

Left Sided Atrophic Kidney and Raised APLA in Association with Arcuate Uterus

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ABSTRACT

Introduction: The kidney is the most frequently involved organ in APS (antiphospholipid syndrome) leading to small atrophic kidney. Arcuate uterus has a single cavity with a convex or flat uterine fundus. The endometrium demonstrates a small fundal cleft or impression (>1.5 cm). Arcuate uterus is the most commonly found uterine anomaly in HSG and is sometimes responsible for causing repeated abortions and infertility.

Case report: A 39 year old female patient presented with secondary infertility and a history of repeated miscarriages. Investigations showed left sided atrophic kidney and raised antiphospholipid antibodies. HSG revealed arcuate uterus.

Conclusion: Raised APLA lead to atrophic kidney and frequent abortions. Co-existence of unilateral cystic atrophic kidney and arcuate uterus could be attributed to urogenital anomaly, contributing to repeated abortion.

Keywords: Atrophic Kidney, APLA (Antiphospholipid Antibodies), Arcuate Uterus.

INTRODUCTION

Here we report a case of left sided cystic small atrophic kidney with raised APLA and arcuate uterus presenting with secondary infertility and bad obstetric history. Renal manifestations of APS (anti phospholipid syndrome) include renal artery stenosis, renal hypertension and infarction.² and renal atrophy. Atrophic kidney is the reduction in the number and size of parenchymal cells. Causes of atrophic kidney are:

Renal Ischemia due to:

- Atherosclerosis of renal artery
- Occlusion of the lumen by antigen antibody complexes as in antiphospholipid syndrome(APS). Renal involvement appears in 2.7% of patients (2).
- Pre existing arterial narrowing or stenosis.
- Compression of blood vessels as a result of renal cyst

Damage to the renal parenchyma as a result of:

- Obstruction in the urinary system
- Long standing kidney infections such as pyelonephritis, polycystic kidney, and other chronic renal diseases that can affect the nephrons.
- Reflux nephropathy.

Studies have shown that renal atrophy and aplasia are frequently associated with uterine anomalies. Raised APLA seem to be the main incriminating factor in this case though the role of possible urogenital anomaly cannot be ruled out. Antiphospholipid Syndrome Criteria was followed.⁶ The case fitted into the criteria according to the laboratory investigations and clinical history.

Arcuate uterus has a single uterine cavity with a convex or flat uterine fundus. The endometrial cavity demonstrates a small fundal cleft or impression (>1.5 cm). The outer contour of the uterus is convex or flat.³ Arcuate uterus is the most commonly

found uterine anomaly in HSG. Though clinically benign it is sometimes associated with obstetric complications. HSG shows saddled shaped fundus. Usually no treatment is required.

CASE REPORT

A 39 year old female patient presented with secondary infertility and a history of repeated miscarriages initially at Gynae OPD, PGIMER, Chandigarh and later in Gynae OPD, Manimajra clinic, Chandigarh. Investigations showed left sided cystic atrophic kidney and raised antiphospholipid antibodies. Hysterosalpingography revealed arcuate uterus, suggesting some anomaly of the kidney as well owing to common development from the intermediate mesoderm.

Past History

- Primary infertility
- history of repeated first trimester abortions
- history of repeated episodes of urinary infection
- History of hypertension for the last 13 years
- Emergency LSCS in 2003, at 35 weeks in view of proteinuria and superimposed preeclampsia.
- Was referred to PGI in 2007 with severe urinary infection. Urine routine showed full field RBCs and albumin 4+. Urea / creatinine 28/0.9, indicating Rapidly Progressive Glomerulonephritis. Renal Doppler showed left sided atrophic kidney and right sided compensatory hypertrophy.

Obstetric / Menstrual History

Obstetric formula: G6P01A5L1.

Delivered a baby girl after 2 years of primary infertility, with LSCS. She had five miscarriages, all in the first two months of pregnancy. Age of menarche was 12 years. There is history of menorrhagia and irregular periods.

Investigations

Lab Investigations (9/3/2014)

1. RFT Blood Urea 29, S. Creatinine 0.9, S. Uric Acid 4.1
2. LFT Total Bilirubin 0.56, Direct Bilirubin 0.08, SGOT 21, SGPT 30
3. Alk. Phosphatase 78
4. Total Proteins 7.2
5. S. Albumin 4.1
6. S. Globulin 3.1

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Figure-1: USG: Left kidney shrunken, 6.9 x 2.8 cm; Right kidney enlarged 12.0 x 4.3 cm Shrunken left kidney with cysts.

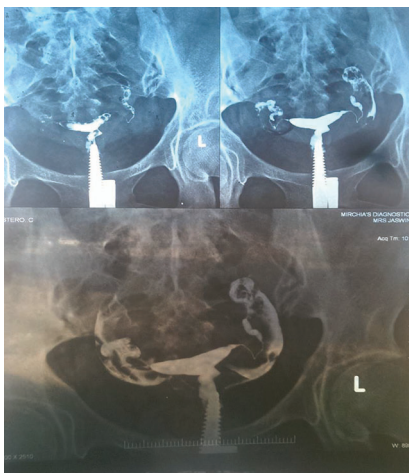


Figure-2: HSG (24/07/2012): Normal Bilateral fallopian tubes with Arcuate type of uterine cavity

7. CBC HB 10.3, TLC 8300
8. Prolactin 10.76 (6/6/2011)
9. TSH 5.79UIU/ML (6/6/2011)
10. anti phospholipid IgG 17.4 μ /ml (<15u/ml is negative), IgM 25.04 μ /ml (<15u/ml is negative).

USG (22/1/2009)

- B/L Renal Doppler Small atrophic left hydronephrotic Kidney with reduced vascularity
- NCCT KUB (16/3/2012)
 - Small cystic atrophic left kidney with compensatory hypertrophied right kidney
- DMSA Scan (21/8/2009)
 - Left kidney not visualised
 - 18/05/2009
 - Urine R/E: Full field RBCs, albumin 4+
 - Urea/Creatinine: 28/0.9 (normal is 15)

DISCUSSION

Antiphospholipid syndrome (APS) is an autoimmune disorder characterized by recurrent vascular thrombosis in the presence of raised antiphospholipid antibodies. According to Alchi, Griffiths and Jayne² The kidney is a major target organ in both primary and secondary APS The kidney is a major target organ in APS leading to renal artery stenosis and sometimes renovascular

hypertension, renal infarction and APS nephropathy.² It is commonly associated with repeated pregnancy losses.

In the present case study vascular compromise secondary to APLA and subsequent hypertension probably caused ischemic changes in the left kidney. This, complicated with chronic renal infection, ultimately resulted in an atrophic kidney (figure-1). The right kidney became hypertrophied to compensate the work load of the left kidney. Besides hindering blood supply to the growing embryo, antiphospholipids reduce HCG release and inhibit trophoblast invasion⁵, which may explain the frequent miscarriages in this case. HSG revealed arcuate uterus (refer pic: 2). A study conducted by Sotirios et al⁵ showed 12.2% of the repeated miscarriages occurring due to arcuate uterus. The case fulfils the criteria for the diagnosis of APS, with likelihood of arcuate uterus as a contributing factor for frequent miscarriages in this case

Incidence

Renal involvement appears in 2.7% of patients with antiphospholipid syndrome¹

Prevalence of congenital uterine anomalies

~6.7% in general populaion.

~7.3% in infertile population

~16.7% in RM population (Repeated Miscarriage)

Prevalence of arcuate uterus ~ 3.9% in general population.³

Prevalence of arcuate uterus ~ 12.2% in RM cases⁵

Embryological Basis of Arcuate Uterus

The uterus is formed at around 8–16 weeks of fetal life from the development of the two paired paramesonephric ducts, called Müllerian ducts. The process involves three main stages

Organogenesis - development of both Müllerian ducts.

Fusion - lower Mullerian ducts fuse to form the upper vagina, cervix and uterus (lateral fusion). Cranial part of the Müllerian ducts remains unfused and forms the Fallopian tubes.

Septal absorption - after the lower Müllerian ducts fuse, a central septum is left which starts to resorb at ~9 weeks. Defects in this stage result in a septate or arcuate uterus.⁴

CONCLUSION

The co existence APS and arcuate uterus is perhaps incidental, the former being an auto immune disorder and the latter, a developmental. It is however worthwhile for a clinician to remember this co existence while planning the management. In the setting of raised APLA, and a past history of repeated abortions, surgical correction of the uterine anomaly, depending on its magnitude, might increase the chances of pregnancy.

REFERENCE

1. Caraba A, Crisan V, Munteanu A, Serban C, Nicoara D, Romosan I. The Kidney in Antiphospholipid Syndrome, In: Antiphospholipid Syndrome, Bulikova A. (Ed). InTech, April 2012 [cited April 2012]. Available at: <http://www.intechopen.com/books/antiphospholipid-syndrome/antiphospholipid-syndrome-and-the-kidney>.
2. Alchi, B., Griffiths, M. and Jayne, D. What nephrologists need to know about antiphospholipid syndrome. *Nephrol. Dial. Transplant.* 2010;25:3147–3154.
3. Morgan MA, Yang N, et al. Arcuate uterus. Available from: <http://radiopaedia.org/articles/arcuate-uterus>
4. Sotirios H, Saravelos, Karen A, Cocksedge, Tin-Chiu Li

Prevalence and diagnosis of congenital uterine anomalies in women with reproductive failure: a critical appraisal Human Reprod Update Medicine and Health. Oxford Journals. 2008;14:415-429.

5. N. Di Simone, P. L. Meroni, N. Del Papa, E. Raschi, D. Caliandro, S. De Carolis, M. A. Khamashta, T. Atsumi, G. R. V. Hughes, G. Balestrieri, A. Tincani, P. Casali, and A. Caruso antiphospholipid antibodies affect trophoblast gonadotropin secretion and invasiveness by binding directly and through adhered β_2 -glycoprotein Arthritis Rheum. 2000;43:140–150.
6. Jose A. Gómez-Puerta, Ricard Cervera. Diagnosis and classification of the antiphospholipid syndrome. Journal of Autoimmunity. 2014;48-49;20-25.

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