

CASE REPORT

Congenital Constriction Band Syndrome

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Abstract

Congenital constriction bands are anomalous bands that encircle a digit or an extremity. Congenital constriction band syndrome is rare condition and is mostly associated with other musculoskeletal disorders. We report such a rare experience.

Keywords

Congenital Constriction Band Syndrome, Amniotic Band Syndrome, Constricting Rings, Streeter Dysplasia

Introduction

Congenital bands, also called ring constrictions, Streeter dysplasia, amniotic bands, annular defects are anomalous bands that encircle, either partially or completely a digit or an extremity(1). Incidence (2) of this condition in new born is 01%. These may involve just the skin but they can be as deep as the underlying skeletal structures and some have caused subtotal or total autoamputation (3). The part distal to the band may appear to be entirely normal, or it may be edematous or even gangrenous. Additional musculoskeletal disorders that may be present include club foot, synactactly or acrosynactactly, hypoplastic nails, hypoplastic fingers, pseudoarthrosis of underlying bones, absence of bones, peripheral nerve defects, distal lymphodema, intrauterine amputations, cleft lip and cleft palate and umbilical hernia (4). Etiology of this syndrome is unknown. This syndrome may be caused by prenatal environmental factors and it appears to be result of excessive contraction of the uterine muscles and hemorrhages from marginal blood sinus. Being a rare condition and mostly associated with other musculoskelton disorders, it is worth reporting.

Case

A boy aged three years borned with full term normal delivery, presented to our hospital for treatment of constriction ring at junction of proximal and middle third

of left leg. This boy was having multiple musculoskeletal defects and constriction bands in other parts of body showing that he was suffering from constriction band syndrome.



Fig 1: Showing constriction ring in left ring finger



Fig2: Showing constriction ring in left lower limb.

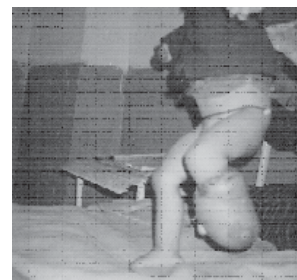


Fig3: Showing release of constriction limb.

Left leg was swollen distal to constriction ring. Soft tissues distal to it were viable. Patient was ambulatory.

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Posterior tibial artery could not be felt at the constriction site. Sensations distal to the constriction ring were intact. Roentengogram of lower extremity revealed marked swelling of the soft tissues. Left foot was having congenital amputation of the toes and was in attitude of Congenital Talipes Equino Varus Deformity (CTEV). There was discharging wound on the anterior aspect of left ankle.

On left hand there was also constriction band on the middle phalynx of ring finger with a absent left little finger. Skin distal to the band was viable with normal voluntary motion at distal interphalangeal joint and was having normal sensations.

Right foot was having congenital amputation of some toes and rest of toes were hypoplastic. Right hand was having acrosyndacty and hypoplastic fingers and thumb. We managed our case by excising the constriction band and multiple Z-Plasty.

Discussion

Patterson (2) in study of fifty two patients of congenital constriction rings had reported two below knee amputations in addition to other musculoskeletal defects. Patterson' classified extremity deformities in ABS into 4 types depending upon the presence of simple ring constriction, by fusion of the distal bony parts, with or without lymphedema, fusion of soft tissue parts, intrauterine amputations

Zych, *et al* (5) in 1983 reported a case of involvement of congenital bands, pseudoarthrosis & impending gangrene of leg. They peromed multiple Z-plasties reported the limb salvage. Greene WB (1) in his study advised a one-stage release for circumferential congenital

constriction bands which was performed in four extremities (three patients). No wound problems occurred, even when there had been marked swelling of the extremity distal to the band. The one-stage release facilitated postoperative care, and there was no need for additional periods of anesthesia or for additional operations, which are necessary when this problem is treated with a release performed in two or three stages.

Samra *et al* (3) in 2006 reported a case severely constricting amniotic band a threatened lower extremity in a neonate they peromed multiple Z-plasties reported the limb salvage after a 6-year functional follow up.

This case of constriction band syndrome was reported for the first time in our hospital. Although this case had been reported by various under mentioned studies but the number of associated musculoskeletal deformities was very high in our case.

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