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Case Report

Amniotic Band Syndrome with Supernumerary Nostril: A Case Report

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Abstract

Amniotic band syndrome is a rare congenital disorder resulting from the entrapment of fetal parts, particularly limbs or digits, in fibrous amniotic bands during intrauterine development. This condition is characterized by a wide spectrum of anomalies affecting multiple systems. Its estimated birth prevalence ranges from 1 in 1,200 to 1 in 15,000 live births. Supernumerary nostril—also referred to as triple or accessory nostril—is another rare congenital anomaly involving the presence of an additional nostril, which may or may not be associated with accessory cartilage. Typically unilateral and isolated, supernumerary nostrils can, in some cases, coexist with other congenital malformations. However, the co-occurrence of supernumerary nostril and ABS has not been documented in the literature to date. In this report, we present a rare case of a female patient born with a supernumerary nostril located above the left nostril in the context of amniotic band syndrome. This appears to be the first documented case of such an association. The aim of this report is to highlight the unusual combination of these two distinct congenital conditions and to contribute to the body of knowledge on the phenotypic variability of ABS. This case underscores the importance of thorough clinical evaluation in patients with congenital anomalies and suggests that rare associations may exist between seemingly unrelated malformations. Further research is needed to explore potential embryological links and to determine whether this association is incidental or part of a broader, yet unidentified, syndrome.

Keywords

Amniotic Bands, Constriction Ring, Supernumerary Nostril, Accessory Nostril

1. Introduction

Amniotic band syndrome (ABS) is a rare congenital disorder caused by entrapment of foetal parts (usually a limb or digits) in fibrous amniotic bands while in utero that presents with complex multisystem anomalies [1-3]. Considering the rarity of the condition with birth prevalence of around 1:1,200 to 1:15,000 live births, it association with supernumerary

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nostril is anecdotal. Supernumerary nostril also known as triple nostril or accessory nostril is characterized by the congenital presence of accessory nostril with or without accessory cartilage. Supernumerary nostril was mostly unilateral and isolated. Also, it can be related with other congenital malformations [4-6].

We report a case of female patient's supernumerary nostril above the left nostril with ABS. As far as we know, this is the first report of such association in the literature.

2. Case Presentation

A 5 days female baby was brought to our department for an accessory opening above the left nostril which had been presented since her birth. The patient and her parents' medical history was non-specific. The age of her mother was 36 years old and there was nothing wrong at parturition. Physical examination showed a left accessory nostril and left unilateral incomplete cleft lip (Figure 1). The size of a supernumerary nostril was about 0.5 cm diameter and not connected to the left nostril. The right one was normal. There was a right columellar deviation. The nasal cavity structure was normal. The patient had constriction band of fingers with syndactyly and short digits along with absence of the 4 last fingers on the right hand (Figure 2). The patient was diagnosed to have amniotic band syndrome associated with supernumerary nostril and unilateral incomplete cleft lip. Patient was lost to follow-up, and was declared deceased by the parent during a phone contact.

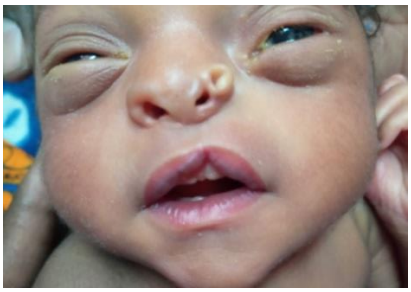

Figure 1. *Clinical photograph revealing accessory nostril above left nostril and left incomplete cleft lip.*

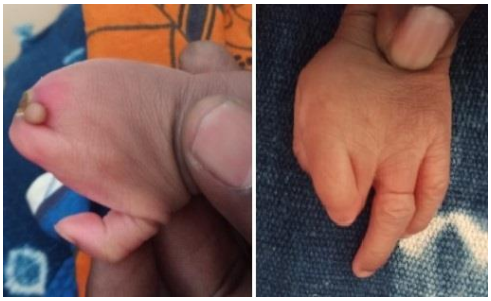

Figure 2. *Clinical photograph revealing syndactyly, short digits on the left hand and absence of the 4 last fingers on the right hand.*

3. Discussion

ABS is a rare condition. The manifestations may be protean in the form of craniofacial deformities, constriction rings in various parts of the body, lymphedema of the digits, and complex multiple congenital anomalies [1, 2, 7]. Predisposing factors for amniotic band syndrome are maternal drug abuse, cigarette smoking, unplanned pregnancy, anorexia, abdominal trauma, infections, amniocentesis, abnormal glycaemic index, very young age of the mother and/or father at the time of conception, populations living at a high altitude, history of febrile illness in the mother in the antenatal period, primigravida, and those women who have vaginal bleeding in the first trimester [1, 7-10]. The pathogenesis of ABS consists of amnion rupture resulting in transient oligohydramnios. The passage of foetus in the extraembryonic coelom through the amniotic defect results in entanglement of foetal body parts with the relatively "sticky" mesoderm on the chorionic surface of the amnion leading to amputations, constriction rings, and skin abrasions [1, 3, 7, 11]. Congenital facial clefts formed as a result of amniotic bands that wrap around the developing foetus may involve the soft tissues of the face and bones [1, 8, 12].

Supernumerary nostrils are exceedingly rare congenital anomalies of unclear aetiology. It is characterized by a nose−like structure arising from the dorsum nasi or ala. It is a very rare congenital anomaly of duplication resulting from embryological defects with less than 50 cases reported after its first description by Lindsey in 1906 [4, 6, 13].

The fissuring of the lateral nasal process during development is the most accepted and cited theory by various authors. This theory may explain the presence of supernumerary nostril occurring in isolation but fails to explain the anomalies associated which are present in significant (45%) percentage of patients. The presence of associated anomalies in supernumerary nostril cases may be due to early significant embryological interruption [6].

The nostril appears similar to the natural ones with vibrissae. It may or may not communicate with the ipsilateral normal nasal cavity, depending on the extent of the anomaly's embryologic progression and with or without accessory cartilage. It can be unilateral (mostly) or bilateral. The external opening of the accessory nostril may be situated above or at the same level or below the normal nostril. It may be situated lateral or medial to the normal nostril. In most of the cases, the supernumerary nostril was located on the left side and above the normal nostril (as seen our case) [4-6, 14, 15].

Associated congenital deformities are seen in 45% of the patients, among them craniofacial malformations are the most common. Reported anomalies are incomplete facial cleft, ipsilateral congenital cataract and microcornea, congenital double columella, patent ductus arteriosus, central incisor fusion, severe sensorineural deafness, ipsilateral/contralateral naso−ocular cleft, cleft palate and lip, hypoplastic heminose, congenital auricular hypoplasia, esophageal atresia, imperforate anus, frontonasal dysplasia, dermoid cyst at the con-

tralateral nasomaxillary groove, and congenital adrenal hyperplasia with clitoromegaly. Furthermore, anophthalmia of the contralateral eye with multiple hemangiomas of head and chest have been reported [4, 6, 14].

The principle of surgery for supernumerary nostril consists of complete removal of soft tissue along with the cartilage between the normal and false nostril. This is followed latter by rhinoplasty for the nostril asymmetry [6, 13]. Saiga proposed a classification of various shapes of supernumerary nostril and ways of treating them. In type 1, the inner nostril is sufficiently large, and symmetry with the unaffected side can be obtained. Treatment is by resection of the outer nostril. This is the type which is most often reported. In type 2, the inner nostril is sufficiently large, and symmetry with the unaffected side can be obtained. Treatment is by resection of the inner nostril. And in type 3, the inner nostril is sufficiently large, and symmetry with the unaffected side cannot be obtained. Although surgery can enlarge the nostril, there is a further problem as to whether or not the inner nostril can be fused with the outer nostril [5].

4. Conclusions

The manifestations of ABS may be protean. Supernumerary nostril is a rare congenital deformation of the nose. To our knowledge, this is the first case of such association in the literature.

Abbreviations

ABS Amniotic Band Syndrome

Author Contributions

Nogognan Ignace Lengane: Conceptualization, writing – original draf

Alain Saga Ouermi: Conceptualization, Writing – review & editing

Hamidou Savadogo: Conceptualization, Writing – review & editing

Ad Bafa Ibrahim Ouattara: Conceptualization, Writing – review & editing

Noe Zaghre: Supervision

Bertin Priva Ouedraogo: Validation

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Data Availability Statement

The data is available from the corresponding author upon reasonable request.

Conflicts of Interest

The authors declare no conflicts of interest.

References

[1] Madan S, Chaudhuri Z. Amniotic Band Syndrome: A Review of 2 Cases. Ophthalmic Plast Reconstr Surg. 2018; 34(4): e110-3.

[2] Phan T, Thi Nguyen P, Nguyen P, Pham H, Ngo Q, La Nguyen P. Amniotic band syndrome leading to severe malformations of the newborn: a case report at Tu Du Hospital, Vietnam, and literature review. Ann Med Surg (Lond). 2023; 85(3): 592-7.

[3] Yang Z, Liang Q, Ouyang K, Piao Z. Amniotic band syndrome with Tessier number 4 and 7 clefts: a case report. Br J Oral Maxillofac Surg. 2019; 57(8): 816-7.

[4] Choi B, Ko S, Shin H. Supernumerary nostril: a case report. Maxillofac Plast Reconstr Surg. 2016; 38(1): 43.

[5] Saiga A, Mitsukawa N. Case of supernumerary nostril. J Plast Reconstr Aesthet Surg. 2013; 66(1): 126-8.

[6] Gupta R, Barolia D, Goyal M. Congenital Hydrocephalus, Corpus Callosum Agenesis, and Prosencephalic Cyst with Supernumerary Nostril: A Neurocristopathy. Asian J Neurosurg. 2018; 13(4): 1239-43.

[7] López-Muñoz E, Becerra-Solano L. An update on amniotic bands sequence. Arch Argent Pediatr. 2018; 116(3): e409-20.

[8] Bouguila J, Ben Khoud N, Ghrissi A, Bellalah Z, Landolsi E, et al. Maladie des brides amniotiques et malformations faciales. Rev Stomatol Chir Maxillofac. 2007; 108: 526-9.

[9] Shirazi Nejad S, Roshan M, Jafarpishe M, Hashemi P, Shahsavan M, Shahsavan M. Prenatal ultrasound detection of fetal amniotic band ingestion in monochorionic diamniotic twin pregnancy: a rare case report. BMC Pregnancy Childbirth. 2024; 24(1): 644.

[10] Muhumad S, Ali A, Moumin H, Egge A, Nour R, Muhumed M. Amniotic band syndrome: Insights from first documented case report in Somalia's low-resource setting. Int J Surg Case Rep. 2025; 129: 111135.

[11] Yadav V, Kumar I, Yadav N, Meena S, Kukreja K. Amniotic Band Syndrome – A Case Report. Journal of Gynecology Research Reviews & Reports. 2023; 5(3): 1-3.

[12] Das D, Das G, Gayen S, Konar A. Median Facial Cleft in Amniotic Band Syndrome. Middle East Afr J Ophthalmol. 2011; 18(2): 192-4.

[13] Widarda I, Soedjana H, Hasibuan L, Lukito R. Nasal reconstruction on supernumerary nostril: A rare case report. Int J Surg Case Rep. 2024; 118: 109627.

[14] Krishna N, Kumar B, Srinivas K, Akurati L. A rare congenital anomaly of nose — Accessory nose : Case report. Indian J Otolaryngol Head Neck Surg. 2006; 58(4): 389-91.

[15] Li C, Pang Q, Zhou H, Alhaskawi A, Dong Y, Lu H. The misdiagnose and treatment of a concealed kind of supernumerary nostril: a case report and review. BMC Surg. 2022; 22: 227.