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CASE REPORT

Amniotic band syndrome associated with orofacial clefts: a report of two cases

O.O. Adeosun[1], O. James[2], V.I. Akinmoladun[3] & T. Owobu[4]

[1]Oral and Maxillofacial Unit, Department of Dental Surgery, Federal Medical Centre, Lokoja, Nigeria
[2]Department of Oral and Maxillofacial Surgery, Faculty of Dentistry, University of Maiduguri, Maiduguri, Nigeria
[3]Department of Oral and Maxillofacial Surgery, University College Hospital, Ibadan, Nigeria
[4]Department of Dental Surgery, Federal Medical Centre, Uguru, Nigeria

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Correspondence to:
Dr O James
Department of Oral and Maxillofacial Surgery,
Faculty of Dentistry
University of Maiduguri
Maiduguri 234
Nigeria
Tel.: +234 703 12715 42
email: olutayo.james@gmail.com

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Abstract

This article presents two cases of amniotic band syndrome associated with orofacial deformities. There was an uncommon presentation in one of them as Tessier nos. 5, 7 and 30 with irregularly distributed amniotic facial band. The surgical repair of the facial clefts and difficulties in overall management in our environment are further discussed.

Introduction

Amniotic band syndrome (ABS) is a set of congenital malformations attributed to amniotic bands that entangle fetal parts during intrauterine life. This results in various birth defects ranging from minor constriction rings and lymphoedema of extremities to multiple anomalies incompatible with life[1]. ABS that involves the orofacial region may present as cleft lip or palate, asymmetric micro-ophthalmia, nasal deformity and oblique facial cleft[2]. These craniofacial anomalies in ABS are typically often bizarre and frequently non-embryological in location[3]. The exact cause of ABS is still unknown. This lack of direct evidence has led to the proposal of two main aetiological theories: the extrinsic and the intrinsic[4]. The extrinsic theory states that the band of ruptured amnion causes an extrinsic compression which then results in constriction rings and other deformities of the developing fetus while the intrinsic theory holds that germ cell deficiencies result in the malformations of the affected parts[4]. Treatment is usually carried out after birth when plastic and reconstructive surgery is considered to treat the resulting deformities. We present two cases of ABS associated with orofacial clefts and the surgical repair of the resultant facial deformities.

Case reports

Case one

A 15 year old lady presented to our unit with a history of congenital upper-lip defect and limb anomalies. There was no previous history of such defect in her

Figure 1 Photograph showing both upper-lip and hands deformities.

family. It was reported that the mother took some traditional medicine for an unknown intermittent ailment during pregnancy.

Findings on clinical examination were unilateral, left-sided, complete cleft of the upper lip with rotated incisors (Fig. 1), amputation of the index and middle fingers of the right hand at inter-phalangeal joints. The little and ring fingers were moderately developed but terminated at the joint between the middle and distal phalanges with fused ends (Fig. 1). On the left hand, the index, middle and ring fingers terminated at inter-phalangeal joints (Fig. 1). There was an amniotic band constriction at the right leg, associated with a swelling distal to the point of constriction (congenital lymphoedema) (Fig. 2). The hallux and the second and third toes of both feet were amputated at the distal ends, while the fourth and fifth toes were slightly developed, with nail beds (Fig. 2).

The lip defect was repaired by the Millard rotation advancement procedure. However, the limb deformities are yet to be corrected.

Case 2

A 9 year old female patient presented to the hospital with a complaint of congenital facial deformity. She was delivered prematurely at home by a traditional birth attendant. There was no history of maternal exposure to teratogens during pregnancy and no family history of such hideous deformity.

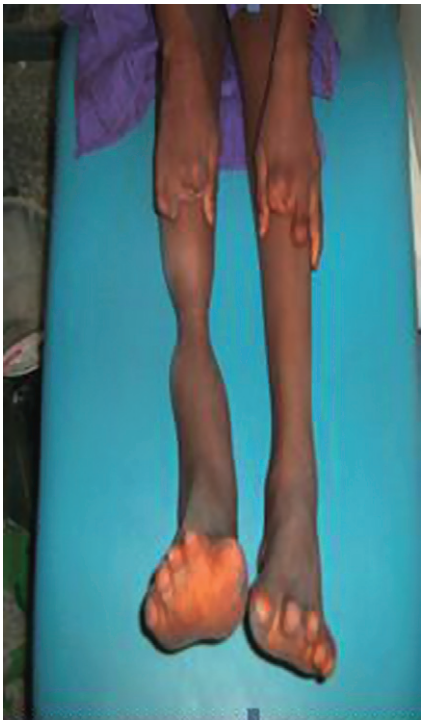

Figure 2 Photograph showing right leg amniotic band constriction with lymphoedema and fingers and toes amputations.

On examination, a depressed scar, about 2 cm in diameter extending from the apex of the v-shaped cleft of the lower lip (Tessier no. 30) to the tragus of the right dysmorphic ear was found. Superiorly, the scar traversed the root of the right zygoma and terminated at the posterior end of the right temporal region. There was also a right-sided commissural cleft (Tessier no. 7) of about 3 cm in length that was associated with a band that extended from its base (Fig. 3). The left facial region revealed a retro-positioned eye with cryptophthalmus; transmitted movement of the eye behind this covering tissue was also observed. Furthermore, there was an oblique facial cleft (Tessier no. 5) that extended from the lateral end of the upper lip to a coloboma at the junction of the middle and lateral third of the left lower eyelid. There was a deformity of the lower lacrimal canaliculus, with the lid retracted inferiorly (Fig. 4). The underlying anterior wall of the maxilla was hypoplastic. Also, the lateral portion of the supra-orbital ridge was slightly depressed and there was an associated diagonal scar extending from it to the hairline of the left temporal area. The cleft of the lower lip was repaired by the V-plasty and the right-sided commissural defect was repaired by the linear suturing technique. Six months later the patient was recalled for the repair of the oblique facial cleft. The lower-eyelid

Figure 3 Photograph showing right dysmorphic ear, amniotic band and right-sided commissural cleft.

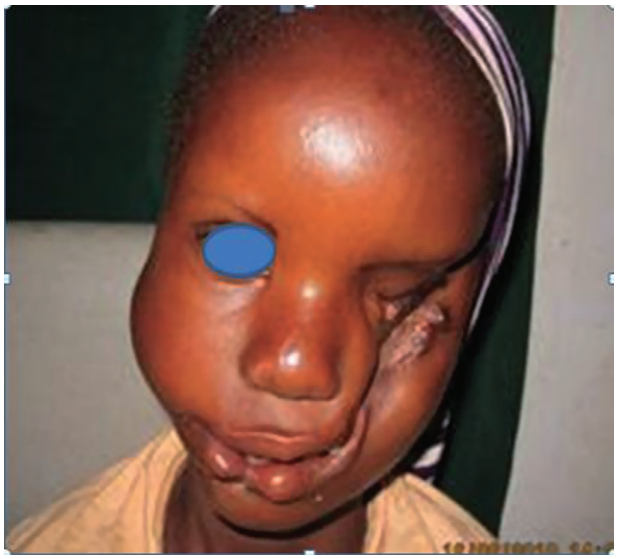

Figure 4 Photograph showing v-shaped lower-lip cleft and oblique facial cleft associated with eye deformities.

part of the cleft was repaired with Z-plasty, while the closure of the labiomaxillary cleft was achieved by direct apposition of adjoining soft tissues in layers (Fig. 5). The surgical correction of the retro-positioned left eye and the lateral canthus are yet to be carried out.

Discussion

ABS is a known cause of fetal malformations, and the anomalies range from mild deformities to severe ones that may be incompatible with post-natal life. The

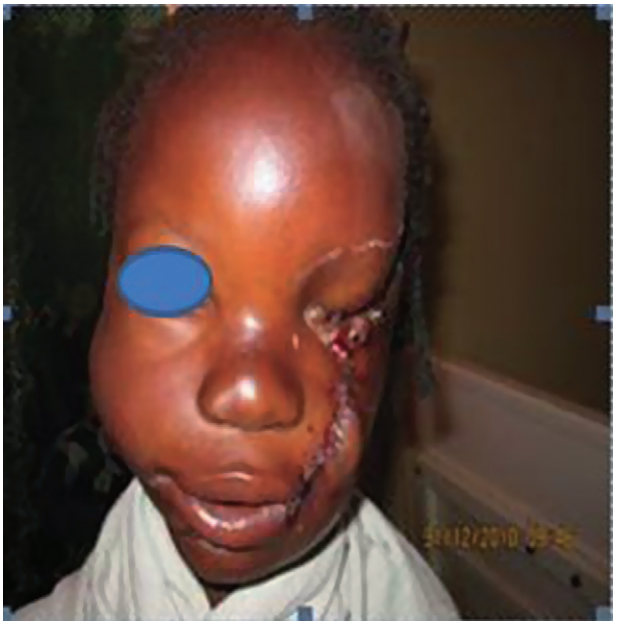

Figure 5 Photograph showing repaired lower-lip cleft, right-sided commissural cleft and oblique facial cleft.

exact incidence is unknown, but reported series suggest that they occur in between 1 in 1200 and 1 in 1500 live births[5] and are commoner in the early pregnancy[6]. According to the extrinsic theory in the aetiology of ABS, the disruption of the amnion allows the embryo or fetus to enter the chorionic cavity and contact the chorionic side of the amnion. Fetal parts may then become entrapped by the fibrous septa that traverse the chorionic space. Entanglement of fetal parts is random and the slash defects so created are non-embryologic in distribution. Compression from these bands will then result in deformities of the developing extremities, trunk and head[7]. In these our case reports, the clinical presentations of digits and toes amputation, leg constricting band with its associated lymphoedema, facial band and clefts could be a result of local compression or adhesion. Therefore, the present case reports may further support the concept of extrinsic theory in the aetiology of ABS[8]. The severity of the band compression on the developing fetus could determine the morphology of the deformity. When bands are superficial, only skin indentation occurs, as it was noted in case 2 on the right facial region. Deeper circumferential bands may cause lymphatic obstruction leading to oedema distal to the band, as shown in case 1 (Fig. 2) which may eventually be associated with lymphatic compromise. Those limbs with lymphatic inadequacy are vulnerable to repeated cellulitis[8]; however, in case 1 with profound lymphatic oedema,

no such clinical history was elicited even though in our society the habit of patients not giving complete clinical history may not be ruled out. Tighter constricting bands have also been noted to cause the narrowing of underlying bone or the amputation of extremities[8]. Therefore, the presence of a circumferential band on the leg with associated extremity anomalies should make this case a good example for the aetiological debate on ABS.

Although severe craniofacial abnormalities caused by ABS are often incompatible with life, a number of these children may still survive even till school age with the accompanying facial anomalies, as seen in these current cases. It has been observed that the presence of amniotic bands in conjunction with an orofacial cleft is part of the clinical features of this syndrome together with other visceral and extremity defects[7,9]. The oblique facial cleft, right-sided commissural and lower-lip clefts with the irregularly distributed amniotic bands in the facial region, as seen in case 2, is a rare and unique one because of the combination of three Tessier clefts[10] presenting in a single patient (Tessier nos. 5, 7 and 30). To the best our knowledge, this is the first case to be presented in our region (Nigeria). This further strongly supports the hypothesis that amniotic bands might have been the aetiological factor, especially with the presence of lateral oro-ocular cleft[11]. These atypical clefts are quite different from those usual ones that occur as a result of the failure of embryological ectodermal and mesodermal structures to migrate or converge in the midline.

The corrective surgical procedures for patients with ABS may range from minor to complex, and the outcome depends on the severity of the deformations. In cases of limb amputation, ambulation is possible with the aid of prosthesis. In our first case, with combined upper-lip cleft and fingers and toes amputations, we recommend prosthetic rehabilitation, but such facilities may not be readily available in a low-resource hospital setting like ours. However, the lip defect was successfully repaired by the Millards rotation advancement procedure[12]. The extensive craniofacial defect of the second case required an advanced facial reconstructive procedure, especially Tessier no. 5 (oblique facial cleft) with the accompanying orbital defects. This is a rare congenital malformation that begins in the upper lip just medial to oral commissure, extending across the maxilla to the infra-orbital rim and floor of the orbit. In an extensive involvement of the orbital region, orbitotomy may be required and during the repair process, certain goals are to be achieved. These include reconstructing the lower eyelid, repositioning the lateral canthus, closure of the labiomaxillary cleft and restoration of the skeletal deformity with bone grafts[12]. In this presenting case only closure of the labiomaxillary cleft was achieved (Fig. 5). This was a result of the inadequacy of both human and material resources. In fact, the problem of management was further aggravated by the rural environment in which our hospital is located. Da Silva *et al*[13]. highlighted the difficulties encountered in the reconstruction of this rare and challenging craniofacial malformation in their own centre, which is more advanced and better equipped. Clefts of the lower lip and oral commissure are also known to be an uncommon congenital deformity that is known to occur as a result of the incomplete merging of embryonic mesenchymal maxillary and mandibular prominences of the first pharyngeal arch[14]. However, the presence of a fibrous band at the base of the commissural cleft and at the apex of the v-shaped lower-lip cleft suggests that ABS may be the primary cause of these anomalies, probably by 'slash effect' disruption and not as a result of the incomplete fusion of developmental structures at designated embryological sites[7,15]. Therefore, we propose that these clefts occurred as a result of ABS. Z- or W-plasty are techniques recommended for the closure of commissural clefts, but these geometric techniques may cause additional scarring and are therefore unnecessary in the repair of transverse facial cleft[15]. Linear suturing technique was then the preferred choice in repairing the commissural cleft because of its simplicity while V-plasty satisfactorily corrected the lower-lip defect[13,16].

The presented cases with peculiar features of orofacial clefts, reduction defects of limbs, leg constriction band with lymphoedema and rare lower-lip cleft support the ideas that these deformities are likely related to ABS.

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