



AMNIOTIC BAND SYNDROME: WHERE MEMBRANE OF LIFE BECOMES BAND OF DEATH

General Surgery

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ABSTRACT

Amniotic band syndrome is a rare congenital disorder that causes multiple malformations in the fetus due to fibrous bands from the disrupted free-floating blind-ending amnion with a complete chorion. A 3 yr old female child presented to plastic surgery OPD with complaints of nonhealing ulcers over the right foot from 2 months and swelling of the right foot from 15 days with no history of trauma. All necessary investigations required for surgery, X-ray, and Doppler of the right lower limb were done. Surgery was planned and constriction band was released up to deep fascia and multiple Z-plasty was done in 2 stages with 4.0 prolene. The post-operative period was uneventful and follow-up done for 1 yr showed excellent cosmetic appearance with no ulceration or swelling.

KEYWORDS

Amniotic Band Syndrome, Constriction Band, Z-plasty.

INTRODUCTION

Amniotic band syndrome (ABS) is a sequence of congenital malformations in the fetus due to fibrous bands ranging from constriction rings, major craniofacial and visceral defects to even fetal death.^{1,2} ABS is not proved to be associated with any genetic or hereditary predisposition. Two main pathogenic theories are proposed in the formation of ABS i.e., exogenous and endogenous theory.³ The endogenous/intrinsic theory was proposed by Streeter and it proposes anomalies in germ cells resulting in defects.⁴ The exogenous theory postulates the intra-uterine disruption during pregnancy followed by events involving the rupture of amnion. The separation of amnion from chorion produces thin strands that entangle digits and toes.^{4,5} Multifactorial process involving vascular compromise is the principal pathogenic stimulus followed by fibrous bands.⁶ Accurate prenatal diagnosis will help in the management of present pregnancy and counseling of future pregnancies.⁷

CASE REPORT

A 3-year-old female child presented to plastic surgery opd with complaints of non-healing ulcers over the right foot from 2 months and swelling in the right foot from 15 days extending upwards to 1/3 rd of the right lower limb. Ulceration and swelling were spontaneous in onset with no history of trauma. On examination, there is a shortening of the right lower limb when compared to the left lower limb with the complete absence of toes. A circumferential band of scarring is seen 7cms above the right ankle joint.



Figure-1 Preoperative image showing constriction band on right lower limb



Figure-2 X ray of the right lower limb in AP and lateral views.

Surgery for the release of the constriction band was planned and all necessary investigations required were done. X-ray of the right lower limb showed a soft-tissue defect without any bony abnormality at the site of the constriction band. Doppler study done was normal. Under General Anaesthesia, constriction band was released up to deep fascia and multiple Z-plasty was done in 2 stages. In the first stage, the constriction band was released posteromedially followed by multiple Z-plasty with 4.0 prolene. The postoperative period was uneventful.

After 6 weeks, in the second stage, the constriction band was released up to deep fascia anterolaterally and multiple Z-plasty was done. The postoperative period was uneventful with suture removal after 1 week in both stages of surgery. Postoperative X-ray and Doppler done to the right lower limb are normal. Follow up done after 6 months and 1 year are normal with good cosmetic appearance.



Figure-3 Intraoperative image showing Z plasty



Figure-4 Follow up image after 1 year

DISCUSSION:

Although 'Amniotic Band Syndrome' is a rare congenital disorder with an incidence of 1 in 15,000 live births, several cases have been reported. The pathogenesis and etiology are still unclear. The main goals for the treatment of the constriction band are functional improvement and better cosmetic appearance.⁸ Timing of repair is a key factor for functional outcome.

Choulakian et al⁸ described the excision of constriction band and direct closure, allowing the fat to reposit naturally.

Muguti et al⁹ described single-stage excision of constriction band and multiple Z-plasty repair. No vascular insufficiency was reported in the postoperative follow-up period.

Napiontek et al¹⁰ described the management of constriction band by multiple Z-plasty in 2 stages. He proposed 2 stage repair to prevent vascular insufficiency.

In the present case, surgery for constriction band is done in 2 stages by multiple Z-plasty. The postoperative period was uneventful. Follow-up done for 1 year showed good cosmetic and functional outcomes.

CONCLUSION:

1. Amniotic band syndrome is a rare congenital disorder associated with multiple malformations in the post-natal period.
2. Prenatal diagnosis helps in the recognition of fetal abnormalities in utero, thereby providing sufficient time for management and counseling.

3. The constriction band in the post-natal period may result in vascular compromise and its complications. The early repair was advised to prevent complications from vascular insufficiency.
4. Z plasty was done in many cases to release the constriction band. The repair was done in 2 stages to prevent vascular insufficiency.
5. Foetal surgery is done nowadays, where, the constriction band is released in utero to prevent postnatal complications.

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