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Unusual Case of Tibial-Hemimelia With Split Foot

Medical Science

KEYWORDS : Tibial hemimelia, Split foot, Jones type 4

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ABSTRACT

Tibial hemimelia is a rare congenital anomaly which is characterized by an aplastic or hypoplastic tibia. This condition actually represents a spectrum of anomalies, ranging from total absence of to mild hypoplasia of tibia. We describe a varied presentation of jones type 4 tibial hemimelia with deformed split foot. To our best knowledge such a deformed association has not been reported before.

Introduction:
Tibial hemimelia is a rare congenital anomaly with an incidence of one per million. This condition actually represents a spectrum of anomalies, ranging from total absence of the tibia to mild hypoplasia of the tibia with intact fibula. It is characterized by marked shortening, bowing of the involved leg, flexion contracture of knee with a skin dimple overlying the proximal tibial region with rigid club foot We report a varied deformation of ectrodactyly with tibial hemimelia.

Case report:
7 month old boy born out of non-consanguineous parents presented with deformity of the left foot along with shortening of the left leg since birth. Pre-natal and neonatal history was uneventful. Child had normal developmental milestones except delay in walking. Multi system congenital anomalies were excluded. On examination of the left lower limb, the limb was short in comparison with the normal limb with split foot. Great toe and second toe were facing backwards and the other toes facing downwards. Heel was small and hypoplastic. On palpation of the bony landmarks, both malleoli and calcaneum were palpable with no talus [Figure:1]. Movements of the ankle were near normal with no distal neurovascular deficit. Radiological evaluation revealed Jones type 4 and kalamachi type 3 tibia hemimelia with inferior tibio-fibular diastasis with talus in between them[Figure:2]. Parents were counselled and syme's amputation was performed. At present, child is two years and walks without support with prosthesis.

Discussion:
Tibial defects are most often unilateral and sporadic. Tibial hemimelia is also associated with different syndromes, which are autosomal dominant in inheritance. Tibial hemimelia has been described with Tibial hemimelia-foot polydactyly-triphalangeal thumbs syndrome, Tibial hemimelia-diplopodia syndrome, Tibial hemimelia-ectrodactyly syndrome, Tibial hemimelia-micromelia-trigono brachycephaly syndrome, Pre axial mirror polydactyly, Split hand/foot syndrome-ectrodactyly.

Ectrodactyly (split foot) has been very well known associated with tibial hemimelia. Autosomal dominant inheritance with variable phenotype. It ranges from isolated minor malformations such as anonychia or syndactyly of fingers or toes, to major anomalies such as split hand/foot with or without tibial aplasia[1,2].

There are several classifications based on radiological description present in literature. Most commonly used is the jones and kalamachi. In Jones type 1, complete absence of tibia with or without the presence of distal femoral physis. In Type 2, presence of only proximal part of tibia. Type 3 describes the presence of distal part of tibia and type 4 is tibio-fibular diastasis with talus in between them3,4.

Management options include syme's amputation or limb salvage with modified custom made prosthesis. Parents should be counselled regarding the outcome of each option. In our case, the child had a deformed split foot, so syme's amputation was performed5.

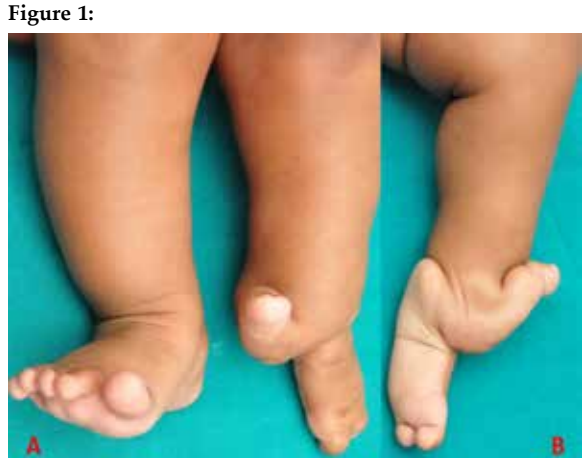
Conclusion:
Tibial hemimelia with ectrodactyly is a known association reported in literature. However a varied deformation of split foot has not been described earlier. This type of tibial hemimelia with deformed split foot and its management is being reported first time in the literature.

Consent: taken

Acknowledgements: None

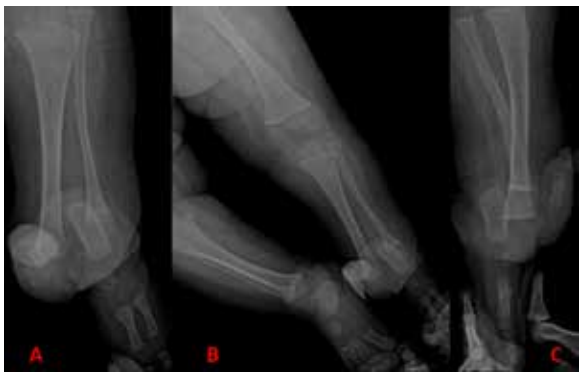
Conflict of interest: None

Figure 1:



Legend: Clinical photograph showing the left tibial hemimelia with split foot anterior-posterior[A] and laterl view[B]

Figure 2:



Legend: Radiograph of the left leg with foot anterior-posterior[A] and lateral view[C] showing Jones type 4 Tibial hemimelia with distal tibio-fibular diastasis with the talus in between them and split rays of the foot.

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