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## Proboscis Lateralis with Rhinosinusitis

### ABSTRACT

**Objective:** This report aims to describe unique manifestations of proboscis lateralis and highlight the importance of a multidisciplinary approach to address the problems that arise from this rare congenital anomaly.

#### Methods:

|                 |                              |
|-----------------|------------------------------|
| <b>Design:</b>  | Case Report                  |
| <b>Setting:</b> | Tertiary Government Hospital |
| <b>Patient:</b> | One                          |

**Results:** A 13-year-old girl diagnosed with proboscis lateralis presented with a trunk-like appendage projecting from the surface of the right supramedial canthal area. She also had clear nasal discharge, nasal congestion, mouth-breathing and snoring since birth. Paranasal Sinus (PNS) CT scan with 3D reconstruction showed agenesis of the right paranasal sinuses and expansile aeration of the left paranasal sinuses. Due to her condition, the drainage system of the paranasal sinuses was obstructed causing chronic rhinosinusitis (CRS). This hindered plans for reconstructive surgery despite medical management, hence, the patient underwent Endoscopic Sinus Surgery (ESS).

**Conclusions:** Proboscis lateralis is a rare congenital anomaly that results in aesthetic problems as well as airway concerns such as rhinosinusitis and obstructive sleep apnea syndrome (OSAS). Management entails a multidisciplinary approach to address functional and aesthetic problems of the patient.

**Keywords:** *proboscis lateralis, chronic rhinosinusitis, obstructive sleep apnea, endoscopic sinus surgery, multidisciplinary approach, plastic surgery, reconstructive surgery*

**Proboscis lateralis** is a rare congenital anomaly of the craniofacial anatomy with an incidence of 1:1,000,000 to 1:100,000 caused by abnormal embryonic nasal development that results in a trunk-like appendage from the facial surface projecting frequently from the region of the medial canthus.<sup>1</sup>

This report aims to describe the unique manifestations of proboscis lateralis associated with chronic rhinosinusitis (CRS) and obstructive sleep apnea syndrome (OSAS), which, to the best of our knowledge, have not been previously described in the English literature. It also aims to highlight the importance of a multidisciplinary approach to address functional and aesthetic problems that arise from this rare condition.

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**CASE REPORT**

This is the case of a 13-year-old girl who presented with a trunk-like appendage projecting from the surface of the right supramedial canthal area and was born preterm at 32 weeks age of gestation via normal spontaneous delivery to a 30 year-old primigravid. Her mother had fever followed by threatened abortion at the sixth week age of gestation and was managed successfully with Isoxsuprine. Her mother denied exposure to teratogens.

Since birth, the patient had also been manifesting with clear nasal discharge, nasal congestion, mouth-breathing and snoring. She was referred to a tertiary hospital for reconstructive surgery of the aesthetic anomaly. However, repeated attempts at reconstructive surgery were deferred due to persistent nasal discharge despite aggressive medical management. This caused her to be quiet and socially withdrawn with low self-esteem.

Three years prior to admission, the patient was seen at our institution by the plastic surgery service but surgery was deferred again due to persistent nasal discharge and sinus infection which could result in failure of the reconstructive surgery.

A few months prior to admission, she was referred to our otorhinolaryngology service for management of chronic nasal discharge and congestion. She had severe manifestations with a Visual Analog Score (VAS) of 9 for CRS. On physical examination, she had hypertelorism and a nasal appendage located at the right supramedial canthal area. (Figure 1)



Figure 1. Preoperative photographs of the patient (published in full with permission)

Paranasal Sinus (PNS) CT scan with 3D reconstruction revealed a rudimentary right nasal structure attached at the right supramedial canthal area with agenesis of frontal, maxillary, ethmoid and sphenoid sinuses on the right. The left sphenoid and maxillary sinuses had expansile aeration with deviation towards the contralateral side. The left anterior ethmoid cells were aerated. (Figure 2)

Because of the combination of CRS, OSAS, hypertelorism and possible depression arising from the aesthetic problem, a multidisciplinary team

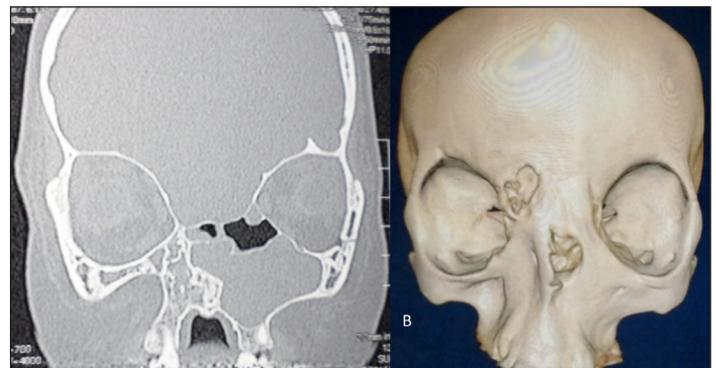
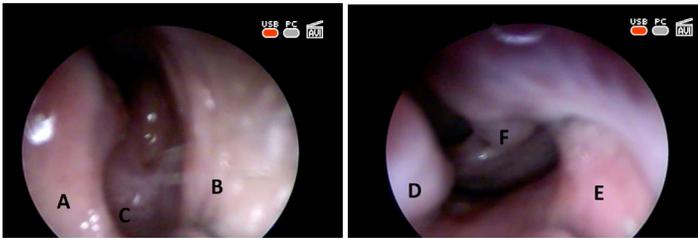


Figure 2. A. Coronal view using 16-slice helical CT scan B. Three-dimensional CT scan reconstruction

approach was necessary. The patient was referred to the snoring clinic to explore treatment of the OSAS and to the ophthalmology department to assess what can be done for the hypertelorism. The ophthalmology service also evaluated the patient’s hypertelorism, weighed risks and benefits and leaned towards conservative management since the condition was not visually disturbing. The patient was likewise referred to the psychiatry department to ensure that the dynamics of a normal growing child in the adolescent stage were evaluated and addressed properly. The pediatrics service addressed the patient’s general health, and developmental pediatrics also played a role in counselling. The otorhinolaryngology, plastic surgery, and anesthesia teams collaborated to plan surgical interventions. The teams on board carefully weighed risks of possible complications of surgery against possible outcomes on the future of a growing child if surgery was delayed or not done.

She first underwent endoscopic sinus surgery. Nasal videoendoscopy showed the clear nasal discharge from the left nasal cavity and from the rudimentary nasal appendage. The endoscope could not be inserted further into the nasopharynx through the left nasal cavity due to the distorted nasal anatomy. (Figure 3) Intraoperatively, the nasal turbinates, uncinat process, and ethmoid air cells were identifiable but were noted to have abnormal structures. Uncinectomy, antrostomy, anterior ethmoidectomy and frontal sinusotomy were performed endoscopically.

Significant clinical improvement was noted on follow-up 9 weeks after the procedure. The patient reported improved nasal airflow, resolution of snoring and decrease in amount and frequency of rhinorrhea. Visual Analog Score (VAS) for CRS decreased from the preoperative score of 9 (severe) to 2 (mild). No complications were noted. In addition to functional improvement, there was also an improvement in self-esteem. With otorhinolaryngology, pediatrics, and anesthesia on board, she successfully underwent initial reconstruction by the plastic surgery service. (Figure 4) Future plans are directed towards a second stage reconstructive surgery after the pubertal growth spurt which occurs at around 16-18 years of age.<sup>2</sup>



**Figure 3.** Preoperative Nasal Videoendoscopy showing obstruction of the nasal cavity. A. Nasal Septum. B. Lateral nasal wall. C. Inferior turbinate. D. Nasal septum. E. Uncinate process. F. Middle turbinate.



**Figure 4.** Before and after photographs (published in full with permission)

## DISCUSSION

Under the Revised Sakamoto Grouping System,<sup>3</sup> this case of proboscis lateralis with hypertelorism, nasal defect but no cleft lip or palate is classified under group III. Proboscis lateralis is commonly associated with median facial clefts which cause hypoplasia or agenesis of one side of the nose.<sup>4,5</sup> This was consistent with the findings in this patient with agenesis of the right side of the nose and nasal sinuses.

However, to the best of our knowledge, there are no published reports in the English literature of proboscis lateralis complicated by the presence of CRS and OSAS. This case showed that proboscis lateralis may be associated with anatomical defects that obstruct drainage of the developed contralateral paranasal sinuses, leading to chronic rhinosinusitis. The patient's chronic and non-resolving sinusitis was due to obstruction of the ostiomeatal unit. The abnormal nasal anatomy caused an obstruction of the left ostiomeatal unit which then resulted in obstruction of drainage of the left paranasal sinuses.

Computed tomography and magnetic resonance imaging have been used as the imaging modalities for preoperative evaluation of patients with this anomaly.<sup>4,5,7-10</sup> In this case, PNS CT scan was crucial for planning the surgical management of the functional airway problem. Previous reports largely focused on the successful outcomes of reconstructive surgery<sup>4,7,9,11,12</sup> but for this case, CRS had to be addressed first. Endoscopic sinus surgery addressed this functional problem by removing the ostiomeatal unit obstruction.

Significant clinical improvement was noted on follow-up. A literature review of eleven studies on the outcomes of ESS in children with CRS showed that success rate ranged from 82 to 100%. Safety profile was excellent with rate of complications as low as 1.4%.<sup>11</sup> However, none of the subjects had congenital anomalies of the nasal cavity. To the best of our knowledge, this is the first report of the successful treatment with ESS of CRS related to proboscis lateralis.

Proboscis lateralis presents with aesthetic and functional problems. ESS played a pivotal role in addressing the functional problems and in paving the way towards definitive reconstruction. The first stage of reconstruction aimed to restore the external anatomy and promote normal development and growth of the nose. Future management includes secondary surgical repair after complete facial growth. However, due to the complex nature of this malformation, achieving a favorable result is often a challenge.<sup>13</sup> Because of this, it is important to have different services working together for holistic and optimal management.

In summary, this paper described the unique manifestations of proboscis lateralis that resulted in chronic rhinosinusitis and OSAS. A multidisciplinary approach involving the otorhinolaryngology, plastic surgery, anesthesiology, pediatrics, psychiatry, and ophthalmology services was essential for the successful management of functional and aesthetic problems arising from proboscis lateralis. The patient is a living testimony to this successful cooperation.

## REFERENCES

- Martin S, Hogan E, Sorenson EP, Cohen-Gadol AA, Tubbs RS, Loukas M. Proboscis lateralis. *Childs Nerv Syst.* 2013 Jun; 29(6): 885-91. DOI: 10.1007/s00381-012-1989-0; PMID: 23354442.
- Bhatt YC, Bakshi HS, Vyas KA, Ambat GS, Laad H, Panse NS. Complete cleft of the upper limb: A very rare anomaly. *Indian J Plast Surg.* 2008 Jul;41(2):195-9. DOI: 10.4103/0970-0358.44842; PMID: 19753263; PMCID: PMC2740503.
- Chauhan DS, Guruprasad Y. Proboscis lateralis. *J Maxillofac Oral Surg.* 2010 Jun; 9(2):162-5. DOI: 10.1007/s12663-010-0046-3; PMID: 22190778; PMCID: PMC3244101.
- Vaid S, Shah D, Rawat S, Shukla R. Proboscis lateralis with ipsilateral sinonasal and olfactory pathway aplasia. *J Pediatr Surg.* 2010 Feb; 45(2):453-6. DOI: 10.1016/j.jpedsurg.2009.10.044; PMID: 20152374.
- Sakamoto Y, Miyamoto J, Nakajima H, Kishi K. New classification scheme of proboscis lateralis based on a review of 50 cases. *Cleft Palate Craniofac J.* 2012 Mar; 49(2):201-7. DOI: 10.1597/10-127.1; PMID: 21219222.
- Verma P, Pal M, Goel A, Singh I, Bansal V. Proboscis lateralis: case report and overview. *Indian J Otolaryngol Head Neck Surg.* 2011 Jul; 63(Suppl 1):36-7. DOI: 10.1007/s12070-011-0182-1; PMID: 22754832; PMCID: PMC3146664.
- Magadum SB, Khairnar P, Hirugade S, Kassa V. Proboscis lateralis of nose—a case report. *Indian J Surg.* 2012 Apr; 74(2):181-3. DOI: 10.1007/s12262-011-0279-5; PMID: 23543614; PMCID: PMC3309082.
- Mohsen N, Mehdi B. Proboscis lateralis: a unique case with choanal atresia and bilateral ophthalmopathy. *Otolaryngology.* 2015 Jan; 5: 183. DOI:10.4172/2161-119X.1000183.
- Franco D, Medeiros J, Faveret P, Franco T. Supernumerary nostril: case report and review of the literature. *J Plast Reconstr Aesthet Surg.* 2008; 61(4): 442-446. DOI: 10.1016/j.bjps.2006.04.007; PMID: 18358435.
- da Silva Freitas R, Alonso N, de Freitas Azzolini T, Busato L, Dall'Oglio Tolazzi AR, Azor de Oliveira E Cruz G, et al. The surgical repair of half-nose. *J Plast Reconstr Aesthet Surg.* 2010 Jan; 63(1): 15-21. DOI: 10.1016/j.bjps.2008.08.040; PMID: 19046661.
- Johns Hopkins Medicine [homepage on the Internet]. Endoscopic Sinus Surgery. Otolaryngology-Head and Neck Surgery. [cited 2015 Dec 07]. Available from: [http://www.hopkinsmedicine.org/otolaryngology/specialty\\_areas/sinus\\_center/procedures/endoscopic\\_sinus\\_surgery.html](http://www.hopkinsmedicine.org/otolaryngology/specialty_areas/sinus_center/procedures/endoscopic_sinus_surgery.html).
- Kapochewa-Barsova G, Nikolovski N. Justification for Rhinoseptoplasty in Children—Our 10 Years Overview. *Open Access Maced J Med Sci.* 2016 Sep 15; 4(3): 397–403. DOI: 10.3889/oamjms.2016.080; PMID: 27703562; PMCID: PMC5042622.
- Bircher A. Proboscis Lateralis. [homepage on the Internet] The Fetus.net. [cited 2017 Feb 09]. Available from: <https://sonoworld.com/Fetus/page.aspx?id=2370>.