European Journal of Plastic Surgery (2022) 45:509–513
https://doi.org/10.1007/s00238-021-01860-y

CASE REPORT

Amniotic band syndrome associated with extremely severe atypical clefts of the orofacial region

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Received: 24 April 2021 / Accepted: 2 July 2021 / Published online: 18 August 2021

Abstract
Amniotic band syndrome (ABS) is not a commonly seen birth defect; however, it may have the potential to be severe and life-threatening requiring adequate attention. We present a severe case of amniotic band syndrome which encircled the head of the neonate tightly causing severe deformity. In this case report, the amniotic band encircled the head causing a severe bilateral Tessier 7 cleft. To our knowledge, this is the most severe type of ABS reported in the literature thus far.
Level of evidence: Level V, risk / therapeutic study.

Keywords Amniotic band syndrome · Transverse orofacial clefts · Tessier 7

Introduction

Amniotic band syndrome (ABS) is an unusual and poorly understood congenital anomaly which might be the cause of multiple fetal anomalies [1]. ABS can trap any part of the fetus during intrauterine life causing various birth defects. These range from minor constriction rings to the extremities to severe anomalies that may be incompatible with life [2]. The incidence of this anomaly is not precisely known, but there are reports suggesting that it occurs between 1 in 1200 and 1 in 15,000 live births [3]. The deformity associated with ABS depends on the area it involves. If it involves the orofacial region, atypical clefts of all types may occur, also affecting the eyes, nose, and ears. According to Seeds et al. [3], fetal malformations associated with this anomaly are grouped into three main categories: limb defects, craniofacial defects, and visceral defects. Additionally, a combination of anomalies may exist, similar to what we have seen in our case. The limb abnormalities include constriction rings, amputations, pseudo syndactyly, abnormal dermatoglyphs, and clubbed feet. The craniofacial abnormalities associated with ABS include anencephaly, encephalocele, cleft lip and palate, nasal deformities, asymmetrical microphthalmia, and abnormal calcification of the skull. Omphalocele and gastroschisis are the most common visceral abnormalities. In our practice, we have seen patients with different malformations associated with ABS; here, we demonstrate one specific case with unusual presentations. We believe this case report will contribute to the existing knowledge in the etiopathology of the anomaly. This case report is part of a bigger study entitled The Role of Genetics and Environmental Factors in the Etiology of Orofacial Clefts in the Ethiopian Population for which ethical approval was obtained from the Institutional Review Board at the Addis Ababa University College of Health Sciences (IRB approval number: 003/10/surg) and renewed yearly. In addition, written informed consent was obtained from the parents.

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Case report

This case report is of a female neonate born at term; the second child in four pregnancies (one previous still birth, a missed abortion, and one healthy child) in a healthy 28-year-old mother. There was no history of trauma or drug intake during pregnancy. History of malformations in the family and consanguinity were both absent. The mother received regular antenatal care and had favorable course until term labor. As part of the antenatal care, ultrasound examination was performed at different stages of the pregnancy, which did not reveal any fetal malformations. Delivery occurred at 41 weeks and 2 days gestational age by cesarean section following the indication of prolonged active first stage of labor and a previous cesarean section scar. The outcome of the pregnancy was a female neonate with a birth weight of 3120 g and Apgar scores of 8 in the first minute and 9 after 5 min. During delivery, the fetal chorionic membrane encircled the head above the ear going through the mouth as seen in Fig. 1. It was detached from the neonate after clamping and ligation.

Initial examination showed multiple malformations consisting of a severe bilateral transverse cleft of the orofacial region extending posteriorly and joined with the other side with a deep furrow on the scalp (Figs. 2 and 3) and two deep indentations on the lower lip simulating bilateral incomplete lower lip cleft (Fig. 3). There were no other clinically identifiable anomalies. Karyotype was normal female (46, XX).

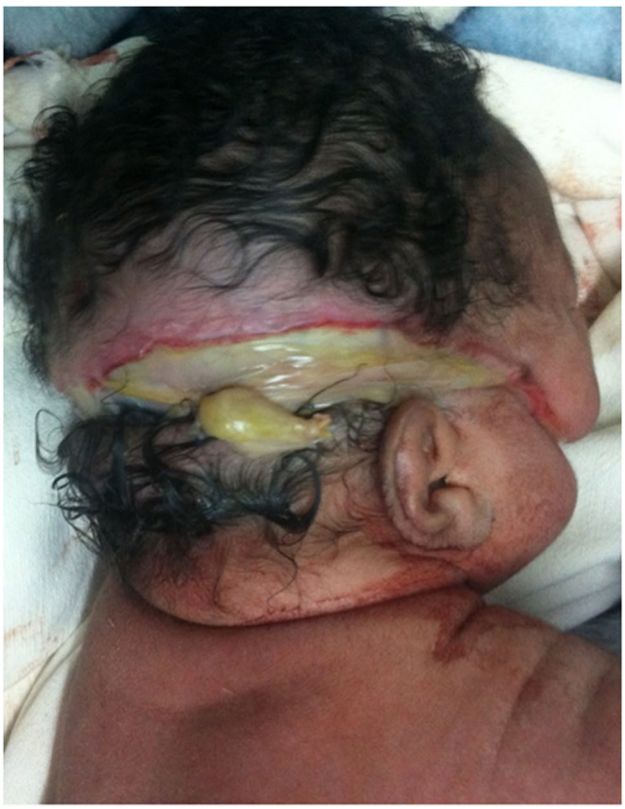

Fig. 1 Amniotic band encircling the head

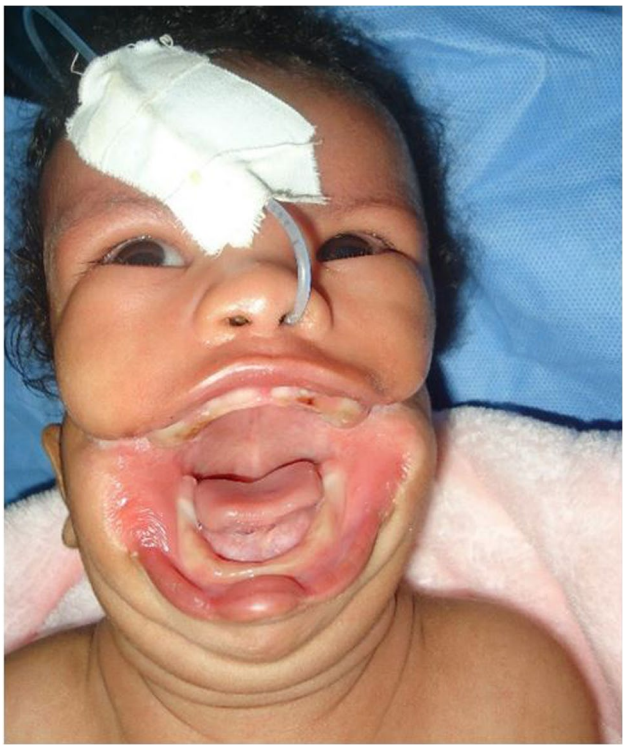

Fig. 2 Front view of the deformity

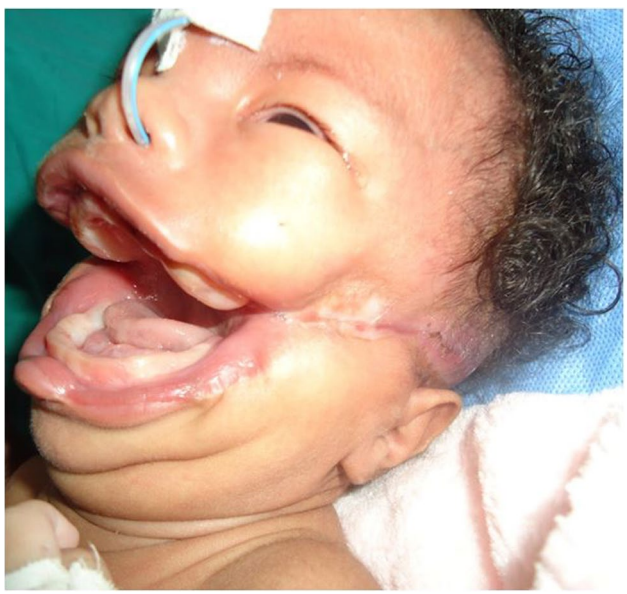

Fig. 3 Side view of the deformity

The patient was referred to Yekatit 12 Hospital Medical College Plastic and Reconstructive Surgery Department following a telephone communication. At presentation, physical examination revealed a 1-week-old neonate who was alert, with nasogastric tube in situ. There was a severe bilateral transverse cleft simulating Tessier 7 cleft and a circumferential scar on the scalp with a palpable defect of the temporal and occipital bones. There were deep indentations on the lower lip causing bilateral incomplete cleft. The patient's mother had reported her eyes as only partially closed during sleep. Both ultrasound and MRI of the central nervous system and abdominal organs showed normal anatomy of examined organs, with the exception of a defect of the temporal and occipital bones. Basic hematologic tests were also within normal ranges. We sought the support of a pediatric anesthesiologist and operated the child with the main aim of improving feeding, speech, and appearance.

Plastic and reconstructive surgeons operated on the neonate using Butow and Botha's four layered techniques with very good outcome [4] at the age of 3 weeks (Fig. 4). This was the youngest patient with orofacial clefts we have operated on with the main intent to improve feeding. Two additional follow-up surgeries were performed at the age of 2 and 4 years old to remove the indentations on the lower lip and improve the side scars. The child is currently 6 years old, has intelligible speech and good appearance, is normal, and is appropriate for age mental and psychomotor development. She cannot close her eyes forcefully or completely during sleep, but there is no corneal exposure. She has no saliva drooling, can eat without difficulty, and only has mild drooling with drinking (Fig. 5). There is a palpable bony defect on the temporal and occipital regions, but it is getting narrower.

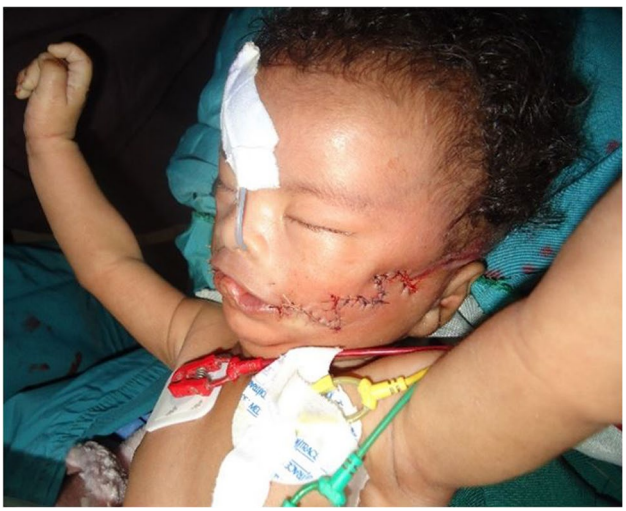

Fig. 4 Immediate postoperative photo

Fig. 5 Follow-up photo

Discussion

We present a severe case of ABS-related facial cleft, an unusual case of a bilateral transverse facial cleft (bilateral Tessier 7). At delivery, a tight amniotic band was found encircling the head passing through the mouth and above the ears. There were indentations on the lower lip causing an incomplete bilateral lower lip cleft. To our knowledge, this is the severest type of Tessier No. 7 cleft reported so far.

The etiology and pathogenesis of ABS is unknown. It is a sporadic disorder [5] which is characterized by constricting rings, acrosyndactyly, and often amputations of the extremities of neonates. Hippocrates recognized this condition as early as 300 BC [6]. He suggested that extrinsic pressure from a ruptured amniotic membrane result in band formation or digital amputation.

After examining many placentae and infants born with amniotic band syndrome, Richard Torpin [7] reintroduced the extrinsic theory (Hippocrates' theory). He noticed lack of a complete amniotic lining in the placentae of neonates born with ABS. Torpin also observed strands of amnion around constricting rings of the digits, and binding strands at the tips of limbs with acrosyndactyly. Similarly, we observed amniotic bands encircling the head of the neonate

and divided after ligation as shown in Fig. 1. This presents additional strong evidence supporting Hippocrates' suggestion regarding the role of ruptured amniotic band in the occurrence of this disorder. Unfortunately, the placenta of our patient was not examined properly to support Torpin's findings. According to Torpin, intrauterine trauma could be the cause of premature rupture of the membranes. There was no history of intrauterine trauma in our case.

Matic and Komazec [8] stated that most cases of ABS are not of genetic origin with no recurrence in siblings or children of affected adults. There was no recurrence in this case study; the patient has an older sister and a younger brother without ABS in either of them.

Although there is no firm evidence of definitive causality, there are studies reporting a connection between ABS and the mother's age especially primiparas under the age of 25 [9, 10], birth prematurity [11], abdominal trauma [10], unsuccessful abortion [12], intrauterine contraception [13], cerclage [14], and chorionic villus sampling [12]. Having little evidence of definite causality for any of the above listed factors, many investigators consider the occurrence of ABS as sporadic with no strong risk factors and gender prevalence [15]. The parents of our patient did not present any of these listed risk factors.

Another attempt to explain the cause of this anomaly was done by Streeter in 1930 [16]. He proposed the intrinsic model suggesting that both the anomaly and the fibrous bands have a common origin as caused by a deviation in the normal development of the early embryo's germinal disc.

The morphology of the deformity on the developing fetus could be determined by the severity of the band compression. As shown in Fig. 2, the deep and circumferential amniotic band caused severe and atypical transverse facial cleft involving deeper structures: temporal and occipital bones and also the facial nerve. The child currently cannot close her eyes tightly. On the other hand, when the bands are superficial, only skin indentations occur.

Prenatal ultrasound examination can sometimes detect amniotic bands, but more often malformations associated with ABS, oligoamnios, and reduction of fetal movements [17]. According to Merrimen [18], it is possible to detect ABS as early as 12 weeks of gestational age. A second trimester routine ultrasound examination can detect most of the defects associated with ABS [17]. In our case, with regular antenatal care inclusive of ultrasound examination, neither amniotic band abnormality nor malformation was detected. This is an area needing improvement in the care of pregnant women in a resource-limited setting. Procuring the latest three-dimensional and four-dimensional ultrasound machines and enabling the professionals to use them is equally important.

The management of children born with birth defects in low- and middle-income countries such as Ethiopia is not simple. It should start with parental counselling inclusive of psychological support to the parents, feeding advice, emphasizing the need for multiple surgeries, and thorough follow-up. The surgical procedure for patients with ABS may range from minor to complex and the outcomes depend on the severity of the deformity. In our case, the surgical management was challenging because of the need to operate at an early neonatal age. Our patient's surgical management is not yet complete, as she will need some additional follow-up surgeries to improve the indentations on the temple regions. The bony defect on the temporal and occipital regions is narrowing but might necessitate bony reconstruction in the future should the bony defect remain open. There are some signs of facial nerve involvement such as the inability to close eyes forcefully, nor during sleep, and mild drooling while drinking.

Conclusions

Amniotic band syndrome is a rare birth defect and has the capacity to threaten life or cause a severe incapacitating deformity. This anomaly may be detected as early as the first trimester of pregnancy if the appropriate facility and expertise is available. Measures can also be taken when the deformity is not compatible with life. Therefore, we need to improve the antenatal follow-up at our institutions; this might include procuring the latest advanced ultrasound machines and providing education for our medical professionals. Furthermore, additional education is needed in the community for the improvement of attitude, beliefs, and understanding of birth defects so that those affected will receive timely treatment and reduce stigma.

Acknowledgements Transforming Faces, Smile Train, and Yekatit 12 Hospital Medical College are acknowledged for supporting the treatment of this child and other children and adults born with orofacial clefts. The authors would like to thank Dr Larry Hollier and Abel Smerica for reading our case report and providing valuable comments.

Funding This study was supported by Grant R00-DE022378 (Azeez Butali) and subaward to Mekonen Eshete and by Smile Train.

Declarations

Ethics approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. Ethical approval was obtained from the Institutional Review Board at the Addis Ababa University College of Health Sciences (IRB approval number: 003/10/surg) and renewed yearly.

Consent to participate Written informed consent was obtained from the parents.

Consent for publication Written informed consent was obtained from the parents including the use of the child's photos.

Conflict of interest Mekonen Eshete is a member of the Smile Train Research and Innovation Advisory Council. Muhidin Abdo Banko, Abiye Hailu, Abeje Brhanu, Peter Mossey, and Azeez Butali declare no competing interests.

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