fiber layer loss [Figure 1c]. A profound visual field depression was also observed in perimetry [Figure 1d and e].

The lateral skull X-ray showed no protrusion of the frontal bone at least up to this time. Neuroimaging investigations

revealed hyperpneumatization of the frontal sinuses as well as sphenoid and ethmoid sinuses [Figure 2]. Furthermore, the expansion of mastoid air cells and calvarium was noticeable. Further examination of the visual pathway revealed the narrowing of both optic canals and optic nerve protrusion into the sphenoid sinus [Figure 3]. Considering other possibilities, genetic and laboratory evaluation was performed to assess toxic (mercury and lead poisoning), nutritional (B12 and folate deficiency), or underlying genetic (Leber hereditary optic neuropathy) conditions; however, other than undiagnosed primary hypothyroidism, no other abnormalities were found. With the diagnosis of PSD, the patient underwent optic nerve decompression through transnasal sphenoidotomy, which was carried out with neurosurgeon-otolaryngologist collaboration after correcting the thyroid function in a single session. After removing the superior turbinate and posterior part of the septum, the sphenoid face was opened, and its air cells were explored and unified with extreme caution not to traumatize the exposed optic nerve. No improvement in the patient's vision was observed for up to 3 months of follow-up after surgery. Unfortunately, due to socioeconomic problems, the patient could not return afterward; however, she did not notice any change in her vision on the last phone call follow-up, which was 9 months after surgery.

DISCUSSION

PSD is characterized by the expansion of one or more paranasal sinuses and thinning of their walls with normal covering mucosa.³ PSD could be categorized as primary, with no underlying structural abnormalities, and secondary, in which other underlying causes could be found. Among secondary causes, skull base meningioma is most noticeable as described by Parizel *et al.*⁴ He and then Scuotto *et al.*⁸ proposed that focal dural traction on adjacent sinus is the cause of bone remodeling and that skull base meningioma should be meticulously sought in any case of PSD. A ball valve mechanism and gas-forming bacterial sinus infection have also been postulated as the cause, though none have been proven.¹ Hormonal imbalance could be another underlying disorder. Parathyroid hormone and Vitamin D imbalance, thyroid hormone, and gonadal hormone dysregulation have been described in this regard, though the exact causative relationship is still doubtful.⁵

This condition affects males almost twice as females and is most prevalent in young adults with a median age of 28.³ The most affected sinuses are frontal sinus followed by sphenoid, maxillary, and ethmoid sinus, respectively.^{2,3} The anatomic variation of the intracranial part of the optic nerve should also be noted in this regard. Dehiscence and protrusion of the intracranial part of the optic nerve into the sphenoid sinus have been shown and are directly related to the amount of sinus pneumatization.⁹ This anatomic variants and their

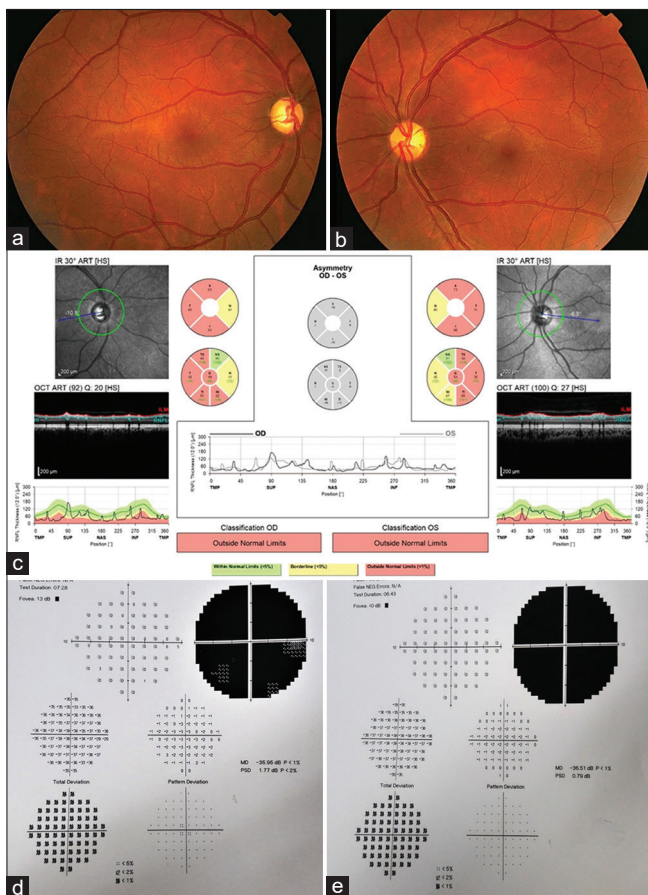


Figure 1: The color fundus photographs of the right (a) and left (b) eyes of the patient show bilateral optic atrophy increased cupping of the right optic disc. Peripapillary circular optical coherence tomography scans show greater than expected retinal nerve fiber layer loss in both eyes (c). The central 10-2 visual field testing of the left (d) and right (e) eye shows severely damaged visual fields in both eyes

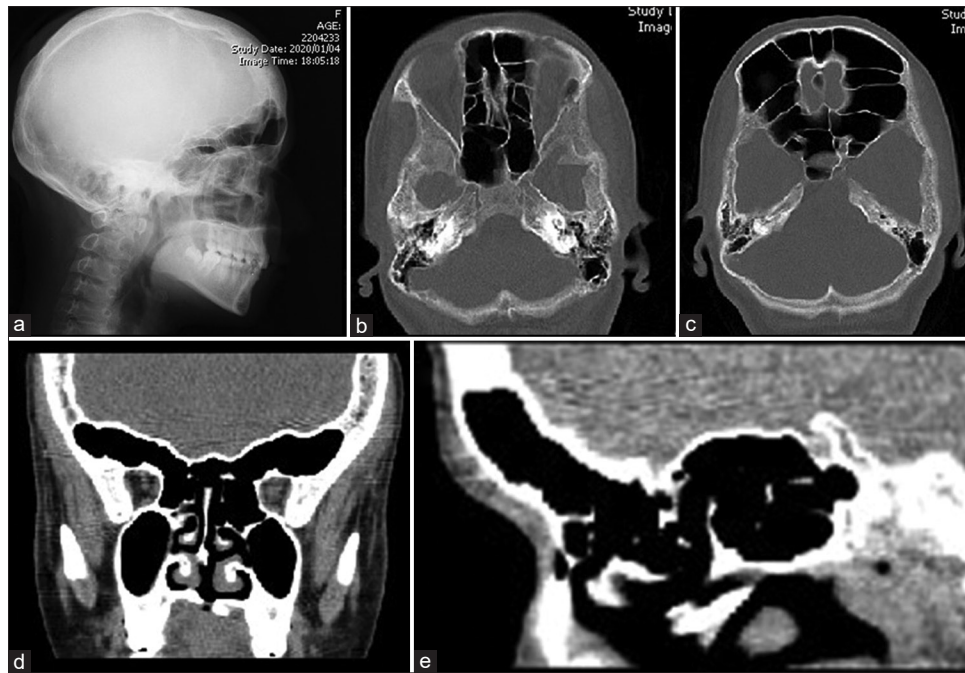


Figure 2: The lateral skull radiograph shows prominent pneumatization of frontal sinuses (a), axial computerized tomography (CT) images render massive expansion of ethmoid (b), and frontal (c) sinuses. Expansion of frontal and ethmoid sinuses is visible in coronal (d) and expansion of sphenoid sinuses in sagittal CT scan images (e)

effects should be kept in mind since almost two-thirds of patients with PSD have been reported to have some kind of visual disturbance, which is a consequence of optic nerve proximity to these structures and possible direct compressive and ischemic effect of air or mucosa on the bulged optic nerve into the sinus or chiasmal compression as a result of the distortion of the tuberculum sellae.^{3,7} The exact mechanism of vision loss is yet to be known. These visual disturbances have been described from temporary amaurosis to visual field defects and permanent unilateral or bilateral loss of vision.^{3,10,11} If PSD is thought to be the cause of vision loss, one should seek possible underlying causes. At least, the part that is related to direct trauma and compression of the optic nerve should be addressed for treatment. That includes creating an outlet for sinus decompression and removing the mucosal lining of the sinuses or the sinus tamponade with fat.⁷

In conclusion, PSD should be considered in any patient with unexplained painless visual loss, especially in the presence of expanded paranasal sinuses, most notably, the sphenoid sinuses. Protrusion of the optic nerve into the sinus cavity is the major risk factor for vision impairment and should be sought out in every case of PSD. However, other causes of gradual painless vision loss should not be neglected. On the other hand, the possible underlying cause of PSD itself should never be overlooked. Careful skull base imaging with contrast should be performed in every patient, looking for meningioma. Brain imaging is also helpful in diagnosing

other associated anomalies, for example, arachnoid cysts. The hematologic and endocrine abnormalities should be considered and thorough laboratory workup should be carried out, particularly for the diagnosis of underlying disturbances in the thyroid, parathyroid, and gonadal hormones. Finally, the vision improvement was not achieved in this patient as a result of permanent optic nerve axonal loss as documented by peripapillary OCT; however, due to the slowly progressive course of the disease and relatively short follow-up period, we could not confirm whether the surgical intervention arrested the progression of vision loss or not. Unlike previous reports of vision recovery after sinus decompression, visual impairment could be permanent in long-lasting cases; thus, early decompression of the affected sinus and optic nerve is highly recommended since there is a chance of vision recovery after sphenoidotomy and decompression, especially in primary cases at early stages.³

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

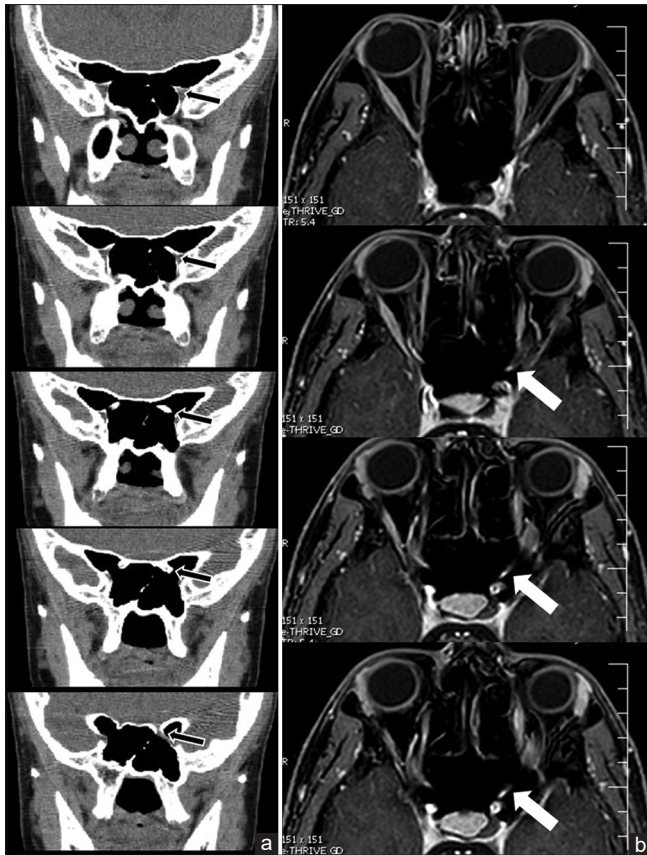


Figure 3: Coronal computerized tomography bone window images depict optic canal narrowing and protrusion of the optic nerve into the sphenoid sinus (black arrows). Note the pneumatization of posterior clinoid processes (panel A). Axial brain magnetic resonance imaging scans with contrast reveal the intracranial portion of the left optic nerve in close contact with the air in the sphenoid sinus (white arrows) (panel B)

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Ricci JA. Pneumosinus dilatans: Over 100 years without an etiology. *J Oral Maxillofac Surg* 2017;75:1519-26.
2. Desai NS, Saboo SS, Khandelwal A, Ricci JA. Pneumosinus dilatans: Is it more than an aesthetic concern? *J Craniofac Surg* 2014;25:418-21.
3. Andrew T, Voglewede JM. Bilateral pneumosinus dilatans of the sphenoid sinuses causing visual loss. *Int J Pediatr Otorhinolaryngol Extra* 2015;10:79-83.
4. Parizel PM, Carpentier K, Van Marck V, Venstermans C, De Belder F, Van Goethem J, *et al.* Pneumosinus dilatans in anterior skull base meningiomas. *Neuroradiology* 2013;55:307-11.
5. Ushas P, Ravi V, Painatt JM, Nair PP. Pneumosinus dilatans multiplex associated with hormonal imbalance. *BMJ Case Rep* 2013;2013:010345.
6. Sanjari MS, Modarreszadeh M, Tarassoly K. Pneumosinus dilatans in a 13 year old female. *Br J Ophthalmol* 2005;89:1537-8.
7. Skolnick CA, Mafee MF, Goodwin JA. Pneumosinus dilatans of the sphenoid sinus presenting with visual loss. *J Neuroophthalmol* 2000;20:259-63.
8. Scutto A, Saracino D, Rotondo M, Izzo A, Urraro F, Cappabianca S, *et al.* Sphenoidal pneumosinus dilatans due to anterior skull base meningiomas-CT and MRI aspects: Report of two new cases and literature review. *Neuroradiol J* 2016;29:295-7.
9. Anusha B, Baharudin A, Philip R, Harvinder S, Shaffie BM. Anatomical variations of the sphenoid sinus and its adjacent structures: A review of existing literature. *Surg Radiol Anat* 2014;36:419-27.
10. Bachor E, Weber R, Kahle G, Draf W. Temporary unilateral amaurosis with pneumosinus dilatans of the sphenoid sinus. *Skull Base Surg* 1994;4:169-75.
11. Kim WJ, Kim MM. Binasal hemianopia caused by pneumosinus dilatans of the sphenoid sinuses. *Indian J Ophthalmol* 2019;67:1772-5.