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Brief Communication

Congenital pseudoarthrosis of the tibia with localised gigantism in a case of congenital constriction band syndrome

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A 6-month-old male child was brought to our institute for deformity in both hands and feet noticed by the parents since birth. He had progressively increasing volume of the left lower limb and no movements of the ankle and toes. The child was born normally and there was no adverse perinatal history. The left lower limb showed a deep, circumferential constriction band near the lower-third of the leg with abnormal mobility at the site with gross enlargement distally. A few nubbins could be felt instead of the toes. No movements could be elicited from the ankle [Figures 1 and 2]. The limb was insensate distal to the constriction band. Pulsations of the dorsalis pedis and the posterior tibial artery could not be palpated. There were constriction bands in the other limbs without any concomitant swelling with acrosyndactylous digits. The right lower limb showed superficial constriction bands over the foot and calf, with minimal distal swelling. The right lower limb had normal sensations and movements. Radiograph of the left lower limb showed a cyst-like lesion in the distal tibia with a pathological fracture similar to type III of Boyd's classification [1] for congenital pseudoarthrosis of the tibia. Radiograph of the right lower limb was normal [Figure 3a and b].

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Although there was no actual neurovascular compromise in the left lower limb, the enlargement of the limb was so much and so grotesque that plastic surgical reconstruction would have entailed an extensive reconstructive process, with a minimal possibility of getting a functional and sensate limb. Considering this, a decision to amputate the left lower limb was taken after informed consent of the parents. The constriction bands of the other limbs were treated with multiple staged Z-plasties. The patient had an uneventful post-operative recovery and all surgical wounds healed well [Figure 4]. A decision regarding ambulation with a prosthesis would be taken later.

Amniotic constriction band syndrome is a group of disorders characterised by a wide range of congenital anomalies, including annular constrictions over the extremities, acrosyndactyly, talipes equinovarus, cleft lip and palate and hemangiomas.[2] According to Paterson's classification of constriction band syndromes,[3] our patient seems to be in type II, with distal lymphedema and sensory deficits. Closed pseudoarthrosis of the tibia, which has been noted earlier in congenital amniotic band syndrome,[1] usually requires no treatment other than adequate splintage. However, this was not possible here due to the associated severe anomalies. Neurocirculatory disturbance along with lymphatic obstruction of such magnitude has not been described before, although a similar case of neurodeficit without circulatory disturbances has been described earlier by Nambi *et al*.[4] In conclusion, this case is presented to discuss the end result of untreated congenital

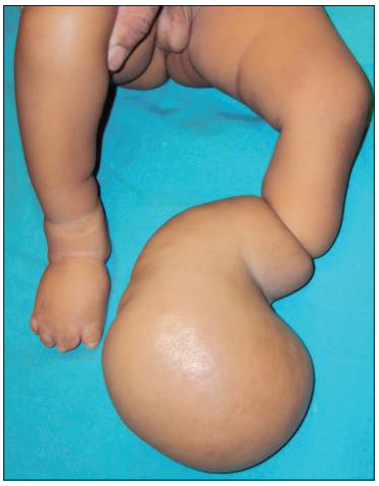

Figure 1: Clinical photograph of a 6-month-old child with constriction band syndrome and severe swelling of the left leg distal to a constriction band. Deformity can be seen at the distal one-third of the leg

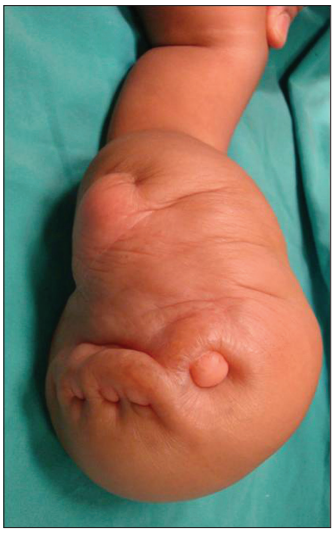

Figure 2: Clinical photograph of the left leg and foot showing severe swelling with nubbins visible. There was no discernible ankle and foot

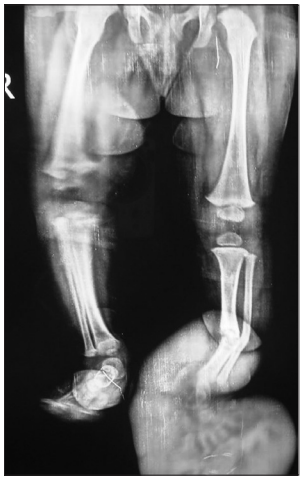

Figure 3a: Radiograph of both lower limbs – anteroposterior view showing cystic lesion in the middle one-third–distal one-third junction of the left tibia with pathological fracture and markedly enlarged soft tissue shadow over the distal third of the leg and the foot. The right lower limb was radiologically normal

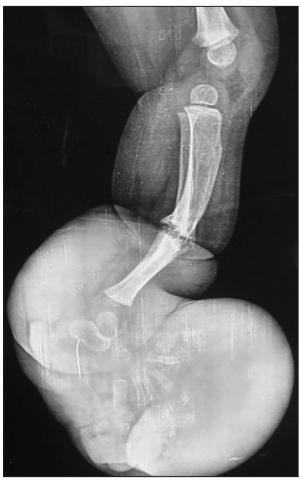

Figure 3b: Radiograph of the left lower limb – lateral view showing cystic lesion in the middle one-third of the tibia with pathological fracture

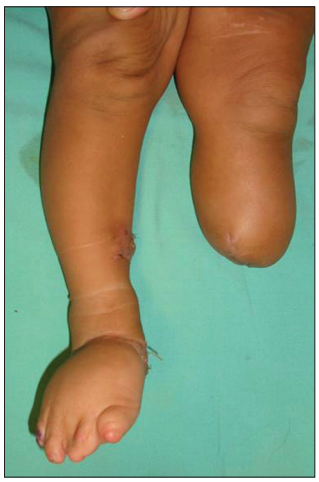

Figure 4: Final clinical photograph of the child with well-healed amputation stump and healing Z-plasties of the right leg

constriction band syndrome as well as to present this unique combination of severe lymphedema leading to localised gigantism and congenital pseudoarthrosis of the tibia in the setting of Streeter's dysplasia.

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