Research Paper

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# Congenital Constriction Ring Syndrome in Lower Limb-A Rare Entity

Medical Science

**KEYWORDS :** Constriction ring syndrome, Congenital anomalies, lower limb

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## ABSTRACT

*The congenital constriction ring syndrome is a rare entity reported with an incidence of about 1 in every 1500 to 15,000 live births. There is no known racial and sex predilection. Familial occurrence is rare, and it is believed that there is no genetic predisposition to this condition.The aetiology of the syndrome is not known; the probable theories that could explain its occurance are: the intrinsic theory, the extrinsic theory and the intrauterine trauma theory. We report a 4 year old male child with congenital constriction ring syndrome in right lower limb without any obvious associated anomalies.*

**INTRODUCTION**

The congenital constriction ring syndrome embraces a group of abnormalities which occur in a variety of combinations, mainly affecting the limbs and rarely the trunk and head. It is associated with fibrous bands that encircle, strangle and even amputate some parts of the foetus.[1] Patterson [2] classified the varieties as follows: (a) simple constriction rings, (b) constriction rings accompanied by deformity of the distal part, with or without lymphoedema, (c) constriction rings accompanied by fusion of distal parts, ranging from mild to gross acrosyndactyly and (d) intrauterine amputations.

According to Patterson, [2] to diagnose this syndrome a patient must have two or more of the above. The deformities are the result of a cascade of events that follow an intrauterine disruption. [3] The aetiology of the syndrome is not known; three main theories attempt to explain the specific cause of the disruption: the intrinsic theory, [4] the extrinsic theory [5] and the intrauterine trauma theory. [6] The condition is not uncommon in Ghanaians but data on it is quite scanty.

**CASE REPORT**

A boy aged four years born by full term normal delivery, constriction ring at junction of proximal and middle third of right leg. Right leg was swollen distal to constriction ring. Soft tissues distal to it were viable. Patient was ambulatory. Posterior tibial artery could not be felt at the constriction site. Sensations distal to the constriction ring were intact. He presented to our hospital because of pain in the affected limb along with edema.

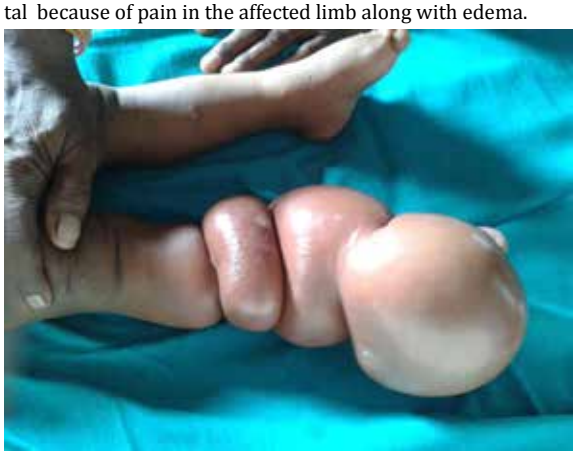

**Fig.1 Congenital constriction of right leg**

**DISCUSSION**

The incidence of constriction ring syndrome varies from 1 in 1500 to 15,000 live births. There is no known racial predilection, and the male preponderance in this study is at variance with the equal sex affectation in other reports. [7] Familial occurrence is rare, and it is believed that there is no genetic predisposition to this condition. [3]

Congenital constriction ring or congenital constriction band syndrome occurs when deep cutaneous creases encircle a limb as if a string were tightly tied around the part. Its frequent association with congenital amputations and acrosyndactyly led to this malformation's designation as a syndrome. Other terms used in the literature include annular band, Streeter dysplasia, intrauterine or congenital amputations, acrosyndactyly and fenestrated syndactyly. [8]

In 1832 Montgomery described a five-month aborted foetus with fibrous bands passing from both hands to the legs and wound around deep grooves in the limbs. [9] Whether the intrauterine amputations and ring constrictions are due to amniotic bands and similar circumferential constricting agents or due to interference with development remains controversial.

The pathogenesis of this disorder is not known for certain, but a number of theories attempt to suggest the aetiology of congenital constriction ring syndrome. The intrinsic theory [4] is the earliest, and it states that these deformities were the result of a "defective germ plasm", within the embryo. Streeter [4] believed that the bands represented macerated sheets of epidermis and the residual of defective local tissue. This theory is supported by localised areas of involvement within the limb and the presence of systemic and internal visceral anomalies.

The second (extrinsic) theory was first described by Torpin. [5] In this theory, the lesions are caused by the strangulating action of the mesodermic bands which occur due to an early rupture of the amnion. After rupturing, the amniotic sac stops growing normally and separates itself from the chorion. The amniotic fluid escapes, causing oligo-hydramnios. The foetus leaves the amniotic sac and lies next to the chorion. Multiple mesodermic bands issuing from the chorionic face of the amnion strangulate the fingers, the limbs and the cranium, inducing the typical lesions. Lack of familial incidence, the transverse disposition of the lesions, the exclusive limitation of the lesions to long digits/limb, the delivery of amputated parts, the presence of engrafted amputated parts on different sites of the body and the absence of associated internal malformations, all support this theory. [5]

The third (intrauterine trauma) theory postulated by Kino [6] believed that congenital constrictions, amputations and acrosyndactyly are caused by intrauterine trauma during pregnancy, which disrupts blood supply to the marginal sinuses of the limb plate.

Surgical correction of a constriction ring should aim at preventing or alleviating distal lymphoedema, separation of an

associated distal fusion and removal of an unsightly groove for the sake of cosmesis. Where intrauterine amputation of digits has occurred there may be opportunities to improve function by transfer of finger stumps from one position to another, deepening of web spaces or free-toe transfer.[12]

This case of constriction band syndrome was reported for the first time in our hospital. Although this case had been reported by various aforementioned studies with the number of associated musculoskeletal deformities, there was no any obvious abnormality in our case. However, detailed work-up of this patient could not be carried out because he did not turn up for the next follow up visit.

**CONCLUSION**

Congenital constriction ring syndrome is of uncertain aetiology and can cause morbidity in the newborn. The syndrome and its complications are amenable to corrective surgery with good results. Early intervention is desirable for a successful outcome.

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