Male Neonate with Polymelia and Left Renal Agenesis: Syndromic Association?

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Abstracts: In the embryo, the limb arises as a condensation of cells from the lateral plate mesoderm and its Ectodermal covering. Congenital malformations involving limb sometimes lead to prenatal mortality, post natal morbidity and psychological effect on parents. Polymelia is one of such condition where diagnostic decisiveness rest on shoulder of radiologist and has an implication on management aspect as far as surgery is concern. We report one unique case having polymelia with left renal agenesis. [Solanki V NJIRM 2015; 6(1):111-113]

Key Words: polymelia, radiography, renal agenesis, accessory limb

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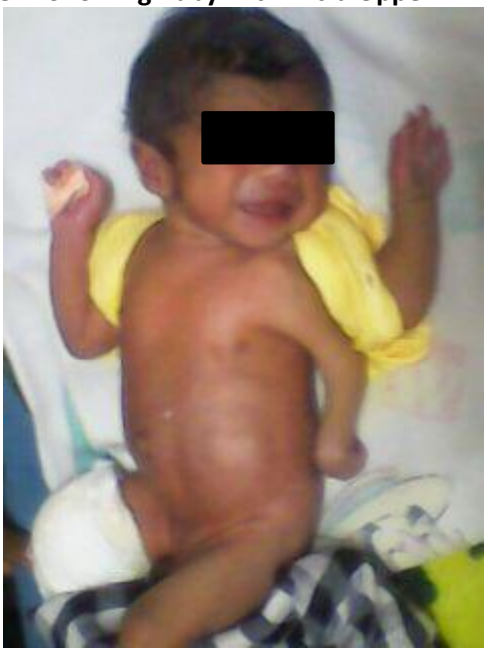
Introduction: Polymelia is a congenital anomaly, which is defined as the presence of accessory limbs attached to various body regions and could be classified as cephalomelia (extra-limb attached to the head), notomelia (extra-limb attached to the back bone), thoracomelia (extra-limb attached to the thorax), and pyromelia (extra-limb attached to the pelvis). These anomalies are usually associated with genetic factors including teratogens, chromosomes, and environmental agents. Here, we report a rare case of thoracomelia, a form of polymelia.

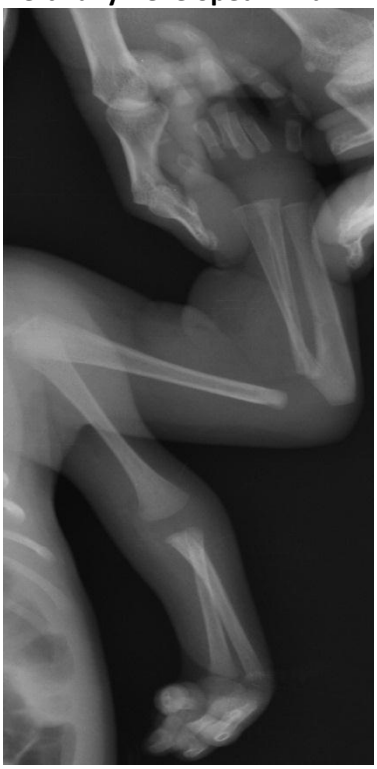
Case Report: A 02 day old male presented to us with an extra upper limb (Figure 1).

Figure 1: Showing Baby with Extra Upper Limb



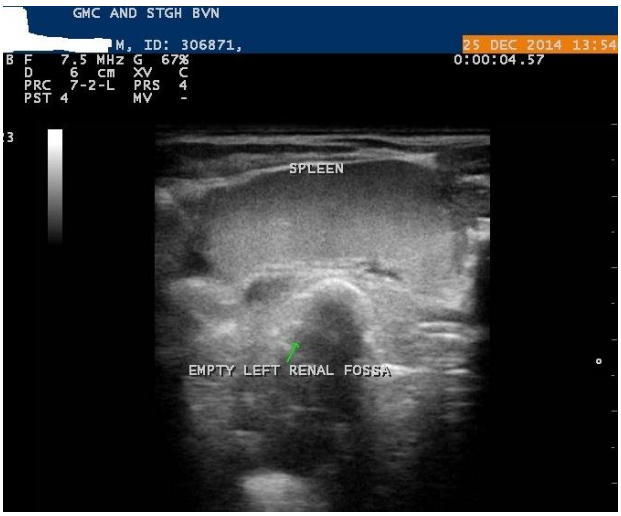
Plain X-ray demonstrated a normal right shoulder joint left shoulder joint with an accessory left fully developed upper limb. The accessory limb had humerus falsely attached to normal scapula of the left shoulder joint. Clinically left shoulder joint movements were normal. This joint had normal flexion and extension movement. Distal most end of the accessory limb had two radiuses, large ulna with six fully developed fingers with no metacarpal ossification (Figure 2).

Figure 2: Radiograph (Baby Gram) Of The Patient Reveals A Normal Left Shoulder Joint With An Accessory Left Fully Developed Limb.



Ultrasonography of abdomen shows absent left kidney with undescended left testis(Figure 3).. There was no history of congenital anomalies in the family members. There was no history of teratogenic drug intake during pregnancy by the mother. The patient has undergone limb resection of accessory limb.

Figure 3: Ultra-sonography Showing Empty Left Renal Fossa



Discussion: Isolated limb duplication is a rare congenital condition and only a few cases have been documented. Various adverse embryo genic influences are responsible for this kind of anomaly. [1, 2, 3]

Limb differentiation occurs roughly between the 4th and 5th weeks of embryonic development,[4] and it follows a dorsal to ventral and proximal to distal pattern, with many factors involved in the process.[2,3]

Initially, two pairs of limb buds – anterior and posterior – protrude from both sides of the embryo, and comprise cells of ectoderm and mesoderm. Their interaction is responsible for cell positioning and limb differentiation. The outer ectodermal layer of the limb bud is termed the apical ectodermal ridge (AER). The zone of proliferating activity (ZPA), another group of cells is located beneath the AER. Both are necessary for limb development. Mesodermal cells in the ZPA stimulate AER formation and the AER maintain the ZPA.[5,6]

As the AER grows further, the induced mesoderm cells, comprising rudimentary parts of the limb, can continue to grow without any developmental interference even if the AER is transplanted to the adjacent region. This leads to an assumption that duplication of the limb arises from the influence of the AER with abnormal splitting creating two sets of limbs.[7]it is possible that one or more of the growth factors produced by the mesonephros take some cells of the intermediary mesoderm out of their renal way to form a supernumerary limb. As our case had left renal agenesis.

Other deformities in our patient such as semi vertebrae, spina bifida, scoliosis, and hip dislocation were absent. We found only a right club foot.

Several factors lead to this anomaly. Intrauterine exposure of thalidomide like teratogenic drug or progesterone like hormonal drug may leads to development of child with limb defect. These defects ranged from absence of the limb (Amelia) or proximal limb elements (phocomelia) to loss of the thumb or polydactyly, polymelia, and other congenital defects. Antenatal anomaly screening of foetus with ultrasonography can be a useful tool to diagnose such conditions *in utero*.

Treatment for such a case like ours requires surgery to detach the soft tissue between false limb and the true shoulder joint.

Conclusion: Radiologist plays a significant role in patients with polymelia antenatal detection, to assess for additional congenital anomalies and neurovascular distribution in accessory limb before surgical intervention.

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