Seid Mohammed, Richard O.E. *Ethiop Med J, 2020, Vol. 58, No. 4*

CASE REPORT

GOLLOP-WOLFGANG COMPLEX IN AN 18 YEARS OLD FEMALE

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ABSTRACT

Skeletal dysplasias are disorders associated with a generalized abnormality in the skeleton. The Gollop-Wolfgang complex is a limb deficiency disorder and an unusual limb malformation with highly variable manifestations. Here, I report a rare case of an 18-year old female patient with Gollop-Wolfgang Complex showing bifurcation of the left femur, bilateral hemimelia and ectrodactyly of the ipsilateral foot. The etiology of Gollop-Wolfgang complex in this patient could be a familial genetic condition, since she had a younger brother with tibial hemimelia and bilateral cleft hands. The clinical and radiographic findings are presented in detail.

INTRODUCTION

Skeletal displasias represent generalized disorders of cartilage and bone (1). Skeletal dysplasia, affecting around 4 million people worldwide is a heterogeneous group of more than 200 disorders, characterized by abnormalities of cartilage and bone growth - resulting in abnormal shape and size of the skeleton and disproportion of the bones. A cumulative international incidence of at least 1:5000 newborns has been estimated (2).

Gollop-Wolfgang Complex (GWC) is a rare congenital limb anomaly characterized by tibial aplasia, ipsilateral bifurcation of the thighbone and ectrodactyly (3). Very often, the anomalies of limbs, heart, digestive and urinary tracts and the lumbosacral vertebrae are also affected (4).

Ectrodactyly involves the deficiency or absence of one or more central digits of the hand or foot and is also known as split hand/split foot malformation (SHFM) (5). The term ectrodactyly has been applied to a variety of malformations of the fingers or toes. But it is probably best reserved for transverse terminal aphalangia (absence of the last bone in the finger or toe, adactylia (total absence of a finger or toes), or acheiria (total absence of one or both hands) (6).

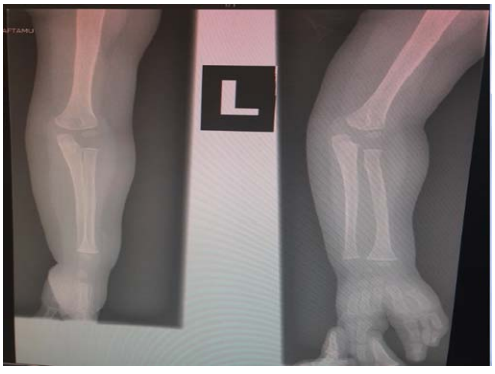
In 1980, Gollop, *et al.* described a case of two brothers with ectrodactyly and unilateral bifurcation of the femur, absence of both tibiae and monodactyly of the feet. In 1984, Wolfgang reported a case of right femoral bifurcation and absence of tibia and bilateral central defects of the hand. Endo, *et al.* found a total of 12 reported cases and added the case of a Japanese girl with a unique form of this malformation complex.

CASE PRESENTATION

An 18 years old female, 9th grade student from Ethiopia, born to a 46-year-old lady, at full term by SVD, presented with limb deformities which included two prominences at her left knee and a short, deformed ipsilateral leg. She also has deformities in the contra lateral leg for which she can't walk on her legs but uses her hands to lift her body and move around. She performs well at school and has better than average scores.

There was no history of exposure to radiation, prenatal teratogenic medications or infections during pregnancy. The mother did not smoke or drink during pregnancy. The child was breast-fed with good appetite for one year.

She has a younger brother with bilateral cleft hands and right leg tibial hemimelia. No other family members are affected. Figures 1,2 and 3 are radiographic images of her affected brother's both upper



extremities and right leg.

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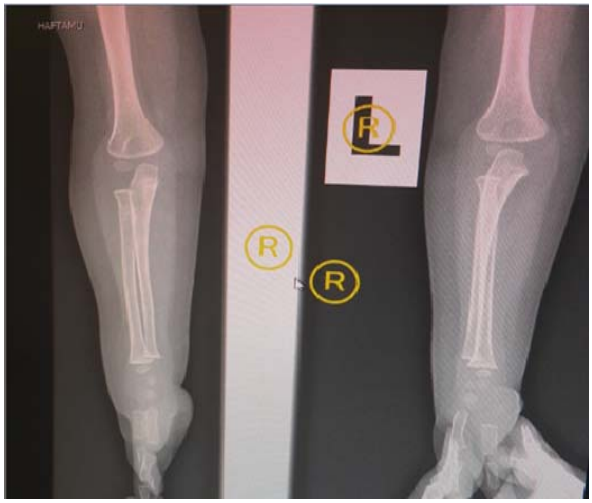


Figure 2: Cleft hand of the right upper extremity (younger brother).

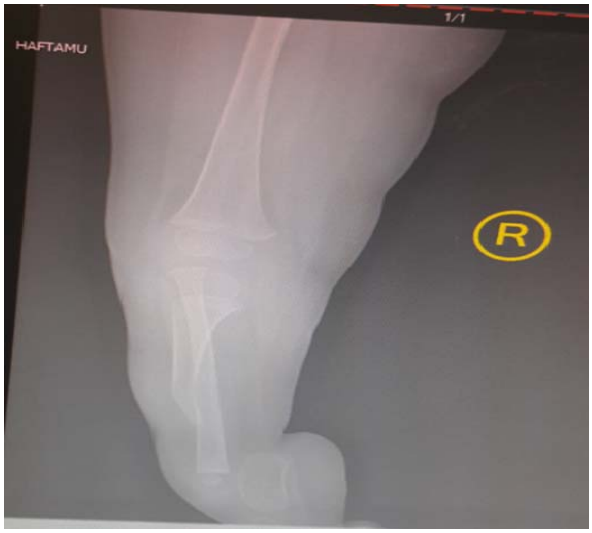


Figure 3: Tibial hemimelia of the right lower extremity (brother).

Coming back to our patient, her physical examination revealed deformed **left** thigh, knee, leg and foot (Figures 4 and 5):

- Deformed left femur with two prominences at the knee
- Short, inverted and hypoplastic foot with absent two central digits
- Patella couldn't be localized
- Fixed flexion deformity of the knee at full flexion (both active and passive)
- Absent quadriceps function
- Distally, she has good capillary filling and sensations are intact

Right foot is flat, internally rotated, and is with rigid equinovarus deformities (Figures 4 and 6).

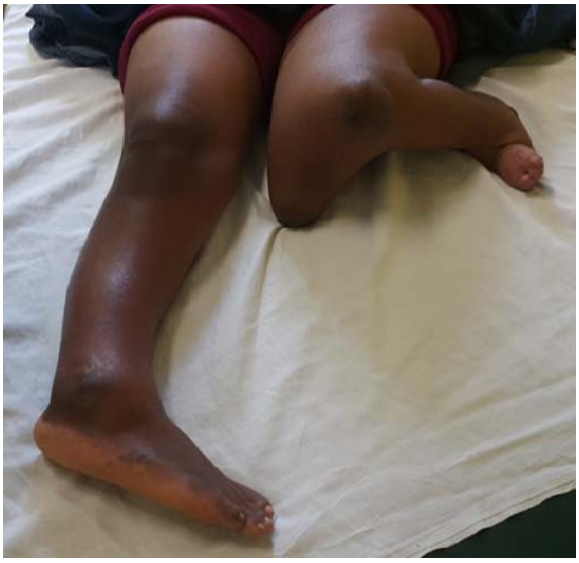
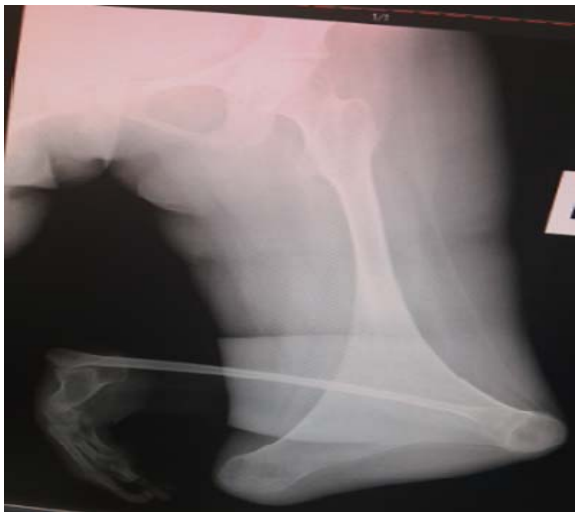


Figure 4: A picture of the lower extremities of our patient.

She has normal upper extremities and normal trunk and pelvis. No cardiac or spinal anomalies was identified. Radiographic images showed bifid **left** femur with ipsilateral tibial agenesis (Figure 5); intact fibula; absent left patella and absence of left foot's 1st and 2nd central digits along with their corresponding metatarsals; there is fusion of 3rd and 4th metatarso phalangeal joints and a single phalanx emerging out of this fusion. Radiographic images of the **right** lower extremity showed distal tibio fibular



diastasis.

Figure 5: Radiographic image of the left lower extremity of our patient.

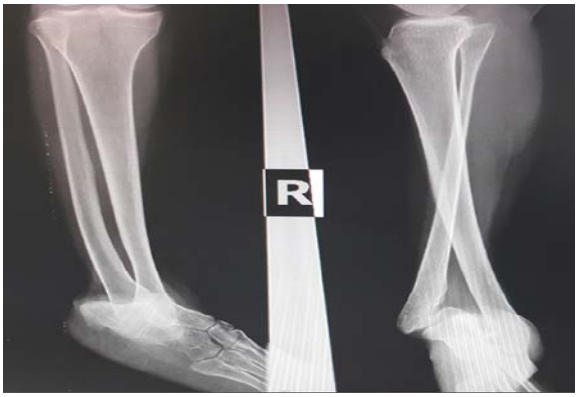


Figure 6: Radiographic image of the right leg and foot of our patient.

DISCUSSION

Our patient has all components of Gollop Wolfgang Complex, which includes tibial aplasia, ipsilateral bifurcation of the thighbone and ectrodactyly.

The etiology of GWC is most likely an error in the complex genetic control of limb development but the exact cause is still unclear. GWC is listed as a "rare disease" by the United States Office of Rare Diseases (ORD) of the National Institute of Health (NIH) (8,9). The fact that our patient has a younger brother with features of skeletal dysplasia signifies familial nature of her condition denoting a possible genetic component as an etiology.

The best treatment option for patients with Gollop-Wolfgang syndrome is early knee disarticulation and resection of the protruded bifurcated femur, followed by fitting of a modern prosthesis. For patients with partial tibial hemimelia, foot centralization, with tibiofibular fusion can be done. But for those with complete tibial hemimelia, even though the required post operative rehabilitation is quite intensive and yet the outcome is uncertain, the possible reconstructive option is fibular transfer (Brown's procedure) and callus distraction lengthening. (5,10,11).

In summary, our patient has: Bifurcated left distal femur + absent left patella + fixed flexion deformity of the left knee + ipsilateral Jones' Ia tibial hemimelia + ipsilatral absent central digits of the foot + contralateral Jones' type 4 tibial hemimelia + deformed right ankle and flat foot + bilateral rigid equino varus deformity of the feet. She has a family member (younger brother) with right side tibial hemimelia and bilateral cleft hands. No other family members are affected. There were no additional associated abnormalities like cleft lip/palate, tibial agenesis, visceral or cardiac anomalies seen in this patient.

Conclusion

This case with the above combination of typical GWC (Gollop Wolfgang Complex) and additional features of bilateralism of leg deformities and the presence of a limb anomaly in a close family member calls for further researches in this rare anomaly especially with regard to its possible familial nature.

Competing Interest

The authors declare that this manuscript was approved by all authors in its current form and that no competing interest exists.

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