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Potters syndrome, A Case Report

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Abstract: *During the dissection of foetuses(obtained from gynecology and obstetric dept of KGH), we observed a foetus showing polycystic horse shoe kidney (cysts of size varing from 1mm --- 5mm) fusion seen in lower pole. The right kidney is smaller and pertained to pelvic region, (suprarenal gland not seen)The left kidney extended up to diaphragm and left suprarenal gland is disc like due to compression. Ureters seen to arise from hilum of both kidneys but are fused in pelvic region and opened into the urinary bladder . The anal agenesis (anal atresia) is observed and anal opening absent (imperforate anus)The rectum opened into the urinary bladder.*

Keywords: *Polycystic horse shoe kidney, Anal atresia*

I. Introduction

Potters syndrome is a rare congenital anamoly seen more predominently in males (2:1). Diagonised usualy at birth and by foetal autopsies in still born , (0.2%-0.4%) . It was first reported by Edith Potter (1946) Foetuses showed renal agenesis or polycystic kidneys (90%) with Lung hypoplasia due to oligohydraminios.40% of cases are fatal (foetuses are still born or die within few minutes after birth) due to severe respiratory insufficiency. Renal failure is the principle defect leading to sequence of events.Premature birth, IUGR, typical compressed faces (Potters faces) are commonly observed findings.

II. Case Report

During the dissection of foetuses(obtained from gynecology and obstetric dept of KGH), we observed a foetus showing polycystic horse shoe kidney (cysts of size varing from 1mm --- 5mm) fusion seen in lower pole. The right kidney is smaller and pertained to pelvic region, (suprarenal gland not seen)The left kidney extended up to diaphragm and left suprarenal gland is disc like due to compression.(Figure .1) Ureters seen to arise from hilum of both kidneys but are fused in pelvic region and opened into the urinary bladder.(Figure1) . The anal agenesis (anal atresia) is observed and anal opening absent (imperforate anus).(Fig; 2)The rectum opened into the urinary bladder.(Fig. 2) Scrotum and penis showed no canalization and are cord like.(Fig 2) Urinary meatus absent.(Fig 2). Ectopia Testes are seen (Inguinal region on both sides and with vas deferens opening into urinary bladder. (Fig.3) Urachus (vitellien duct) seen to be persistent and opened into the umblicus.(Fig.3) Face is seen to be flattened, ear are low set , compressed against head and are posteriorly placed. (Fig.4) Foetus usually is small for date.(Low birth weight) APGAR score:> o7 maternal age > 20 yrs , 15 % normal show normal vaginal deliveries. Bilateral dysplastic kidney or bilateral polycystic kidneys seen in 22 – 25 % of cases. Hypoplastic lungs are seen in 8o% of cases.

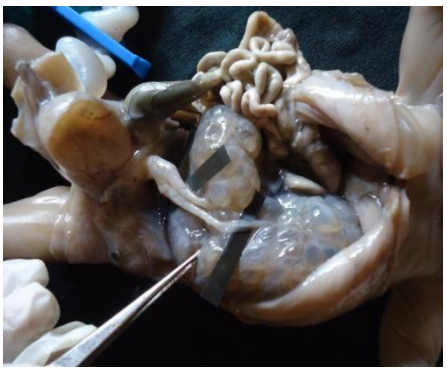

Fig. 1 Showing fused Rt& Lt lower poles with Ureters opening into bladder with common opening. (Horse shoe)

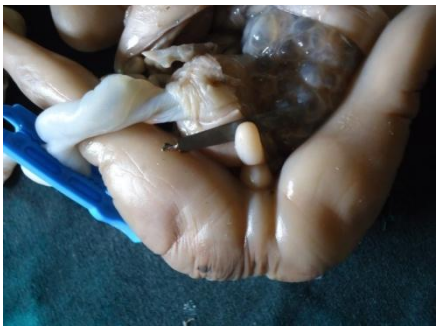

Fig 2 Showing anal atresia and anal opening absent. Scrotum and penis non canalised.

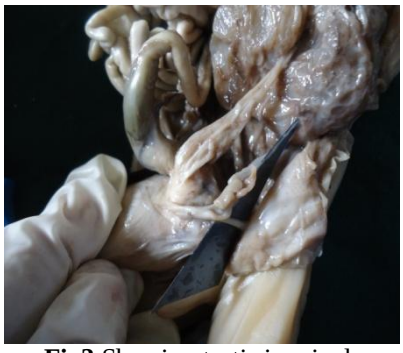

Fig3 Showing testis inguinal.

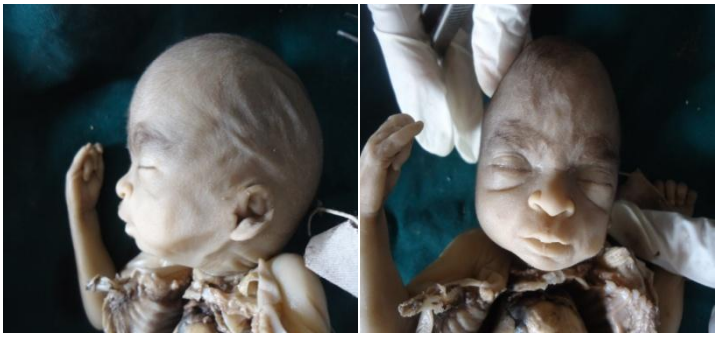

Fig4 Defect in face.

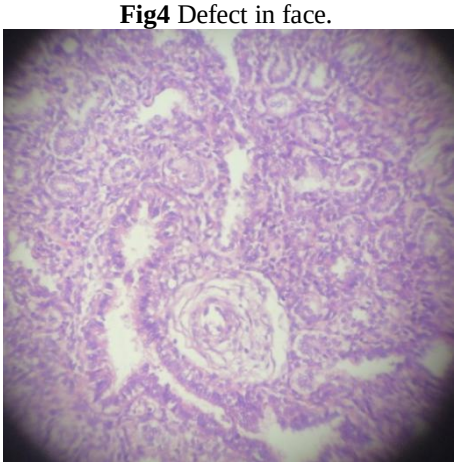

Fig 5: Histology of foetal lung

III. Discussion

➢ There is a developmental link between the lungs and kidneys. Urine from foetal kidneys maintains the volume of amniotic fluid which is required for normal foetal lung development. Renal dysplasias and agenesis are a spectrum of severe malfromations that show numerous ducts which are surrounded by undifferenciated cells. Nephrons fail to develop, Ureteric bud fail to branch so that the collecting ducts never form. It also occurs due to interaction between metanephric mesoderm and uretric bud. Mutation in genes that regulate GDNF

(responsible for branching and growth of ureteric bud, Bilateral renal agenesis (1 in 1000 births) results in renal failure and baby presents with Potters syndrome, anuria, oligohydraminos, hypoplastic lungs. 80% casesshow urogenetal anomalies. Oligohydraminos compression of foetus in uterine cavity causes abnormal facies. Congenital polycystic kidneys is both an autosomal recessive(in children).occurs in 1 in 5000 births and autosomal dominant(adults).ARPK ….is a progressing disorder,kidneys are large renal failure seen in infancy. Fusion of ureters results due to failure of splitting of ureteric bud. Horse shoe kidney results due to arrested acent of the kidneys by IMA &IMV & susequent approximation of metanephric tissue (usually at lower pole forms isthmus). Developmental defects in the mesonephric and the paramesonephric ducts may have a common genetic. Basis and Schimke and King used the term 'hereditary urogenital. Adysplasia for the combination of anomalies of the mullerian duct. With developmental errors of the urinary tract. Non working or mutated genes on the long arm of chromosome 10(10q) result in the abnormal development of the urogenital tract patients with the monosomy of chromosome 10q26, six patients had urinary anomalies such as vesicoureteral reflux and hypoplastic kidney, and 8 had genital anomalies such as micropenis, hypospadias, cryptorchidism, and hypoplastic labia majora. of urogenital development in mice who lacked the Emx2 genes on the distal 10q chromosome. Skinner et al. [16] identified 10 different heterozygous mutations in the RET gene. In vitro functional expression studies showed that the mutations resulted in either constitutive RET phosphorylation or absent phosphorylation. Yang et al. [17] observed a significant association between the primary vesicoureteral reflux and a G691S polymorphism in the RET gene among French Canadian patients with Potter's syndrome. The stillborn foetus which was under study was a destitute (unclaimed child, without parents), which was brought to the department and as such, his prenatal history could not be probed. As the foetus had cystic kidneys with urogenital defects, a possible genetic condition was thought of and it could not be further probed as the parents could not be traced.

IV. Conclusion

Fetal polycystic horseshoe kidney with associated hind gut developmental variations is a very rare presentation the infants usually are still born or die immediately after birth due to respiratory insufficiency (cardio pulmonary resuscitation is mandatory handling such cases in the labour room). Ultra sonograph during the 1st trimester of pregnancy helps to identify these cases early which enables counseling the parents for taking wise decision regarding the termination of pregnancy and reducing the physical and psychological trauma of to the parents Ectopia testes….testes leaves the abdomen but does not reach the scrotum.(defect in distal attachments of gubernaculum). Imperforate anus …failure rupture of anal membrane ,non development of ectodermal proctoduem, atresia of lower segment of rectum. Common associated defects are: cardiac, tracheal &duodenal atresias, cleft lip & palate, brain anomalies, GIT anomalies.

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