

## PROBOSIS LATERALIS

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### ABSTRACT

Proboscis lateralis is an embryological inconsistency which is identified with the middle facial cleft. This outcomes in fragmented arrangement of one side of nose. This underdeveloped nose structure is situated close to the nose but off the midline. It seems like a cylinder and is appended at the undeveloped combination line anytime between the foremost maxillary interaction and the frontonasal cycle. This issue ought to be tended to at an early age and with stage shrewd methodology as this proboscis itself is a valuable design for remaking. We report an instance of proboscis lateralis in a 1-year-old female patient.

**Keywords:** Face/abnormalities, Nose/abnormalities, Nose/pathology.

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### INTRODUCTION

Proboscis lateralis is a rare congenital anomaly. Its incidence is reported to be <1 in 100000.<sup>1</sup> Proboscis lateralis is an embryological anomaly which is related to the median facial cleft resulting in rudimentary form of one side of nose. This rudimentary nose structure is located near the nose but off the midline. It appears like a tube and is attached at the embryonic fusion line at any point between the anterior maxillary process and the frontonasal process.<sup>2</sup> Paul Tessier classified arrhinias into total arrhinias, heminasal arrhinias & proboscis lateralis.<sup>3</sup> According to various studies the various localization points for rudimentary nose fragment may be at the embryonic fusion line between the anterior maxillary process and the frontonasal process. Different levels include:- Level 1: Supramedialcanthal area. Level 2: Medial canthal area. Level 3: Infra medial canthal area. Level 4: Supranostril area.<sup>3,4</sup>

To have a total assessment of this peculiarity Computerized tomographic check is significant which helps in evaluating development of facial and skull bones just as intracranial correspondence. This issue ought to be tended to at an early age and with stage shrewd methodology as this proboscis itself is a helpful structure for remaking.<sup>5</sup> We report an instance of proboscis lateralis in a 1 year old female patient. We have made an internet search for such cases reported earlier but could find only one case reported so far in medical journals of Pakistan,<sup>6</sup> and this case report makes it 2<sup>nd</sup> case.

### CASE REPORT

A 1-year old baby girl was brought to ENT

out patient by the parents for the evaluation of a soft and rounded trunk like tubular structure about 3cm in length originating from near to left medial canthus (Figure-1). A diagnosis of proboscis lateralis was made keeping in view its obvious appearance. It was a consanguineous marriage case with no family and drug history with no history of exposure to radiation.

On general examination the child was healthy and on local examination the left side of nose was not developed. The nasal airway was unilateral. The eye and lips or palate were normal. The right side of nose was also normal and patent. The septum was totally deviated to the left. The reconstructive surgery was planned stage wise. After assessment and work up for general anaesthesia stage 1 surgery was performed. It was planned to use the tubular structure to make left side of the nose so that the same structure is available in future for reconstructive surgery. An incision was made on the inner side of the tubular structure (Figure-2).

This incision was extended on the lateral aspect of hypoplastic left side of nose. The anterior choana was fashioned. The posterior bony choana was opened using the dilator (Figure-3). After opening the posterior choana, A size 7 nasogastric tube was passed upto nasopharynx and the tubular structure was fashioned over the tube making the left lateral nose and ala. The wound was closed in layers (Figure-4).

Anterior nasal packing with soft paraffin gauze was done for one day (Figure-4). The stitches were removed on 7<sup>th</sup> post-operative day. The tube was kept in the nose for 1 month (Figure-5). Parents were counselled for regular followup visits and later stage septo-rhinoplasty.

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Figure-1: 1-year old child with proboscis lateralis.



Figure-2: Incision line.



Figure-3: Nasogastric tube passed after dilating posterior choana with dilator.



Figure-4: Post operative day 1.



Figure-5: 7th postoperative day.

## DISCUSSION

Proboscis lateralis is a rare disease and is scarcely found in the medical literature of Pakistan. This is probably the second case to be reported and that is why a need was felt to highlight this case.<sup>6</sup> Proboscis lateralis (PL) has a typical presentation of a tube-like structure which originates from medial canthal area and is hanging on atrophic side of nose which may be having a central epithelium lined tract. This epithelium is either squamous and/or ciliated respiratory epithelium which expresses mucoid secretions from inside. This PL may be associated with heminasal hypoplasia or choanal atresia,<sup>1</sup> which was present in our case. The precise mechanism of formation of this pathology is not known but theories suggest that there may be imperfect fusion of nasal and maxillary processes.<sup>2</sup> However, it was pointed out by Rontal and Duritz that the imperfect fusion theory does not explain the presence of other associated pathologies so it may be a direct insult to the nasal placode which leads to the formation of proboscis lateralis and associated pathologies.<sup>3</sup> On the basis of presence or absence of other pathologies associated it is classified as follows.

Group-1 consists of isolated proboscis lateralis without any other anomaly (9%). Group-2 is PL with ipsilateral nasal defect (23%). Group-3 is PL with ipsilateral nasal and Ocular adnexal defect (47%). Group-4 is group-3 plus additional cleft lip or palate (21%).<sup>5</sup> Our patient was placed in group-2.

Mainstay of treatment is surgery. The use of Proboscis part is advocated by many authors so was

used by our team for management of the case however a complete excision followed by reconstruction using flaps is mostly recommended by the plastic surgeons.<sup>7,8</sup>

## CONCLUSION

Depending upon the group of the defects (mentioned in discussion) the treatment options can be planned either by ENT team or in collaboration with Eye and plastic surgeons.

**Conflict of Interest:** None.

## Authors' Contribution

MWA: Direct contribution, data collection, SSW: Proof reading, compilation, FF: Data collection, AW: Data collection, typing, proof reading.

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