Innovative Journal of Medical and Health Science
Received 15 Aug 2020 | Accepted 20 Sep 2020 | Published Online 31 Oct 2020

DOI: https://doi.org/10.15520/ijmhs.v10i10.3133
I Jour Med Health Science 10 (10), 1346–1348 (2020)
ISSN (O) 2589-9341 | (P) 2589-9341 IF:1.6

CASE REPORT

Case report of Partial arrhinia: An extremely rare congenital abnormality

Alen Kinyina[1*] | Benson Mulokozi[2] | Sarah M. Chamos[2] | Jasmine Chadewa[3]

[1]Open University of Tanzania

[2]Sekou Toure Regional Referral Hospital-Mwanza

[3]Jhpiego-Tanzania

Abstract
Arrhinia or nasal agenesis is an extremely rare congenital malformation. It may occur as an isolated defect or with other congenital anomalies. It is characterized by absence or incomplete nasal chambers and the palate is very arched or hypoplastic. We report this case due to its extremely rarity.
Keywords: Partial arrhinia, nasal, congenital anomalies

1 | INTRODUCTION

The absence of an external nose is one of a complex malformation syndrome characterized by the lack of nasal cavities, microphthalmia, high arched palate, coloboma of iris, and microtia[1].The first arrhinia case was reported in 1931[2], and only 43 cases has been reported in world's English literatures [1,6] .It is believed to be due to mutations in the gene SMCHD1 and It can be associated with facioscapulohumeral muscular dystrophy type 2[3].

2 | CASE REPORT

A 26 years old woman, Gravida 3 Para 1 was referred from the Primary Health Care (PHC) center to Sekou Toure Regional referral hospital. She was referred due to cervical dystocia with prematurity and polyhydramnios. She has one living child and history of abortion at 12 weeks GA in the previous pregnancy. The woman had two ANC visits and there is no history of family congenital anomalies, medical or previous surgical history.

After admission, the clinical assessments and ultrasound scan were done which revealed the single tone baby at GA of 35 weeks, FHR of 136 bpm and polyhydramnios; the contractions were moderate. The report did not reveal the presence of any fetal anomalies. During per vaginal examination the cervix was thick, 5cm dilated; the presenting part was not felt.

Caesarean section was done, a male baby 2.1kg with low Apgar score was extracted .On postnatal examination the baby had neither nostril nor nasal chambers, there were no right or left palpable alar cartilage and the baby had nasal dorsum with small central dimple. There was a wide nasal bridge and hypertelorism was noted. The other deformities were noted on hands (polydactyl) and ambiguous genitalia. Resuscitation was done to stabilize the newborn although the condition continued to deteriorate and the baby died after 20 minutes.

Supplementary information The online version of this article (https://doi.org/10.15520/ijmhs.v10i10.3133) contains supplementary material, which is available to authorized users.

Corresponding Author: *Alen Kinyina*
Open University of Tanzania
Email: alenkinyina@gmail.com.

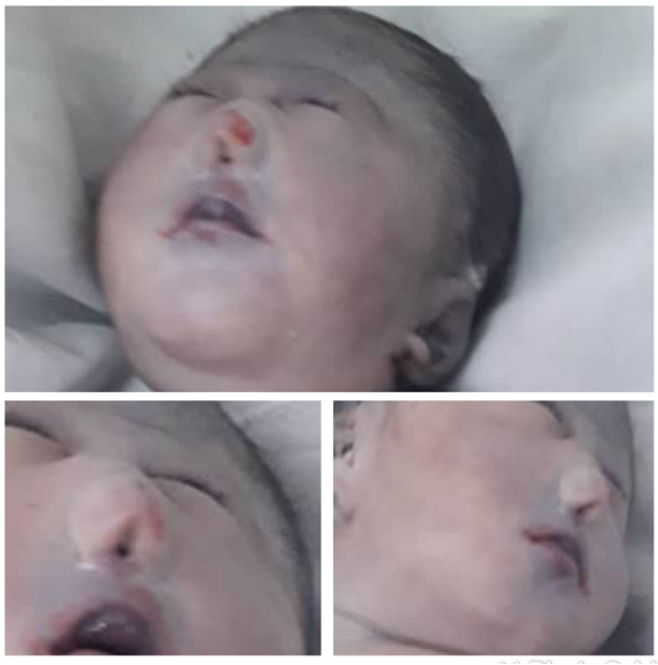

FIGURE 1: Clinical Photograph showing Partial arrhinia

3 | DISCUSSION

Most nasal anomalies are associated with genetic syndrome, and some are occurring due to known exposure of fetus to teratogenic drugs and environment during critical period of embryogenesis [1].That results to delay or failure in mesoderm reinforcement of tissues from neural crest [4].It is believed to be caused by mutations in the gene and can be associated with facioscapulohumeral muscular dystrophy type 2 [3].Because arrhinia is exceptionally rare the factor of gene deformity and pathogenesis is still not fully understood [3].The prevalence is unknown, however 43 cases has been reported in world's English literature [1,6].

The initial management is the same like in choanal atresia, the baby is given time to wait for gaining of mouth reflex ,good coordination of swallowing and breathing during feeding which is approximately 2-3 days of life[4]. It is recommended to do nasal prosthetic without creation of nasal passage [4].The nursing care is very important to the child to facilitate breathing and feeding [4].

In early management the surgical correction of nasal airway or tracheostomy tube or placement of orogastric tube can be created [6]. Reconstruction is very difficult and requires multidisplinary team such as otorhinolaryngologist, cranial facial or plastic surgeons and prosthodontists[4].If reconstructive attempt is needed are best delayed until midfacial development at age of 5-6 years when mucosal and skin lined flaps from external and transpalatal areas can be rotated [4,8].

Several cases are born without complications and they have normal growth development and reach adult stage without surgical treatment[5,6].However some cases reported clinical consequence of congenital arrhinia such as difficulty in breathing and difficult feeding like how it happened in this case[4,6].As it was reported of good prognosis of several cases, the prognosis depends on the presence of other anomalies or complications like how it happened in this case where by; the contributing factor for death of this baby was due to birth asphyxia a common complication associated with prematurity which is among the leading cause of neonatal mortality[7]However, the partial arrhinia accelerated its severity.

In this case the diagnosis during prenatal was missed on ultrasound. The mother had poor ANC follow up during pregnancy which is a missed opportunity for early detection of polyhydramnios and other possible assessments. This could have given good opportunity for adequate psychological counseling to the parents. The genetic studies were not possible due to financial constraints and limited resources at our hospital.

4 | CONCLUSION

The prognosis of this case was poor due to complication of prematurity. Good prognosis in this exceptionally partial arrhinia has been reported in other similar cases and early management and reconstruction is possible. For women with polyhydramnios there is always suspicion of fetal

congenital anomalies, hence thorough assessment and clinical examination during ANC should be done in order to enhance adequate psychological counseling to parents and birth preparedness.

REFERENCES

1. Solanke S, Mahanti S, Parulekar SV (2018).Unilateral ArhiniaJournal of Postgraduate Gynecology & Obstetrics . Medical College and K.E.M. Hospital, Parel, Mumbai.
2. Blair VP , Brown JB. Nasal abnormalities, fancied and real. Surg Gynecol Obstet. 1931; (53):796–819.
3. Shaw ND, Brand H, Kupchinsky ZA, Bengani H, Plummer L, Jones TI.SMCHD1 mutations associated with a rare muscular dystrophy can also cause isolated arhinia and Bosma arhinia microphthalmia syndrome. Available at https:// www.nature.com/articles/ng.3743
4. Pashley NRT. Congenital anomalies of the nose. In: Cummings CW, Fredrickson JM, Harker LE, et al., eds. Otolaryngology-Head and Neck Surgery. 2nd ed. St. Louis, MO: Mosby Year Book; 1993:702–712.
5. Mao-mao Zhang,Yang-hong Hu,Wei He, and Kui-kui Hu.Congenital arhinia.American Journal of case report. 2014; 15: 115–118 available at https://www.ncbi.nlm.nih.gov/pmc/ articles/PMC3966695/
6. Meng LJ, Huang ZL, Niu L. [Congenital arhinia: one case report] Zhonghua Er Bi Yan Hou Tou Jing Wai Ke Za Zhi. 2009;44(4):343–44.
7. WHO (2020) Newborns: Improving survival and well being . Available at https://www.who.int/ news-room/fact-sheets/detail/newborns-reducing-mortality
8. Muhlbauer W, Schmidt A, Fairley J. Simultaneous construction of an internal and external nose in an infant with arhinia. Plast Reconstr Surg. 1993;91(4):720–25

How to cite this article: Kinyina A., Mulokozi B., Chamos S.M., Chadewa J. **Case report of Partial arrhinia: An extremely rare congenital abnormality**. Innovative Journal of Medical and Health Science. 2020;1346−1348. https://doi.org/ 10.15520/ijmhs.v10i10.3133