CASE REPORT

Gollop-Wolfgang Complex in a New Born with Morton's Toe and Congenital Heart Disease

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Abstract:

Gollop-Wolfgang complex is a rare anomaly comprising of bifid femur, tibial aplasia or hypoplasia and cleft of hands with variations and other systems involvement. A case of Gollop-Wolfgang complex is reported here, which was associated with other rare anomalies such as bilateral Morton's toe, ventricular septal defect and congenital talepus equinovarus.

Keywords: Gollop-Wolfgang complex, Bifid femur, Tibial aplasia, Ectrodactyly

Introduction:

Gollop-Wolfgang Complex (GWC) is an association of congenital limb malformation characterized by bifid femur, absent or hypoplasia of tibia and ulna and ectrodactyly. There are around 200 cases of GWC reported as rare anomaly by United States Office of Rare Disease (ORD) worldwide [1]. It is occasionally associated with Morton's toe, congenital heart defects, cleft lip, cleft palate and tracheo-esophageal fistula.

Case Report:

A full term male newborn presented at the of age 36 hours, born via unassisted normal vaginal delivery was admitted to neonatal intensive care unit in a tertiary care hospital with visible deformities on second day of life. He was born to a non-consanguineous parents without any family history of diseases associated with inborn errors of metabolism, chromosomal abnormalities or congenital malformations.

Antenatal period was uncomplicated. Mother was a 24 year old second para with no history miscarriages or any exposure to radiation. Laboratory investigations of mother for toxoplasma rubella, cytomegalovirus, herpes simplex, syphilis and human immunodeficiency virus infections were normal during antenatal period. Antenatal ultrasonography was not performed. On clinical examination, occipitofrontal circumference of newborn was 34.5 cm (95^{th}centile), weight 2.395kg (25^{th}centile) and length 49.5cm (95^{th} centile) according to Indian Academy of Paediatrics Chart. His heart rate was 146/min, respiratory rate 48/min and oxygen saturation (SpO_2) was 98% at room air.

His rectal temperature was 36.8^0C. There was a swelling in the distal one third of right thigh and X-ray of right lower limb showed a bifid femur and tibial aplasia (Fig. 1). There was bilateral congenital talipes equinovarus. Left hand showed ectrodactyly, also known as lobster claw deformity which was confirmed clinically and radiologically (Fig. 2). Both feet had Morton's Toe also called Greek Toe / Royal Toe4 and Congenital Talipes Equinovarus (CTEV) (Fig. 3). On auscultation of the precordium, a loud pansystolic murmur was heard almost all over the precordium. 2-D Echocardiography showed a Ventricular Septal Defect (VSD) and mild right ventricular hypertrophy.

Examination of respiratory system, Central Nervous Sytem (CNS) and Gastrointestinal Tract (GIT) systems were normal. Although there was a swelling in right thigh, the movement over right knee joint was normal.

Ultrasonography of abdomen was normal. Venous blood was obtained for laboratory tests. Complete blood counts, blood sugar, serum electrolytes, Urea and Creatinine, Alanine Aminotransferase (ALT), Aspartate Aminotransferase (AST) serum alkaline phosphatase, serum albumin, serum globulin and serum calcium was normal. Karyotyping was not performed, due to economic constraints. Patient was referred to the orthopaedic surgeon and radiologist findings were confirmed. As according to the orthopaedic surgeon the bony deformities were complex, patient was advised to consult a paediatric orthopaedic surgeon.

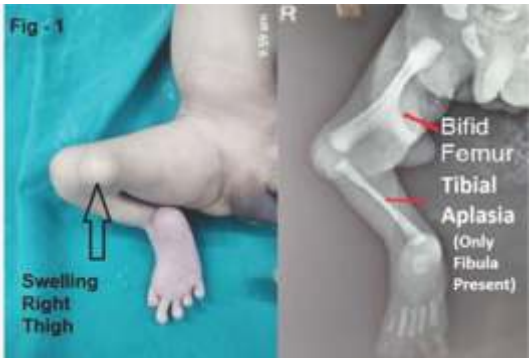

Fig. 1: Showing Bifid Femur and Tibial Aplasia

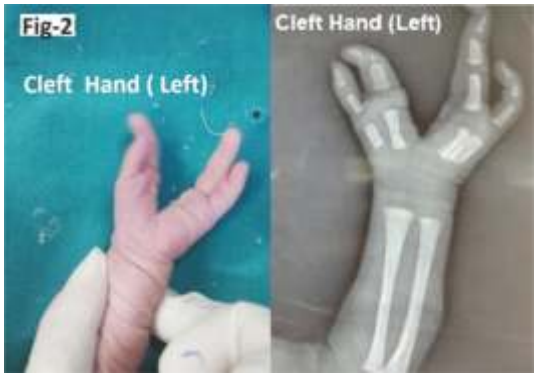

Fig. 2: Showing Ectrodactyly

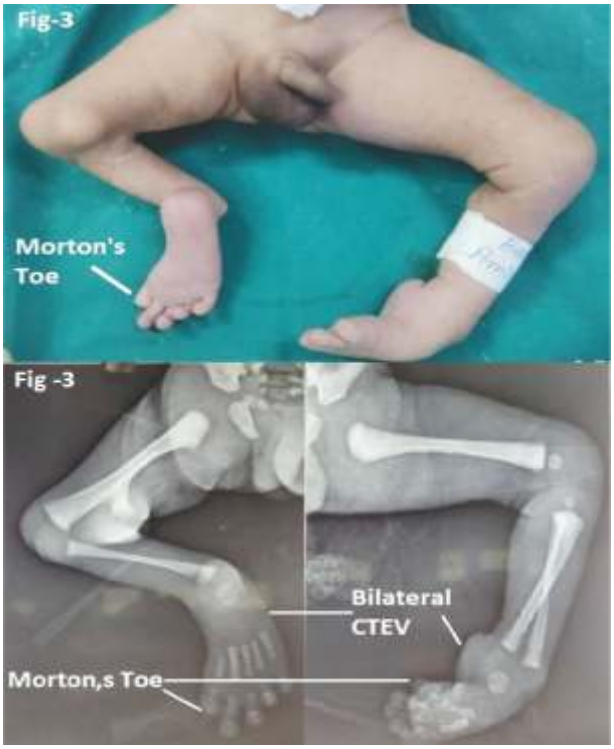

Fig. 3: Showing Morton's Toe and CTEV

Discussion:

In 1984, Wolfgang reported complex congenital anomalies of the lower extremities: femoral bifurcation, tibial hemimelia and diastasis of the ankle [2]. In 1986, the term 'Gollop-Wolfgang complex' was introduced by Lurie and Ilyina as they concluded that the association of hand ectrodactyly and femoral bifurcation was not coincidental [3]. Our patient had all the major features of GWC which are, bifid femur, tibial aplasia and ectrodactyly. Brachymetatarsia is a condition in which the metatarsal epiphysis closes prematurely yielding a pathologically shortened metatarsal length. Brachymetatarsia of the first metatarsal leading to shorter first toe compared to 2nd toe is also known as Morton's toe [4].

Morton's toe was present in both lower limbs in our patient. Though diagnosis of GWC by antenatal

ultrasonography was difficult, New American Institute of Ultrasound Medicine guidelines has improved the possibility of detection of GWC during the antenatal period [5]. GWC is inherited autosomal dominant or autosomal recessive [6]. Antiepileptic agent has been considered as one of the etiology of GWC most notably sodium valproate has been documented [7] and also in3 siblings of a mother taking carbamazepine during antenatal period [8]. Our patient was most likely to be sporadic in inheritance. Literature search for association of GWC and congenital heart disease yielded few case reports with citations available that too stating broadly as congenital heart disease, the type of which were not specified. Only one case report of GWC with ventricular septal defect was described by Celentano *et al.* [9].

Conclusion:

Gollop-Wolfgang Complex is a rarely reported defect with multiple bony and other variable manifestations involving multiple systems. Its exact etiology is not known but various factors are postulated such as genetic defect and anticonvulsant drugs. Improved methods of ultrasonography in antenatal period has increased chances of early detection of defect.

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