GOLLOP WOLFGANG COMPLEX - A CASE REPORT
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Abstract: Around 205 limb abnormalities have been described. Congenital absence of tibia is a rare and severe lower limb malformation with an incidence of approximately 11,000,000 live births. Here we present an infant who had multiple limb deformities bifurcated femur, absent tibia, ectrodactyly of upper lower limbs. These features typically fitted into a complex called GOLLOP WOLFGANG COMPLEX (GWC).

Keyword: gollop wolfgang complex, Ectrodactyly, bifurcated femur, tibial aplasia.

INTRODUCTION: Congenital limb deficiencies have an incidence of 1:1,000,000. Only a few cases of GWC have been described in the literature. It was first described in 1575 by Ambroise Pare, the complex was named after Gollop & Wolfgang who described the same in 1980 & 1984 respectively. It is classified under syndromic Split Hand Leg Malformation. GWC is differentiated from its close variant TIBIAL APLASIA – ECTRODACTYL syndrome by presence of bifurcation of femur more commonly in the former. It is inherited as autosomal dominant with variable or reduced penetrance.

CASE SUMMERY: 38 days old male infant third born of non-consanguineous parents presented with complaints of multiple limb deformities. Antenatal history was not significant for drug intake / radiation exposure. Antenatal sonogram was normal. Child was delivered by full term normal delivery & had an uneventful natal period.

middle finger absent, rudimentary thumb
On examination cry & activity of the child was good. vitals were stable. He had retrognathia, ectrodactyly of all 4 limbs and protrusion of distal part of thigh on left side. Other systems were normal.

Ectrodactyly

LEFT UPPER LIMB: showing all metacarpels present & phalanges of middle finger absent. X-RAY BOTH LOWER LIMBS:
Shows bifurcated femur & absent tibia.

ECHOCARDIOGRAM & USG ABDOMEN: normal study middle finger phalynx absent. left sided bifurcated femur & absent tibia.
DISCUSSION:
Our case had the classical clinical and radiological features of GWC. Ectrodactyly is defined as Cong. absence of one or more fingers or toes. The diagnosis of GWC is mainly clinical and confirmed by x-rays of upper & lower limbs. Its clinical features are aplasia of middle fingers (74%) with absence of only MCP or phalanges, deficiency of ulna (12%), tibial aplasia or hypoplasia (59%) with preaxial oligodactyly or monodactyly of foot which is held in equino varus and bifurcation of the distal femur (8%) primarily unilateral – classic form of GWC. They will also have normal intelligence and short stature. Karyotyping may not be useful in confirmation. Carriers are diagnosed in AD forms. Antenatal diagnosis is possible – often missed due to the position of the fetus in utero. Anomaly scan should be done at 20 to 22 weeks for diagnosis.

TREATMENT:
• Genetic counselling.
• Surgical & prosthetic treatment.
• If tibial anlage present – tibio fibular synostosis provided there is a good quadriceps function.
• If tibial anlage absent – knee disarticulation.

REFERENCES:
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