

Congenital Constriction Ring Syndrome Associated With Club Foot And Contralateral Absence Of Part Of Tibia And Fibula- A Case Report

M Moosa, M Wani, I Wani, A Bashir, A Hussain, B Mir, M Halwai

Citation

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Abstract

Congenital constriction ring syndrome also known as streeter bands or dysplasia, annular grooves or defects, amniotic band syndrome and intrauterine amputation is a name given to a cluster of congenital malformations grouped together, believed to be caused by fibrous amniotic bands tightly tied around the limbs or digits. It is characterized by syndactyly, hypoplasia, brachydactyly, symphalangism, symbrachydactyly, club foot, cleft lip, cleft palate, cranial defects. We present a case, 4 year old male child with bilateral lower limb constriction rings with a club foot deformity in one and absent distal tibia and fibula with a rudimentary foot in another limb.

CASE PRESENTATION

A male child, Asian Indian in origin was brought to us at the age of 4 years with constriction rings bilateral legs (fig1). Parents of the patient had never sought a consultation before for reasons of ignorance and poverty. The malformations were present since birth of the child who was born by a normal vaginal delivery at full term with cephalic presentation. The child had one constriction ring on right lower limb with club foot deformity (fig 2) and two constriction rings on left lower limb with a rudimentary foot (fig 3). Skeletal survey of the patient showed absent distal tibia and fibula on left side (fig 4). The patient was operated for his club foot deformity on right side using modified McKay procedure and is still under our follow up.

Figure 1

Figure 1- bilateral constriction rings in lower limbs



Figure 2

Figure 2- Right leg with a constriction ring and associated club foot



Figure 3

Figure 3-left lower limb with two constriction rings and a rudimentary foot



Figure 4

Figure 4- X-ray of the patient's lower limbs showing bony anomalies.



DISCUSSION

Congenital ring syndrome or congenital constriction band syndrome is synonymous with the terms like streeter bands or dysplasia, annular grooves or defects, amniotic band syndrome and intrauterine amputation. Patterson [1] reported an incidence of per 15,000 births. Flatt's [2] in his series reported an incidence of 2 % of constriction bands. Kino [3] suggested the cause of this syndrome to be an external affect of amniotic adhesions formed in utero after hemorrhages in the distal rays. Patterson [1] and Streeter[4] theorized failure of development of subcutaneous tissue in the same manner that normal skin creases are formed.

Patterson [1] included four types of deformity in congenital ring syndrome : (1) a simple ring usually occurring transversely , but occasionally obliquely, around the limb or digit; (2) a deeper ring often associated with abnormality of the part distally, usually lymphoedema; (3) fenestrated syndactyly or lateral fusion of adjacent digits at their distal ends with proximal fenestration between the intervening skin and soft tissue; and (4) intrauterine amputation , in which the

soft tissues are more affected than the bone . Syndactyly, hypoplasia, brachydactyly, symphalangism, symbrachydactyly have been reported in 80% of patients with congenital constriction rings and club foot, cleft lip, cleft palate and cranial defects have been reported in 40% to 50% of patients with this syndrome. These malformations are usually asymmetrical. With deeper rings, the superficial blood vessels that run across the ring are absent, although deep vessels are intact. Digits distal to the rings may be shortened or completely amputated.

The prevalence of clubfeet with constriction bands ranges from 12 to 56 % [5-7]. Cowell and Hensinger in their series of 14 patients reported clubfeet among 25 patients with congenital constriction band syndrome [8]. The location of constriction bands are divided into 4 zones. Zone 1 bands occur between the greater trochanter and the knee. Zone 2 bands occur between the knee and the ankle. Zone 3 bands occur between the ankle and metatarsophalangeal joints. Zone 4 bands are limited to toes. Severity of bands is also considered. Grade 1 bands are subcutaneous, not to the level of fascia. Grade 2 bands are to the level of fascia and not compromise the circulation to the distal extremity. Grade 3

bands are to the level of fascia, such that lymphedema or circulatory compromise necessitates surgical release. Grade 4 bands include all congenital amputations [9].

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Author Information

Mohammad Moosa, MS Orthopaedics

Department of orthopedics, Government hospital for bone and joint surgery

Mubashir Wani, Post graduate student

Department of orthopedics, Government hospital for bone and joint surgery

Iqbal Wani, MS Orthopaedics

Department of orthopedics, Government hospital for bone and joint surgery

Arshad Bashir, MS Orthopaedics

Department of orthopedics, Government hospital for bone and joint surgery

Anwar Hussain, MS Orthopaedics

Department of orthopedics, Government hospital for bone and joint surgery

Bashir Mir, MS Orthopaedics

Department of orthopedics, Government hospital for bone and joint surgery

Manzoor Halwai, MS Orthopaedics

Department of orthopedics, Government hospital for bone and joint surgery