

Clinical Quiz

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A child with congenital constriction bands

Clinical Presentation

A 2-month old female baby was brought to the Orthopedic Outpatient Department with complaints of constriction bands in both lower legs since birth. Her parents also complained of deformities of both feet with absence and fusion of toes in her right foot. The child was a full term normal delivery. On examination, constriction bands were present in the lower legs bilaterally, along with bilateral clubfeet. There was also absence of little and great toes in her right foot, with fusion of second and third toes (Figure 1). The rest of the skeletal survey was normal.



Figure 1 - Clinical photograph of patient showing constriction bands in both lower legs (thin arrows) along with bilateral club-feet. Also note absence of little and great toes in right foot, with fusion of second and third toes (thick arrow).

Questions

1. What is the diagnosis?
2. What other congenital deformities are associated with this syndrome?
3. What is the management?

Clinical Quiz

Answers

1. The baby is suffering from congenital constriction band syndrome. It is also known as amniotic band sequence, Streeter's dysplasia, congenital bands or rings, and amniotic adhesions mutilations.¹
2. Congenital constriction band syndrome may be associated with clubfeet, syndactyly, acrosyndactyly, hypoplastic nails and fingers, pseudo-arthritis of underlying bones, absence of bones, peripheral nerve defects, cleft lip, cleft palate, umbilical hernia, microphthalmia, and hydrocephalus.^{1,2}
3. Treatment of constriction bands depends upon the depth of involvement of bands (superficial or deep). Superficial bands usually do not require any surgical release because they do not cause neurovascular deficit, distal swelling, or gangrene of the extremity. In such cases, surgical release is carried out only to improve cosmesis, especially in females.¹ Deep bands in contrast can result in neurovascular damage of inconsistent intensity, requiring surgical release to prevent progressive distal swelling, vascular compromise, and gangrene. Surgical releases comprises of Z-plasty, W-plasty, and excision of the constriction band with soft tissue reconstruction. The procedure can be carried out in one stage,³ or in sequential stages 6 to 12 weeks apart.⁴

Discussion

Congenital constriction band syndrome is a rare congenital abnormality characterized by a set of congenital birth defects caused by entrapment of fetal parts, typically an extremity or digits in fibrous amniotic bands while in the uterus. Male and females are affected equally with an incidence of 0.01%.⁵ Clinically, it may manifest as benign superficial constriction bands to in-utero, auto-amputation, or gangrene of the extremity requiring urgent amputation following birth. The part distal to the band may be normal or may be edematous. In our patient, bilateral clubfeet deformity was present distal to the constriction band along with absence of little and great toes, with fusion of second and third toes of the right foot. However, there was no distal swelling or neurovascular compromise (Figure 1). Congenital constriction band syndrome is due to incomplete split of the amniotic sac. Only amnion is ruptured leaving chorion integral. The fibrous bands of the ruptured amnion glide in the amniotic fluid, which surround and entrap some part of the fetus leading to congenital anomalies.⁵ Treatment consists of surgical release. The aim of the surgery for a congenital constriction band should be complete excision of the constricted tissue to curtail the risk of recurrence and conserve circulation.³

References

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