Original Article

Pattern of constriction band syndrome

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ABSTRACT

Objective

To ascertain presentation of constriction band syndrome and its association with other congenital anomalies seen at our institution.

Patients and Methods

This retrospective study was conducted at the department of Plastic and Reconstructive Surgery, Hayatabad Medical Complex, Peshawar from January 2000 to June 2009. Twenty seven patients with constriction band syndrome were included in the study. All were admitted through the outpatient department. The location and degree of each constricting ring and associated anomalies were recorded. Surgery was performed in stages with an average interval of 3 to 6 months.

Results

Out of 27 patients, 11 were males and 16 were females. Age range was 1-18 years (mean 8.19 years). Upper limbs involvement was seen in 15 patients on the right side and 16 on the left side, with total number of 31 digits involved on the right and 38 digits on the left side respectively. The most commonly involved digits were middle and ring finger. Lower limbs were involved in 8 patients on the right side and 11 patients on the left side. Nine toes were involved on the right side and 11 on the left side. Most frequently involved toes were second and third toe. Intrauterine amputations were more common on the right side. The most common associated anomalies were syndactyly, acrosyndactyly, hypoplastic phalanges, lymphedema and Talipes Equino Varus.

Conclusion

Constriction band syndrome had varied presentation and is cause of morbidity in the newborn. We noted three new associated conditions in this syndrome previously not reported. These were strawberry hemangioma of the face, polydactyly and congenital first web contracture increasing the list of associated anomalies in constriction band syndrome.

The syndrome and its complications are amenable to corrective surgery with good results. Early intervention is needed for a successful outcome. (Rawal Med J 2010;35:).

Key words

Constriction band syndrome, syndactyly, polydactyly.

INTRODUCTION

Constriction band syndrome is an uncommon congenital abnormality with multiple disfiguring and disabling manifestations.[1-4] Constriction bands, also called congenital rings, Streeter dysplasia, amniotic bands and annular defects are anomalous bands that encircle, either partially or completely a digit or an extremity.[5] A wide spectrum of clinical deformities are encountered ranging from simple ring constrictions, minor digital defects to major craniofacial and visceral defects.[2-4,6] This syndrome was first recognized by Portal[7] in 1685. It occurs in approximately 1 in 1200 to 1 in 15000 live births.[4,6-10] Upper and lower extremities malformations are the most common and consist of asymmetric digital ring constrictions, distal atrophy, congenital intrauterine amputations, and acrosyndactyly and lymph edema.[2,4,5,7,11] Inheritance is sporadic, although association with other anomalies, such as club feet or cleft lip and palate has been described.[9] Patterson classified constriction bands into four types: (a) simple constrictions only; (b) constrictions with distal deformity; (c) constrictions with fusion of distal parts; and (d) intrauterine amputations.[9,10,12] The etiology of the syndrome is not known; three main theories attempt to explain the specific cause: the intrinsic theory, the extrinsic theory and the intrauterine trauma theory.[9,10] The condition is not an uncommon in our population but data is lacking. Aim of this study was to ascertain presentation of constriction band syndrome in our center and its association with other congenital anomalies.

PATIENTS AND METHODS

This retrospective study was conducted at the department of Plastic and Reconstructive Surgery Hayatabad Medical Complex, Peshawar from January 2000 to June 2009. Patients presenting with the features of the constriction band syndrome were included in the study. Those with congenital amputation of a limb were not included in the study unless they demonstrated constriction rings at sites other than the amputated limb. A total of 27 patients admitted through the outpatient department. Written informed consent was obtained from all patients. Detailed history was taken and complete physical examination was performed which specifically noted constrictions of the extremities including digits and accompanying anomalies. The location and degree of each ring was recorded and they

were classified into four types according to Patterson criteria.[12] X-rays of the affected parts in both anteroposterior and lateral views were taken. Surgical procedures performed included excision of constriction rings followed by Z-plasty. Patients with acrosyndactyly/syndactyly had their fingers or toes separated; the site of tissue loss was either closed by local flaps or graft. Most of these procedures were staged with an average interval of 3 to 6 months.

RESULTS

Out of 27 patients, 11 were males (40.74%) and 16 were females (59.25%). Age range was 1-18 years (mean 8.19). Family history of consanguinity was present in 6 patients (22.22%) while there was only one patient with positive family history of constriction band syndrome (3.70%).

Table 1. Sites affected by constriction band syndrome.

Upper limb	**Right side**	**Left side**	**Lower limb**	**Right side**	**Left side**
Fingers: Thumb	3	5	Toes : Great toe	1	2
Index	6	9	Second toe	3	3
Middle	8	10	Third toe	3	2
Ring	8	9	Fourth toe	1	2
Little	6	5	Fifth toe	1	2
Hands	1	2	Foot	0	0
Wrist	2	0	Ankle	1	0
Forearm	0	1	Leg	3	5
Arm	1	0	Thigh	0	1

There were 2 patients with type1 constrictions (7.40%), 8 patients with type2 (26.62%), 11 patients with type3 (40.74%) and 6 patients with type 4 constrictions (22.22%). The sites of constrictions in the limbs and digits and associated anomalies are given in table-1 and table-2.

Table 2. Associated anomalies in constriction band syndrome.

Areas affected	Right side	Left side
Hand anomalies		
Syndactyly	7	9
Acroyndactyly	4	6
Hypoplastic phalanges	5	5
Lymphedema	3	2
Foot anomalies		
Talipes equinovarus	2	3
Syndactyly	5	5
Acroyndactyly	0	1
Lymphedema	1	1
Other anomalies		
Strawberry hemangioma	1(cheek)	---
Polydactyly	1	1
Congenital first web contracture	1	1

Upper limbs involvement were seen in 15 patients on the right side and 16 on the left side with total number of 31 digits involved on the right and 38 digits on the left side respectively. The most commonly involved sites are shown in table 1. Intrauterine amputations involved both upper and lower limbs (Fig 1 and 2). The most common associated anomalies are shown in table 2.

Fig 1. Constriction bands involving thumb and ring fingers with associated syndactyly.

Fig 2. Constriction bands involving distal third of right leg.

In the lower limbs, a total number of 9 amputations (Fig 3).

Fig 3. Intra-uterine amputation of lower limbs.

Other anomalies we noted with this syndrome were Strawberry hemangioma (1), Polydactyly (2), and congenital first web contracture (2).

DISCUSSION

There was female preponderance in our study which is at variance with the equal gender affection in other reports.[13] Familial occurrence is very rare as seen in only one patient in our study and there is no known genetic predisposition.[14] The pathogenesis of this disorder is not known but a number of theories attempt to suggest the etiology. The intrinsic theory states that these deformities were the result of a ‘defective germ plasm’ within the embryo. In 1930 Streeter supported this theory by adding that the bands represents macerated sheets of epidermis and the residual of defective local tissue.[15] Extrinsic theory proposed

that the lesions are caused by the strangulating action of the mesodermic bands which occurs due to an early rupture of amnion.[4,10,16] The amniotic bands are composed of either acellular fibrous tissue or fibrous tissue contracting fibroblast, caused by squamous cells.[17] The third theory (intrauterine trauma theory) postulated by Kino[18] believes that congenital constrictions, amputations and acrosyndactyly are caused by intrauterine trauma during pregnancy which disrupt blood supply to the marginal sinuses of the limb space.

In our study, there was one case of positive family history of constriction band syndrome so far not reported in the literature. Similarly, family history of consanguinity was present in 6 (22.22%) patients, as reported by others.[19] These findings in our study suggest some hereditary or familial factors for this condition. Constriction bands mostly involved the upper limbs as noted by Mosses[19] and Kino,[18] while involvement of the great toe was least common in our study. The constriction bands involved mainly the long digits of hands and feet and they were more on the left side than the right as reported earlier.[10,18,19]

The numbers of associated anomalies in our study were lower than in other studies.[19] Other known malformations associated with constriction ring syndrome include craniofacial defects like cleft lip/palate and neural tube defects, leg length discrepancies and bone abnormalities.[4,7,10,20] We noted three new associated conditions with this syndrome previously not mentioned. These are strawberry haemangima of the face in one case, polydactyly and congenital first web contracture in two cases each, increasing the list of associated anomalies in constriction band syndrome. Mild simple constrictions may not require any treatment, while more severe constrictions and distal deformity can be treated with Z-plasty.[9,10] The procedure is staged, with only half the circumference corrected at the first operation to prevent vascular compromise.[9] Those with syndactyly/ acrosyndactyly are treated early to allow unimpeded growth of the fused digits.[9,10]

CONCLUSION

Constriction band syndrome is of unknown etiology and varied presentation and causes morbidity in the newborn. We noted three new associated conditions consisting of strawberry haemangima of the face, polydactyly and congenital first web contracture. The syndrome and its complications can be easily diagnosed clinically and are amenable to corrective surgery with good results. Early intervention is needed for a successful outcome.

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Received: June 21, 2010 Accepted: July 17, 2010

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