International Journal of Anatomy and Research,
Int J Anat Res 2016, Vol 4(4):3097-99. ISSN 2321-4287
DOI: http://dx.doi.org/10.16965/ijar.2016.411

Case Report

POTTER'S SYNDROME: A CASE REPORT

A.K.Manicka Vasuki *[1], M.Nirmaladevi [2], Deborah Joy Hebzibah [3], M.Jamuna [4], K.Radhika [5], K.Kalyana Sundaram [6].

*[1] Assistant Professor, Department of Anatomy, PSG Institute Of Medical Sciences And Research, Coimbatore, Tamil Nadu, India.

[2] Associate Professor, Department of Anatomy, PSG Institute Of Medical Sciences And Research, Coimbatore, Tamil Nadu, India.

[3] Assistant Professor, Department of Anatomy, PSG Institute Of Medical Sciences And Research, Coimbatore, Tamil Nadu, India.

[4] Professor and HOD, Department of Anatomy, PSG Institute Of Medical Sciences And Research, Coimbatore, Tamil Nadu, India.

[5] Assistant Professor, Department of Anatomy, PSG Institute Of Medical Sciences And Research, Coimbatore, Tamil Nadu, India.

[6] Associate Professor, Department of Anaesthesiology, Coimbatore Medical College, Coimbatore, Tamil Nadu, India.

ABSTRACT

Renal agenesis occurs as a result from a lack of induction of metanephric blastema by the ureteral bud. This may be secondary to defect in the formation of mesonephric duct or ureteral bud maldevelopment. Postnatal involution of multicystic kidneys may also result in solitary kidney. This may be associated with VACTERL SYNDROME or may be associated with ipsilateral urogenital anomalies. During dissection of 19 weeks old male fetus in the department of Anatomy, We found bilateral renal agenesis with Pulmonary hypoplasia. Prenatal ultrasound showed Oligohydramnios with bilateral renal agenesis. Bilateral renal agenesis may also be associated with pulmonary hypoplasia. The newborn would die shortly after birth, due to severe pulmonary hypoplasia. Hence we have to look for other anomalies in renal agenesis. The risk of other anomalies in the subsequent pregnancy is also increased because the condition may be an autosomal dominant or recessive disorder.

KEY WORDS: Agenesis of kidney, fetus, pulmonary hypoplasia, oligohydramnios.

Address for Correspondence: Dr.A.K.Manicka Vasuki, Assistant Professor, Anatomy Department, PSG Institute Of Medical Sciences and Research, Coimbatore, Tamil Nadu, India.
E-Mail: vasukikalyan01@gmail.com

Access this Article online

Quick Response code

DOI: 10.16965/ijar.2016.411

Web site: International Journal of Anatomy and Research
ISSN 2321-4287
www.ijmhr.org/ijar.htm

Received: 23 Sep 2016
Peer Review: 24 Sep 2016
Revised: None

Accepted: 02 Nov 2016
Published (O): 30 Nov 2016
Published (P): 30 Nov 2016

INTRODUCTION

Bilateral renal agenesis is an uncommon prenatal diagnosis.The incidence is one or two per 10,000 births. It is a lethal anomaly with 50% of the fetuses being stillborn. Rest of the fetus if born, may die from pulmonary hypoplasia. Associated adrenal agenesis is seen in 8 – 10% cases. It may be associated with Vertebral, Anorectal, Cardiac, Tracheo – esophageal and limb anomalies. It may be associated with

genital tract abnormalities like undescended testis, seminal vesicle cysts, uterus didelphys and vaginal atresia.

The development of Kidney begins in the fourth week of intrauterine life from intraembryonic mesoderm. Nephrogenic cord derived from intermediate mesoderm forms a longitudinal ridge on posterior abdominal wall on each side of the dorsal aorta.The Neprogenic cord forms three successive kidneys: Pronephros, Mesonephros and Metanephros – succeeding each other in time and space. The last to develop retained as permanent Kidney.

The Pronephros forms at the beginning of fourth week in the cervical region. It is nonfunctional and completely regresses. However, the Pronephros duct – which opens in the Cloaca persists, which is annexed by Mesonephros and forms Mesonephros duct.

The Mesonephros forms at the end of fourth week in thoracolumbar region. It is functional for short period and completely regresses. A series of excretory tubules develop in Mesonephros which drains into Mesonephros duct.

Metanephros forms at the beginning of third month in the sacral region. It persists permanently in humans. It drains into Ureter.Each Kidney develops from two distinct sources: 1. Metanephros, 2. Ureteric bud.

Metanephros forms Secretary System and the Ureteric bud which arises from Mesonephric duct, forms collecting system of Kidney. Later communication between the two systems takes place.

If there is a defect in the formation of Mesonephric duct or Ureteric bud, then renal agenesis occurs because of lack of induction of metanephric blastema.

MATERIALS AND METHODS

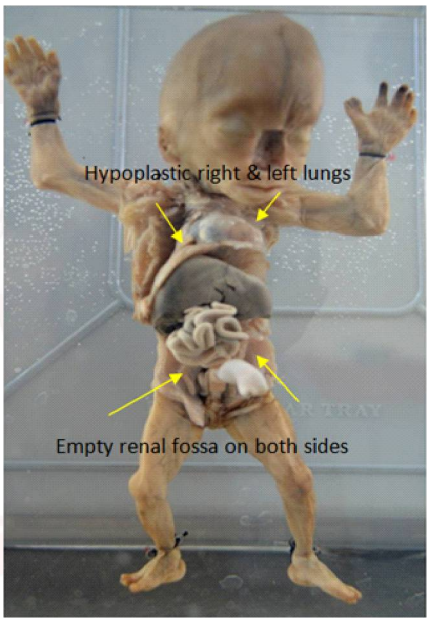
The fetus of 19 weeks collected from Department of Obsterics and Gynaecology, PSG IMS & R, Coimbatore. Permission obtained from concerned authorities and Institutional Ethical Clearance was also taken. Immediately after collection, gestational age was determined by Crown – Rump Length and maternal history. During dissection of 19 weeks old terminated male fetus in the department of Anatomy, a congenital anomaly of Bilateral renal agenesis with Pulmonary hypoplasia was observed.

Prenatal ultrasound of the fetus showed Bilateral renal agenesis with Oligohydramnios.

OBSERVATIONS

1. On opening the peritoneal cavity, there was no peritoneal fluid.
2. Both renal fossa were found to be empty. Urinary bladder could not be delineated.
3. Both lungs were found to be hypoplastic.
4. Typical facies of Potter's facies was found.

Fig. 1: Shows features of Potter's syndrome.



DISCUSSION

Kidneys are seen on either side of the spine in the posterior abdomen at 12 weeks of gestation. Bilateral renal agenesis is an incidence of 1 : 10,000. It is usually sporadic in nature. It may present with family history. Newborns with bilateral renal agenesis have low set floppy ears, broad, flat nose, redundant dehydrated skin, wide set eyes, prominent fold in the inner canthus of each eye, parrot beak nose and receding chin. These features are known as Potter's facies. Because of Pulmonary hypoplasia, these babies die due to respiratory failure within few hours of birth.

Hetal parikh [1] observed bilateral renal agenesis with lying down adrenal sign in antenatal ultrasound of 18 weeks fetus. The autopsy of

the fetus showed features of Potter's facies with pulmonary hypoplasia. Both renal fossa and pelvis were empty. This is similar to our presentation. In our study, We observed Bilateral renal agenesis, Potter's facies with pulmonary hypoplasia.

A.Mishra [2] observed right renal agenesis with compensatory hypertrophic changes in left kidney in 30 years old man. There was a dilated distal part of right ureter with hypoplastic, cystic right seminal vesicle.

Prashin C unadkat [3] observed right renal agenesis with compensatory hypertrophic left kidney accidentally in a 34 years old man who came for right inguinal hernia. The hernia sac contained right testis which was removed for suspicion of malignant change in it. There was no change in Malignancy, but the testis was found to be atrophic by microscopic examination.

Maki Kashiwagi [4] found bilateral renal agenesis, Phocomelia and single umbilical artery in antenatal ultrasound of 23 years old female with 17 weeks pregnancy who took regular abuse of Cocaine and Heroin. The terminated fetus at autopsy confirmed bilateral renal agenesis with a single umbilical artery with right upper limb reduction defect. Absent elbow joint, forked hand on the right side (rudimentary humerus, complete ulna and two carpal bones, three metacarpal bones and two fingers) were found.

Abdelmageed Abdelrahman [5] observed right renal agenesis with compensatory hypertrophic left kidney and Uterus didelphys in a 17 years old girl who presented with right iliac fossa pain. She was evaluated and found to have large cystic mass in the pelvis which was found to be haematometra on laparatomy,which was aspirated vaginally. During the surgery, Uterus didelphys and right renal agenesis were confirmed.

Doron Kabiri [6] observed Herlyn – Werner – Wuderlich syndrome, a combination of Uterus didelphys and obstructed hemivagina with bilateral renal agenesis in 20 years woman who came with lower abdominal pain and urinary retention.

CONCLUSION

Renal agenesis is a fairly common congenital anomaly with an unknown etiology. Renal agenesis may be unilateral or bilateral. This occurs with an incidence of 1 in 10,000births with a male – female ratio of 3:1. It may be familial. However it is diagnosed antenatally because of absence of amniotic fluid. Even though antenatal diagnosis of bilateral renal agenesis is uncommon, this condition is important. It is a lethal condition with 50% of the fetuses being stillborn and rest of them may die within few hours of birth due to severe pulmonary hypoplasia.

In the presence of renal agenesis, other anomalies like pulmonary hypoplasia, undescended testis, mullerian agenesis, vertebral, Anorectal, limb anomalies should be looked for. If renal agenesis is unilateral without any other anomalies, no need for termination of the fetus. After the birth the fetus should be observed periodically.

In Bilateral renal agenesis with/without other anomalies termination of the fetus should be done in view of worst prognosis.

Conflicts of Interests: **None**

REFERENCES

[1]. Hetel Parikh, Sameer Raniga et al, Antenatal diagnosis of Bilateral renal agenesis with Potter's sequence: A rare case, I.J.Clinical Practice,Mar 2012;22(10):526-28.
[2]. A.Mishra, Renal agenesis : report of an interesting case, BJR, Aug 2007;80(956):167-69.
[3]. Prashin C.Unadkat, Radha verma et al, Right undescended testis with ipsilateral renal agenesis, I.J.Sci.Study, Oct 2014;2(7):240-41.
[4]. Maki Kashiwagi, Rabih Chaoni et al, Fetal bilateral renal agenesis, Phocomelia and single umbilical artery associated with cocaine abuse in early pregnancy, Birth defects Research (Part A),2003;67:951-52.
[5]. Abdelmageed Abdelrahman, A case of Uterus didelphys and unilateral renal agenesis, Sudan Med.J, Apr 2013;49(1):41-43.
[6]. Doron Kabiri, Yaara Arzy etal, Herlyn – Werner – Wuderlich syndrome: uterus didelphys and obstructive hemivagina with unilateral renal agenesis, IMAJ,Jan2013;15:66.
[7]. A.Nickavar, M.Razzaghy – Azar et al, Pheochromocytoma and renal agenesis, Acta Medica Iranica,2008;46(1):81-83.
[8]. Shreeprasad.P.Patankar, Vijay Kalrao,et al. Mayer – Rokitansky syndrome with Anorectal malformation, I.J.Pediatrics, Dec 2013;71(12):1133-35.
[9]. Anas M.Ajazami, Ranad Shaheen et al, Am.J.um. Genet.,Sep.2011;85(3):414-18.

How to cite this article: A.K.Manicka Vasuki, M.Nirmaladevi, Deborah Joy Hebzibah, M.Jamuna, K.Radhika, K.Kalyana Sundaram. POTTER'S SYNDROME: A CASE REPORT. Int J Anat Res 2016;4(4):3097-3099. **DOI:** 10.16965/ijar.2016.411