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One-Stage Emergency Surgical Release of Amniotic Constriction Band in Streeter's Dysplasia with Clubfoot- A Case Report.

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Abstract

Streeter's dysplasia is a rare condition that occurs in 1 in 1200 – 15,000 live births. Timely intervention is necessary to save the limb in cases where there is vascular compromise. A 7-day-old neonate presented with Streeter's Dysplasia and clubfoot that was swollen and cyanosed. There was a circumferential amniotic constriction band in the lower third of the Left leg causing vascular compromise. There was another semicircular band in the mid-foot region. Single stage urgent circumferential band excision and multiple Z-plasties for skin cover were performed. Circulation to the affected portion was restored immediately. The swollen foot was treated by debulking and the clubfoot deformity by the Ponseti method.

Keywords: Streeter's dysplasia, Vascular compromise, Constriction ring, One stage release, Clubfoot, Debulking.

Introduction

Streeter's dysplasia, also known as amniotic band syndrome (ABS) is a rare congenital abnormality that is found in 1 in 1200 to 1 in 15000 live births. It is a sporadic condition characterized by constriction rings. It comprises a wide spectrum of clinical abnormalities mainly occurring in the limbs that may be associated with deformities, acrosyndactyly and amputations [1]. The incidence of clubfoot in Streeter's dysplasia varies from 17 to 23% [2, 3]. Baker and Rudolph suggested a triad associated with the constriction band comprising of clubfoot, syndactyly and lymphoedema beyond the constriction band [4]. Other associated anomalies include tibial pseudoarthrosis, bowing of the tibia, facial cleft lip and palate, renal and cardiac abnormalities [5]. The earliest account of ABS was given by Hippocrates, who described the condition and postulated the theory of amniotic band as a causative mechanism responsible for the constriction, deformities and auto-amputations [6]. George Streeter (1930) introduced his theory of germ plasma defect as a cause of this condition [7]. Patterson (1960) described the lack of mesodermal development in the area of constriction band [2]. Torpin (1965) reintroduced the original theory proposed by Hippocrates that freely floating bands in the amniotic fluid caused by the ruptured amniotic membrane wraps around the limbs and other body parts leading to this condition [8]. This notion has since been confirmed by 3D and 4D ultrasound techniques [9]. Resolution of amniotic bands do occur spontaneously

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Figure 1: Circumferential constriction band in the lower third of the leg, semicircular band in the mid-foot, associated clubfoot deformity and cyanosed foot.

in-utero. Serial ultrasound studies on a patient have shown the in-utero process of limb strangulation and subsequent gradual disappearance of an amniotic band attached to a fetal elbow [10].

Treatment is usually surgical by one or two-stage resection of the band and skin cover by Z or W-V plasty [11,12]. Ponseti technique is considered as an effective treatment modality in treating clubfoot associated with amniotic band syndrome [13].

Case Report

A 7-day-old neonate presented with a circumferential constriction band in the lower third of the left leg and an additional semicircular band in the mid-foot region. There was an associated club foot deformity (Fig. 1a). There was a history of cyanosis of the foot since birth. The sole and toes were blue, suggesting a vascular compromise (Fig. 1b). The foot was grossly swollen and the toes were somewhat embedded in the swollen foot. It was a bulky foot due to lymphoedema. X-ray showed soft tissue circumferential constriction in the lower third of the leg and another semi-circular constriction in the mid-foot region and altered talocalcaneal relationship, with reduced talocalcaneal angle on anteroposterior and lateral radiographs (Fig. 2).

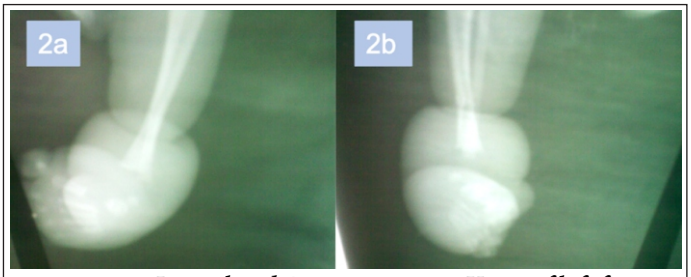

Figure 2: Lateral and Anteroposterior X-ray of left foot showing circumferential constriction in the left lower third of the leg and another at mid foot at the time of presentation.

The patient underwent treatment in a staged manner:

Stage 1 (Single stage band release)- The patient underwent a successful excision of the circumferential band urgently in one stage (Fig. 3a). The circumferential band was excised along with the deep fascia and the dissection went right up to the bone anteriorly (tibia) and tendons and muscles all around. Multiple Z plasties were done circumferentially and satisfactory skin cover was achieved (Fig. 3b, 3c). The foot and the toes turned pink immediately after the excision of the band. 10 days after the band release, sutures were removed. Circulation was satisfactory and foot and toes were pink. The foot, however, was too bulky for satisfactory casting. (Fig. 3d)

Stage 2 (De-bulking of the foot)- Debulking of the foot was performed, ten days after the suture removal by the primary surgeon. A transverse incision was made across the foot. Copious amount of accumulated fluid seeped out. A significant bulk of fibrofatty tissue was excised. Wound was sutured and foot splinted (Fig. 4). The purpose of debulking was to facilitate treatment of the clubfoot by Ponseti method and improve the cosmetic appearance of the foot.

Stage 3 (Deformity correction by Ponseti Method)- Clubfoot

Figure 3: Excised band and multiple z-plasties and foot after suture removal.

Figure 4: Debulking of the foot

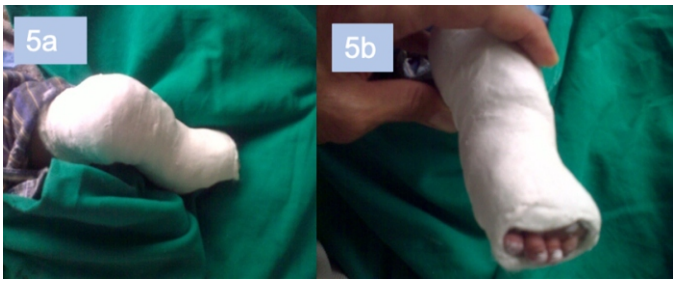

Figure 5: Deformity correction by Ponseti method

deformity correction began 10 days after de-bulking, once the wound had healed. Manipulation and casting were performed by Ponseti method at weekly intervals (Fig. 5a,5b). Percutaneous Tendo-Achilles tenotomy was undertaken after four casts. Foot abduction orthosis was provided following cast removal. Parents were instructed regarding foot abduction orthosis use. Patient compliance was satisfactory (Fig. 6).

Follow up at 8 years of age showed no residual deformity with a painless, functional foot that was free of callosities. The child could walk, run and perform all activities of daily living without limitation. Well healed scars were noted on the anterior, medial and posterior aspects of the left lower limb (Fig. 7a, 7b, 7c). Ankle and sub-talar movements were full and pain free. Calf wasting was 1.5 cm on the affected side. The foot was marginally bulkier (1 cm circumferentially). He wore normal footwear (Fig. 7d). Radiographs showed satisfactory appearances (Fig. 7e, 7f).

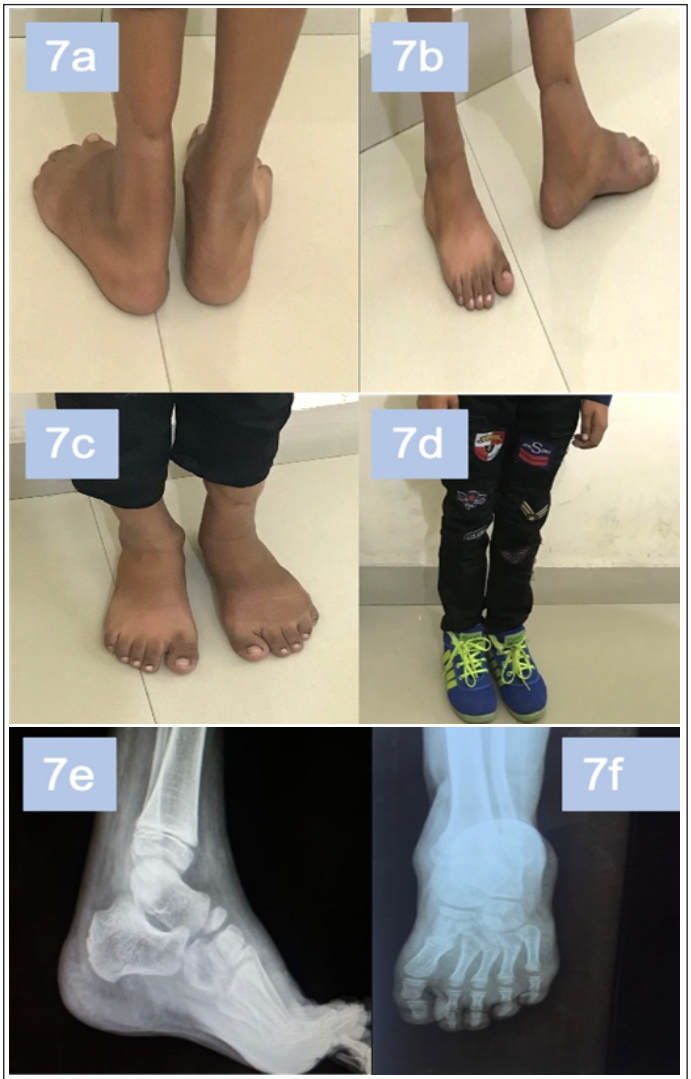

Figure 7: Clinical and X-ray images after eight years follow up

Figure 6: After band excision, de bulking of foot and clubfoot correction by ponseti method.

Discussion

Amniotic band syndrome is a rare congenital disorder resulting from entrapment of various fetal parts in utero by floating amniotic bands [8].

Patterson classified Streeter's dysplasia into four types [2]:

Type 1: Simple constriction ring

Type 2: Deformity distal to ring with or without lymphedema & neurovascular compromise

Type 3: Fusion of toes or fingers (Acrosyndactyly)

Type 4: Amputation (intrauterine)

Basheer et al presented a case of amniotic band syndrome in both the legs (type 1 on the left leg and type 3 on the right leg) in a one-day old neonate resulting in lymphedema, fracture in the right tibia but no vascular compromise. Single stage release provided satisfactory results but swelling recurred in the right foot after 6 months requiring repeat excision [14]. In this case, in addition to single stage circumferential band excision, debulking of the foot was performed that restored the normal contour of the foot and there was no recurrence of swelling. Waiswa et al presented a case of amniotic band syndrome type 3 in a three-month old infant involving the right leg with lymphedema, clubfoot and no neurovascular compromise. The lymphedema resolved 2 days after single stage release [15]. Naqvi et al presented a case with type 4 ABS in a four-month-old child involving both legs with vascular compromise bilaterally and lymphedema with associated clubfoot, auto-amputated right little finger and anterolateral bowing of the right tibia. Two stage release was performed with no postoperative complication. The edema reduced gradually and

wound healed completely with improved vascularity status 5. Nabi et al presented a case showing type 2 ABS in one-day-old neonate involving the left leg with compromised vascularity. Two stage release was done with no post-operative complication [11].

Controversy exists between one-stage and two-stage excision of the band. Single stage surgery appears to provide similar results to two-stage surgery [12]. One-stage release can lead to the recurrence of the band in a patient with severe lymphoedema [14]. There was no recurrence of the band in our patient at eight years' follow up. A good recovery can be achieved with one stage release, as illustrated by this case.

Conclusion

Streeter's dysplasia resulting in lymphoedema and vascular compromise requires urgent surgical excision of the constriction band. Timely intervention is important in avoiding complications and restoring function. Ponseti method was effective in correcting the clubfoot and the debulking procedure in restoring normal foot contour.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his/her consent for his/her images and other clinical information to be reported in the Journal. The patient understands that his/her name and initials will not be published, and due efforts will be made to conceal his/her identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil; **Source of support:** None

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