

Proboscis Lateralis : Report Of One Congolese Case

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Abstract:

Introduction: Proboscis lateralis is known as a rare anomaly related to facial fusion defect. The optimal care of the patient with a multidisciplinary approach that may involve an ophthalmologist, plastic surgeon, oro-maxillofacial surgeon or otolaryngologist has necessary.

Case-report: It's a case note of a 1 day-old female infant was consulted into our department because of the complaint of nasal anomaly. The physical examination revealed a tubular structure originated from the medial canthal region, an absent left nasal structure or right heminasal hypoplasia with a normal right nasal structure. According to those signs, the diagnostic of proboscis lateralis was given.

Discussion: PL is known as a rare anomaly related to facial fusion defect. The precise embryologic mechanism responsible for the development of PL has not been defined. Nowadays, it does not seem to be any racial predilection in proboscis lateralis. In this case, it is very difficult to prove the association between: the embryologic mechanism of PL, the bad follow up of pregnancy, and the prenatal history of exposure to alcohol, drugs auto medication

Conclusion: Ignorance and poor management, and lack of equipment yield impossible the care of proboscis lateralis's patient in our context.

Keywords: Lateralis proboscis, multidisciplinary approach, infant, Congolese.

INTRODUCTION

Proboscis lateralis reveals a rudimentary nasal appendage or structure that is located off-center from the vertical midline of the face. PL is known as a rare anomaly related to facial fusion defect with an incidence of 1/100.000 live births [1,2]. The proboscis is a tubular structure measuring 2-3 cm long and about 1 cm diameter arising from the medial canthal region. So, it is possible for the tubular to contain a small lumen along its length [3]. Sometimes, the nose is normal and heminasal aplasia or hypoplasia is generally associated with proboscis in few cases of literature. The management should start early in childhood to avoid psychological consequences related to this deformity. For the heminosal reconstruction, use of proboscis itself is the best option [2]. In our area, we do not have data concerning this malformation. The aim of this work is to describe that congenital malformation in Congolese people.

PATIENT AND OBSERVATION

A 1 day-old female infant was consulted into our department because of the complaint of nasal anomaly. In the family history, no consanguinity between the parents and no congenital abnormalities or defects. The pregnancy had not been completely followed by an obstetrician. The prenatal history was positive for exposure to alcohol, drugs. The physical examination revealed a tubular structure measuring 2 cm long and about 1 cm diameter originated from the medial canthal region, an absent left nasal structure or right

heminasal hypoplasia with a normal right nasal structure. There was an accompanying abnormality of eyelid and the nasal airway was unilaterally patent (Fig 1,2). According to those signs, the diagnostic of proboscis lateralis was given. We did not do CT scanner, radiographic examine, and reconstruction nose method because of possibility of the family to go under surgery.

DISCUSSION

Proboscis lateralis was first mentioned in 1861 by Forster in his monograph congenital malformations of the Human Body [4]. Later in 1884, Selenkoff reported this anomaly in the autopsy findings of a 34-year old Finnish farmer [5]. PL is known as a rare anomaly, with an incidence of 1/100.000 live births [1,2]. Guerrero shows the incidence in male seems to be higher than in females, with a 3:1 male/female ratio. And the reason for this sex difference is unknown [6]. For Eroglu, the proboscis is a typically obvious deformity of the facial structures, which can be diagnosed prenatally by ultrasound or at birth [3]. In the literature, the proboscis lateralis is divided into four clinical groups based on the associated deformities [7,8]. Group 1 consists of isolated PL without other associated anomalies (9%) and this group is the least common type of proboscis lateralis. Group 2 consists of PL with associated ipsilateral nasal defect (23%), this group is the most common of PL. Group 3 consists of PL with associated ipsilateral nasal and ocular and/or ocular adnexal defect (47%). Group 4 includes the features of group 3 with the addition of cleft lip and/or palate (21%). In

our study, the sex was female and the category was Group 3 of PL. Nowadays, it does not seem to be any racial predilection in proboscis lateralis. Embryologic development of the face and nose is a complex process. Thus, the embryologic defect that results in proboscis lateralis appears to involve the nasal placode, which is a primary organizer of nasal area of midface. Duplication of the nasal placode, which is very rare, may generate a lateral proboscis in the absence of any other facial anomalies. The precise embryologic mechanism responsible for the development of PL has not been defined. Some theories include imperfect fusion of the lateral nasal and maxillary processes and aberrant fusion of the maxillary process of the affected side to the medial nasal process (globular process) [7]. Rontal and Duritz correctly point out that these theories do not adequately explain the associated anomalies and suggest a primary insult to the nasal placode as the likely mechanism for PL development [9]. In this case, it is very difficult to prove the association between: the embryologic mechanism of PL, the bad follow up of pregnancy, and the prenatal history of exposure to alcohol, drugs auto medication. Ideally, radiographs and Computed tomography should be done for determining the extent of the bony and soft tissue of the anomaly. In general, the treatment for lateral proboscis is surgical repair after complete facial growth. However, due to the complex nature of this malformation, achieving an esthetic result is often a challenge [10]. There are several nasal reconstruction methods using the proboscis as a donor [11,12]. Because of the variety of maxillofacial and ocular disease seen with proboscis lateralis, optimal care of the patient needs a multidisciplinary approach that may involve an ophthalmologist, plastic surgeon, oro-maxillofacial surgeon or otolaryngologist and a psychologist. We recommended the patient to go under surgery with a multidisciplinary approach. Finally on one side, poverty, ignorance and sociocultures believes of parents; on the other side poor management and lack of equipment make impossible the optimal care of proboscis lateralis's patient in our society.

CONCLUSION

Proboscis lateralis is known as a rare congenital anomaly with a characteristic appearance. It is an ideal donor tissue to reconstruct the nose in PL associated with hemi nasal aplasia. The management of optimal care needs a multidisciplinary approach, and that attitude stays difficult in our context. The vital prognostic of the patient depends of others associates malformations. The functional and esthetic prognostic depend of nasal reconstruction methods using the proboscis as a donor.

Conflits d'intérêts :

Les auteurs ne déclarent aucun conflit d'intérêts

Contributions des auteurs :

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Figure 1: an absent right nasal structure or right heminasal hypoplasia with a normal left nasal structure



Figure 2: tubular structure measuring 2 cm long and about 1 cm diameter originated from the medial canthal region