Scholars Journal of Applied Medical Sciences
Abbreviated Key Title: Sch J App Med Sci
ISSN 2347-954X (Print) | ISSN 2320-6691 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Pediatric

Case Report: Tibial Hemimelia

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DOI: 10.36347/sjams.2023.v11i06.006 | **Received:** 10.04.2023 | **Accepted:** 22.05.2023 | **Published:** 07.06.2023

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Abstract **Case Report**

Unilateral tibial hemimelia is an anomaly characterized by the absence of the tibia. Tibial hemimelia is the rarest form of congenital longitudinal deficiency of the tibia with an incidence of 1 in 1,000,000 live births. It is characterized by deficiency of tibia with relatively intact fibula. It causes marked shortening of the limb with a severe equinovarus deformity. It may occur in isolated form or combined with other congenital anomalies. Some cases of tibia hemimelia are genetically transmitted as autosomal dominant or recessive, whereas others are sporadic. The treatment is always difficult and challenging. Amputation was the preferred treatment option specially in complete tibial absence; however, a conservative management is used in other types of the deformity.

Keywords: Tibial hemimelia, tibial deficiency, Jones classification.

I. INTRODUCTION

Tibial hemimelia is the rarest of the lower extremity deficiencies. It results from a disruption of the lower limb developmental field during embryogenesis due to slow or even abort of chondrification process, which results in leg length discrepancy. Affected leg appears short and deformed with knee and ankle involvement, and the foot will be in an abnormal position. Jones classification is simple widely used radiographic classification [1], and farther modified by Kalamchi and Dawe in 1985 [2]. Recent classification by Paley in 2003 is also based on Jones classification [3]. These classifications help to determine an appropriate intervention in terms of limb salvage surgery or amputation. We present a case of a 4-weeks-old boy with type II tibial hemimelia according to Jones classification.

II. CASE PRESENTATION

We report the case of a 4-weeks-old boy with a deformed right leg, second out of 2 children of non-consanguineous parents, with no relevant family history. He was born by spontaneous vaginal delivery at 39 weeks of an uncomplicated pregnancy, average birth weight and height. On examination the baby was not dysmorphic, the hip was stable, and the right lower limb was short with varus deformity (Fig. 1) with no other associated malformations. Quadriceps function was good. Psychomotor development was normal. Radiograph of the leg revealed presence of proximal tibia with absence of distal tibia, and the fibula is intact (Fig. 2). The surgical procedure that our surgeons proposed to the parents consisted of soft tissue distraction between the proximal tibial residual remnant and fibula, and distal fibula and foot, followed by centralization of fibula under the proximal tibial remnant, and foot under the distal fibula.

Fig. 1: Clinical photograph showing severe shortening associated with varus deformity

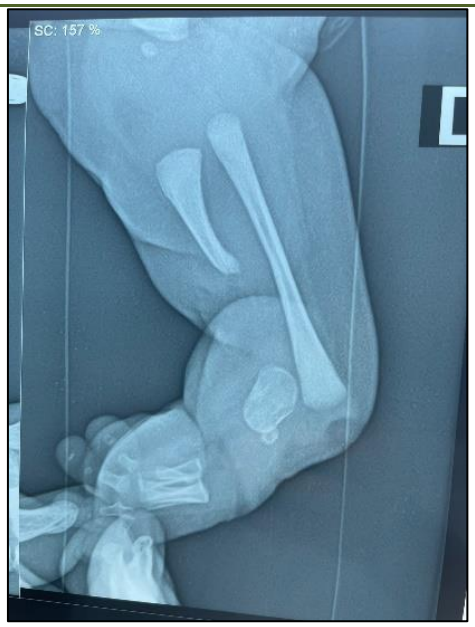

Fig. 2: Radiograph showing intact fibula with presence of a proximal tibia and an absent distal tibial segment

III. DISCUSSION

Several classifications for tibial hemimelia have been proposed. Jones in 1978 firstly published his classification based on X-ray founding [1]. Thirty years later, a new classification was introduced by Weber taking into account X-ray and cartilaginous anlage [4]. According to Jones classification, our case corresponds to type II.

The Jones type II tibial hemimelia has good recovery potential for the reason that the proximal portion of the tibia is ossified and visible on X-rays at birth and there is good knee function. Management of this type has many options, depending on the radiological type and quadriceps function. It was treated in the past by tibiofibular synostosis followed by a Syme or Boyd amputation [1, 2, 5]. However, recent practice has shifted toward limb preservation using tibiofibular synostosis and centralization of foot followed by length restoration using callus distraction [6-8]. The former operation is usually carried out around 10–12 months of age and the latter performed around 3–7 years of age [5, 9].

IV. CONCLUSION

Tibial hemimelia is a rare congenital anomaly. Knowledge of the different aspects of this pathology is essential for an adequate global management.

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