

Case Report

A RADIOLOGICAL STUDY OF POTTER'S SYNDROME BABY IN UTERO

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ABSTRACT

The objective of present study is to report a case of fetus showing Polycystic Kidney with oligohydramnios by ultrasound study. This case was studied in the Department of Radiology of Velammal Medical College Hospital & Research Institute, Madurai. This study was done in the month of May 2013. A mother of 26 years old with 34 weeks amenorrhoea underwent ultra sound examination dated 28.05.2013 at our Velammal Medical College Teaching Hospital which showed 38 weeks of amenorrhoea with enlarged bilateral autosomal recessive polycystic kidneys with severe oligohydramnios. Ultra sound also showed placenta in fundal and anterior wall region of the uterus. Features suggestive of Autosomal bilateral enlarged recessive polycystic kidney with oligohydramnios were present. There is no evidence of pelvicalyceal dilatation. Medullary pyramids shape is maintained. Has shown a single live fetus with enlarged polycystic kidneys associated with gross oligohydramnios. There is no history of low set ears and involvement of lungs, no widely placed epicanthic folds. This is study of bilateral enlarged recessive polycystic kidney with oligohydramnios is suggestive of Potter's Syndrome.

Key Words: Oligohydramnios, Bilateral Polycystic Kidney, Low Set of Ears, Cystic diseases, Hypoplastic Lungs, Premature Rupture of Membranes

INTRODUCTION

Potter's Syndrome is syndrome having both renal and non renal features. Non renal features are i) Alterations in the development of face, associated with ii) Late deficiency of fetal growth, iii) aberrant hand and foot positioning and iv) hypoplasia of lungs.

These are secondary manifestations to prolonged compression of fetus due to oligohydramnios. Oligohydramnios is a result of either renal agenesis or defective development of urinary system causing deficiency of flow of urine into amniotic space or in production of urine (Ioan *et al.*, 1965).

There will be also hypoplasia of abdominal muscles due to compression of large kidneys on developing anterior abdominal musculature.

This condition is seen prune-belly syndrome along with nonrenal features of Potter's syndrome. The pathogenesis lies in teratogenic role seen in cytomegalovirus inclusion disease and other viruses, so there will be coexistence of both Potter syndrome and prune-belly syndromes (Arun *et al.*, 1977).

Three fatal cases of prune-belly syndrome were associated with nonrenal features of Potter syndrome which has been reported by Arun *et al.*, (1977).

MATERIALS AND METHODS

A mother of 26 years old with 34 weeks amenorrhoea underwent ultra sound examination dated 28.05.2013 at our Velammal Medical College Teaching Hospital in the Department of Radiology which

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showed 38 weeks of amenorrhoea with enlarged bilateral autosomal recessive polycystic kidneys with severe oligohydramnios. There was no hypoplasia of lungs.

RESULTS AND DISCUSSION

Results

Ultrasound report done on female aged 26 years old on 28.05.2013 at our Velammal Medical College Hospital Showed Single live foetus in cephalic presentation with the foetal spine to the maternal left. Foetal movements were present along with foetal cardiac pulsations and foetal heart rate of 134bpm. Ultrasound also showed placenta in fundal and anterior wall region of the uterus. Features were suggestive of Autosomal Recessive Bilateral Enlarged polycystic kidney with oligohydramnios. Liquor is not seen in four pockets – Severe oligohydramnios. Bilateral fetal kidneys are echogenic and grossly enlarged in size. Right kidney measures 9.0 x 4.3 cm and left kidney measures 8.6 x 4.1cm. There is no evidence of pelvicalyceal dilatation. Medullary pyramids shape is maintained.

Foetal Biometry is given in the following table:

PARAMETER	MEASUREMENTS	GESTATIONAL AGE	
Biparietal Diameter	84 mm	33 Wks	6 Days
Head Circumference	304 mm	33 Wks	6 Days
Abdomen Circumference	375 mm	41 Wks	3 Days
Femoral Length	65 mm	33 Wks	6 Days
Effective Foetal Weight	3740 gms approximately	-----	

Discussion

This term Potter's Syndrome was coined by Edith Potter, a pathologist in 1946, which has typical physical appearance associated with hypoplasia of lungs in the neonates and give rise to characteristics facial appearance along with agenesis of both the lungs in infants. This is due to oligohydramnios and compression of fetus in the utero (Edith, 1946). There is sequence of events that occurs, hence it is also known as Potter's Sequence or oligohydramnios sequence commonly used in available literatures.

If the amniotic fluid is less than normal corresponding to period of amenorrhoea, then oligohydramnios occurs. This may be due to decrease production of urine secondary to renal agenesis on both sides and obstruction to urinary tract, and sometimes prolonged rupture of membranes (Preus *et al.*, 1977; Palmer *et al.*, 2001). Pulmonary failure is the cause of death in Potter's Syndrome & bilateral agenesis is incompatible after birth (extrauterine life). In 33% foetus die in uterus & nowadays in 70% survival rate have been reported among 23 infants who had oligohydramnios & pulmonary hypoplasia antenatally (Woods and Brandon, 2007). Agenesis of both kidneys occurs in about 1 of 5000 fetuses (Slickers *et al.*, 2008). The other associated maternal high risk factors to cause agenesis of both kidneys are maternal body mass index more than 30, and smoking (Klaassen *et al.*, 2007; Paces-Fessy *et al.*, 2012). Other anomalies that occur along with bilateral renal agenesis are anomalies of vertebrae, atresia of anal canal, defects of cardia, tracheoesophageal fistula, renal defects, and defects of limbs (Kumar *et al.*, 1997). Cardiovascular anomalies, skeletal anomalies, and central nervous systems anomalies and caudal dysplasia syndrome are also found (Vanderheyden *et al.*, 2003; Uematsu *et al.*, 2006; Schmidt and Kubli, 1982; Tonni *et al.*, 2008). Alan *et al.*, (1975) have stated that a fetus may be born with normal kidney but may have nonrenal features. These features are due to uterine compression caused by premature rupture of membranes followed by amniotic fluid leakage (Alan and Thomas, 1975). A retrospective study was done on 80 cases of Potter sequence due to a renal or urologic abnormality. The abnormalities findings were renal agenesis seen in 21.5%, cystic dysplasia seen in 47.5%, obstructive uropathy in 25%; and others in 5.25%. Multiple congenital anomalies were seen in 15 patients. Three had aneuploidy, autosomal recessive syndromes were seen in 4 patients and in 8 cases, and cause was idiopathic (Cynthia *et al.*, 1984). It is Uroplakins IIIa is a protein that is secreted in mammalian urothelia is said to be

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involved in early defective development of kidney like renal hypoplasia dysplasia (Schonfelder *et al.*, 2006). Some noted genetic disorders like renal coloboma syndrome (*PAX2* mutation) & branchio-oto-renal syndrome (*EYA1* mutation) are usually with agenesis of kidney or dysplastic kidney anomalies (Lu *et al.*, 2009; Tabatabaeifar *et al.*, 2009). It is males who have higher incidence of Potter's Syndrome with higher rate of Eagle-Barrett (prune belly) syndrome (Curry *et al.*, 1984). A study has been done in 12 European countries showed data from 20 registries that was collected on 709030 live births, still births, induced abortions in which 95 cases were Agenesis of both kidneys and 86 cases were diagnosed prenatally (Colquhoun-Kerr *et al.*, 1999). Another study was made in Europe which had 17 registries. In this study 4366 cases were reported having 11 congenital anomalies of severe variety. There were 257 cases of agenesis of kidney on both sides (bilateral renal agenesis) (Prouty and Myers, 1987).



Figure 1: Radiographs showing a single live fetus with bilateral enlarged polycystic kidneys

Present Study

Has shown a single live fetus in cephalic presentation with the foetal spine to the maternal left with enlarged polycystic kidneys associated gross oligohydramnios. There was no history of low set ears and involvement of lungs, no widely placed epicanthic folds. There was neither hypoplasia of lungs nor hypoplasia of Anterior Abdominal wall. There were associated genetic disorders. NO anomalies of vertebrae, atresia of anal canal, defects of cardia, tracheoesophageal fistula, nor defects of limbs and central nervous systems. No multiple congenital anomalies found radiologically.

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Conclusion

This is study of bilateral enlarged recessive polycystic kidney with oligohydramnios suggestive of Potter's Syndrome is of rare variety among congenital anomalies, hence studied.

Take Home Message

Since it is rare variety of congenital anomaly, it has be reported to bring awareness to all surgeons especially to urologists. Incidences of such conditions of anomalies can be brought down by giving proper health education and awareness regarding consanguinity marriages and its effect to the public by health workers, NGOs, and Voluntary organizations. Hence studied and reported.

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