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Case Report

Congenital Constriction Band Syndrome: A Case Report and Literature Review

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Abstract: Congenital constriction band syndrome is a congenital disorder caused by entrapment of parts in fibrous band while in utero. A review of the operating register revealed only two cases in three years. We are presenting a case report of a patient and a literature review pointing out the rarity, aetiology, mechanisms, diagnosis and different treatment options. The condition has good prognosis.
Keywords: Congenital, band syndrome,

INTRODUCTION

Congenital constriction band syndrome or amniotic syndrome band is a congenital disorder caused by entrapment of fetal parts in fibrous band while in utero [1]. It is a rare congenital hand anomaly. However the incidence is on the increase in literature. It has different names which suggest that the real cause of the disease is not yet known. However there have been speculations that it could be due to entanglement by fragments of ruptured amniotic membrane in utero or it could be due to vascular disruption to certain parts of the body. After birth, the diagnosis is clinical but in utero, it is made by using 3D or 4D Doppler scan or MRI. Presentation could be varied, usually the limb deformities are accompanied with other body anomalies. Treatment is both conservative and surgical with good prognosis.

CASE REPORT

A 2-year-old child presented with unilateral congenital constriction rings at the distal third of the legs since birth. Mother attended antenatal clinic at a Government General Hospital. She had high grade fever in the first trimester of pregnancy for which she was treated for malaria with sulfadoxine- pyrimethamine. No history of bleeding, intake of herbal preparation nor drugs not prescribed by a clinician. No family history of hand and foot deformity. Both parents were public servants. Further evaluation of child did not reveal any other congenital abnormality. Multiple z-plasty was used for the surgery. The outcome was satisfactory as shown below. Follow up period was one year.

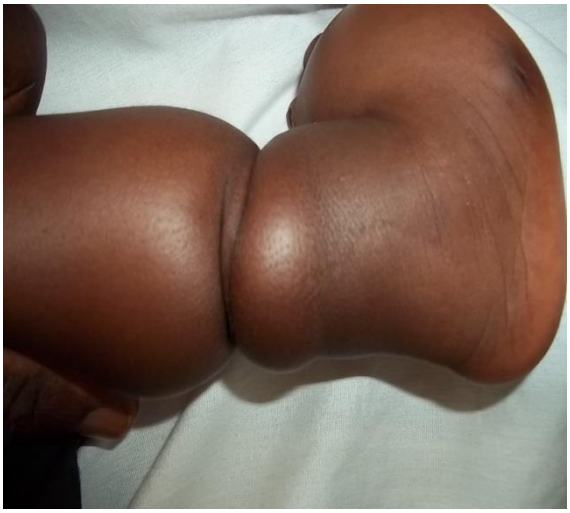

Fig-1: Congenital Constriction Band Syndrome Before Surgery

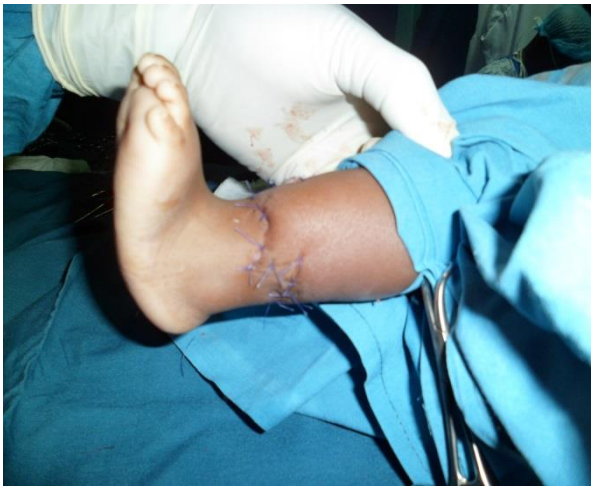

Fig-2: Congenital Constriction Band Syndrome During Surgery

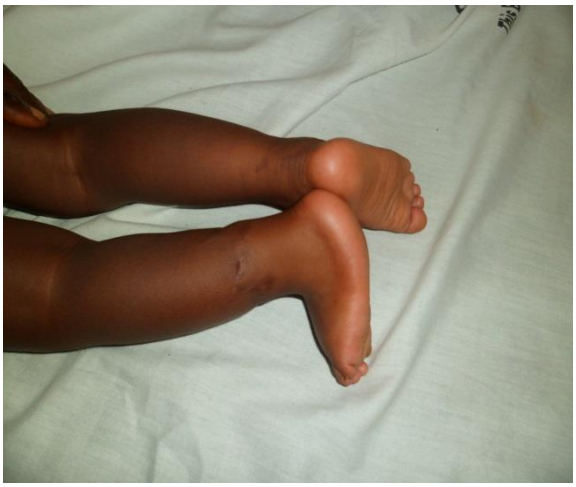

Fig-3: Congenital Constriction Band Syndrome After Surgery

DISCUSSION

It was once believed to have an incidence of 1:100,000, recent literature support the incidence today as 1:1200- 1:1500 births [2, 3]. No distinct sex predilection has been determined. Prenatal risk factors associated with amniotic band syndrome include prematurity (<37weeks), low birth weight(<2500g), maternal illness in pregnancy or placental haemorrhage in trauma, attempted abortion in the first trimester are highly associated findings [2]. Family history seldom reveals any direct inheritance pattern, since the syndrome occurs in no particular association with known genetic or chromosomal disorders. Karotypes are virtually always normal and the syndrome is almost always sporadic in nature. The incidence of malformation seen in the hand is two times as common as accompanied foot deformities [3].

Aetiologically, several factors have been suggested to be the cause of amniotic band syndrome. Firstly, damage to the amniotic sac has been implicated as a cause of amniotic band syndrome under the extrinsic theory. The exact cause of the amniotic tear is not known .Secondly, environmental factors have been implicated. Some researchers speculated that abdominal trauma which could lead to blunt trauma to the placenta has been shown to cause amniotic band syndrome in some cases .Thirdly, it has been reported that intense uterine contractions caused by a drug known as misoprostol (a prostaglandin E analogue) has resulted in amniotic band syndrome. It has been used to induce abortion. If the pregnancy continues after 6 to 8 weeks of gestation the infant may have the amniotic band syndrome. Finally, genetic factors may predispose infants to the development of amniotic band syndrome .The disorder is not expressed unless it is triggered or activated under certain circumstances such as particular environmental factors (multifactorial inheritance). Researchers have discussed the possibility that genetic factors influence the development of amniotic band syndrome in certain cases [4-6].

The mechanisms of congenital constriction band are complex and controversial. Several different theories have been proposed to explain the complex mechanisms that underlie amniotic band syndrome. The two theories are the extrinsic and intrinsic ones. The extrinsic or amniotic band theory for the development of amniotic band syndrome is that strands of tissue separate from the inner layer (amnion) of the amniotic

sac. The amniotic sac is composed of two main layers – the outer layer (chorion) and the inner layer called the amnion .According to this theory, amniotic band syndrome occurs when the inner layer (amnion)of the amniotic sac ruptures , thereby exposing the fetus to strands of fibrous tissue that may float freely in the amniotic fluid and still remain partially attached to the amniotic sac. The bands of tissue can wrap and constrict the fingers, toes, arms, legs, and other parts of the developing fetus. The symptoms that occur due to amniotic bands depend on specific part of the body affected by these strands of tissue and how tightly they have wrapped around the body part. The second theory is the intrinsic or the vascular disruption theory which was proposed because some researchers noted that while the above theory explains some cases of amniotic band syndrome it is insufficient to explain all cases. The external theory fails to explain why there is intact amniotic sac in some infants with amniotic band syndrome. Why there are a high number of malformations affecting internal organs in some cases and why some infants have defects of the body not affected by amniotic constriction bands. The intrinsic theory attributes the development of amniotic band syndrome to impaired blood flow to specific parts of the developing fetus (vascular disruption). The exact cause of impaired blood supply is not known but it is thought to be vascular injury leading to haemorrhage with eventual tissue loss in the affected part of the body. The intrinsic theory attributes the presence of constriction bands as a secondary effect of the impaired blood flow and subsequent damage to the fetus. In a 1987 article in the journal of Tetralogy, Webster et al demonstrated this theory of vascular disruption with an intact amnion in animal models [6].

The signs and symptoms of amniotic band syndrome vary greatly from one infant to another [7]. Some infants develop severe and even life-threatening complications. No two infants can develop the same number of or types of abnormalities. [8] Amniotic band syndrome develops at any point during the first 20 weeks of pregnancy [9]. Generally, the most severe complications occur when amniotic band syndrome develops early in the first trimester [10, 11]. Several different patterns have been identified with amniotic band syndrome. The three most common patterns are amniotic band syndrome characterized by one or more limbs being affected; limb-body-wall complex; and amniotic band syndrome characterized by abnormalities of the head and face (craniofacial abnormalities), certain birth defects of the brain and spinal cord and serious malformation of the arms and legs [12-14]. Most infants with amniotic band syndrome have some form of deformity of the arms and legs or fingers and toes. One or more limbs may be affected. Upper limbs may be affected more than the lower limbs [15]. Some infants may have bands of tissues that encircle or gradually constrict certain fetal areas. These bands can cause superficial constrictions without damage to the subcutaneous tissue. [16] Another pattern associated with amniotic constriction band is referred to as limb-body- wall complex [17, 18]. The infants have abdominal wall defects with defects of the arms and legs and other abnormalities. The infant may present with encephalocele, omphalocele, through a fissure in the abdominal wall (abdominoschisis) or the chest wall (thoracoschisis) and a variety of defects affecting the arms and legs [19]. Additional abnormalities can occur in infants with limb –body –wall complex [20]. A third pattern associated with amniotic band syndrome involves craniofacial abnormalities such as incomplete closure of the roof of the mouth (cleft palate), cleft lip, facial cleft, microphthalmia, cloanal atresia and malformations affecting the size and shape of the skull [21]. In severe cases the neural tube defects could lead to anencephaly. In some cases, serious complications of the arms and legs called terminal transverse limb deficiencies. In such cases affected infants may be missing a portion or all of a limb, ranging from one finger or toe to an entire arm or leg. In 1961, Paterson used a classification that is still widely used today [22, 23]. The classification is as follows: (A) Simple ring constriction; (B) Ring constriction accompanied by deformity of the distal part with or without lymphedema;(C) Ring constrictions accompanied by fusion of distal parts ranging from mild to severe acrosyndactyly, and (D)Uterine amputation.

Diagnosis of amniotic band syndrome after child birth is clinical. However, the diagnosis in intrauterine life is often difficult. 3D ultrasonography and Magnetic Resonance Imaging can be used for more detailed and accurate diagnosis of bands and the resulting damage or danger to the fetus. Although the earliest may be difficult to detect by ultrasound and are more often diagnosed by the effects they have on fetal anatomy. The visualization of amniotic bands attaching to fetus with restriction of motion, constrictions rings on the extremities and irregular amputation of digits with syndactyly, on prenatal ultrasonography is diagnostic. The latest ultrasound techniques of 3D and 4D ultrasound contribute to more sensitive prenatal diagnosis of the condition. In complicated cases, fetal MRI may be helpful.

Surgery is the mainstay of treatment. The goal of surgery is to release the constriction rings and acrosyndactyly so that the child may have an improved appearance and obtain basic hand functions such as grip and pinch. Acute exacerbation of the distal edema or nerve palsy is an indication for emergent excision for the ring to relieve the tension [7, 22]. Distal edema can rapidly progress in the first week after birth. In these cases, expeditious release can resolve this condition. Correction of constriction ring can be performed at 3months of age if necessary [9]. In cases of acrosyndactyly surgical treatment is performed between 6 and 12 months of age [9, 23].

Regarding anaesthetic management, the child should be evaluated properly for any hidden systemic anomaly during pre-anaesthetic checkup. Anaesthetist should always be prepared for difficult airway in such cases. In addition to deformities in amniotic band syndrome, Paediatric airway is challenging because of unique anatomy (smaller size vocal cord between 9-24, with an anterior angulation, a large and floppy epiglottis, a large occiput) and physiology (frequent upper airway obstruction under general anaesthesia, higher metabolism and faster desaturation during period of apnea). Difficult airway cart with different sizes of endotracheal tubes, laryngeal tubes, laryngeal mask airway, paediatric fiberscope, etc were kept ready although they were not used. Adequate covering of extremities with warm blankets, elevation of room temperature and warm infusion fluids were measures taken to prevent hypothermia. Extubation should be done when there is regular spontaneous breathing, vigorous movements of all limbs, good oxygen saturation and absence of significant hypothermia [24].

There are few surgical techniques that are used for the release of congenital constriction rings. The management strategy of constriction band syndrome depends upon the extent of the associated anomalies. Lately, there have been some attempts of prenatal constriction band treatment using fetoscopic laser cutting of amniotic bands before their compression on the fetus makes malformation [8]. Surgical correction of constriction ring includes the excision of the constricted ring and the excessive subcutaneous fibrous tissue and fat. This is followed by multiple z-plasty flaps to make the scar less conspicuous and also distributing the lines of tension [10, 25]. W- plasty [26], Mutaf procedure [27,29,30], triangular- plasty, quadrangular–plasty, replacement of z-plasty with direct closure, sine plasty [28] combined with removing fibrous groove and fasciotomy in two stages are some of the procedures being used. It should be noted that infants require symptomatic and supportive care. For example, in infants with incomplete development of the lungs and associated respiratory insufficiency, treatment may include oxygen support measures and other supportive therapies as required. Based on the complexity of the associated organ system involvement, a multi-disciplinary approach may be adopted.

The most severe postoperative complications of constriction ring release are the potential vascular problems. In the wrist, forearm arm, leg, thigh and the trunk, one-stage excision of constriction ring is acceptable because deep main arterial and venous vessels are protected [21,23]. However, the distal blood flow is supplied via perforators from the distal arteries [21]. A one-stage operation may fail to preserve the venae comitantes and endanger the distal circulation. Therefore constriction ring corrections of the digits are best performed in two stages at 6 to 12 weeks a part [30].

What is the expected outcome? The resection of constriction ring and the correction of acrosyndactyly can improve both appearance and function. In the case of severe deformity with distal finger amputations and acrosyndactyly, the limited functional benefit with reconstruction should be weighed against the risk of scar formation [29]. There is satisfactory prognosis.

In conclusion, congenital constriction band syndrome is a rare condition in our facility. However, literature search has revealed that the condition is on the increase. Successful results can be achieved with excision of the fibrotic band and the correction of the subcutaneous deficiency under the constriction. Infants with congenital constriction ring deformities should be referred for surgery early, particularly if there is vascular compromise.

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