CONTRAST INDUCED NEPHROPATHY & PELVIURETERIC JUNCTION OBSTRUCTION IN A PATIENT WITH MAYER–ROKITANSKY–KUSTER–HAUSER SYNDROME & SOLITARY ECTOPIC KIDNEY- A CASE REPORT

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ABSTRACT Mayer–Rokitansky–Kuster–Hauser (MRKH) syndrome is a rare congenital syndrome with the absence of uterus and vagina, affecting 1 in 5000 women. The patient has normal secondary sexual characters and the first sign of the syndrome is primary amenorrhea. There are two types of the condition in this syndrome type A without associated anomalies, and type B, which is associated with other gynecological and non-gynecological anomalies. The renal agenesis is most common in type B. The contrast-induced nephropathy is the foremost risk factor in renal agenesis. The contrast induced nephropathy is a serious complication resulting from the administration of contrast media which leads to impairment of renal function. Here we reported a rare case of a 34-year-old female patient with a history of MRKH syndrome and Solitary Ectopic Kidney presenting with fever, dyspnea, and diarrhea. The patient previously admitted to her local hospital for fever, nausea, and vomiting. The diagnostic workup was done with blood investigation, ultrasonography, and CT contrast of the abdomen. The patient's total count, blood urea, and serum creatinine levels showed a substantial increase on the following day. The patient was diagnosed to have Mullerian agenesis type II, with contrast-induced nephropathy and uteropelvic obstruction. Incidentally she was also found to have dengue fever with thrombocytopenia. She was managed with hemodialysis and DJ stenting. Patients with solitary ectopic kidney have a high risk of developing contrast induced nephropathy and one should exercise great prudence when ordering radiological investigations in these patients.

KEYWORDS Mullerian agenesis, Single kidney, Contrast-induced nephropathy, Pelviureteric junction obstruction

Introduction

Mayer–Rokitansky–Kuster–Hauser (MRKH) syndrome is a pathological condition characterized by primary amenorrhea, infertility, congenital aplasia of the uterus and upper vagina. It is also known as Mullerian agenesis characterized by malformation of (paramesonephric) Mullerian duct.[1] This condition was first described by the scientists Mayer(1829), Rokitansky (1838), Kuster (1910), and Hauser and Schreiner (1961) and later it was named as MRKH syndrome.[2][3] MRKH syndrome is a congenital disorder, with an incidence of approximately 1 in 4500 newborn female children.[1][4-6].The patient with this condition is born with normal 46, XX female karyotype with normal secondary sexual characteristics, external genitalia, regular sexual hormonal profile[1] [4]. This condition is divided into two categories, Type 1 (isolated) or Rokitansky sequence and Type 2 (associated) or MURCS association (Mullerian agenesis, renal agenesis, and Cervicothoracic Somite anomalies), were in Type 1 the structures of Mullerian duct is only affected, in Type 2 the malformation of structures of Mullerian duct is associated with abnormalities of renal, skeletal and heart[7][8]. Unilateral renal agenesis is considered to be a risk factor for contrast induced nephropathy. Contrast-induced nephropathy (CIN) is the impairment of kidney function—measured as either fractional rise in serum creatinine (S.Cr) of 50 -100% from baseline or a

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0.5 mg/dL (44 µmol/L) increase in absolute SCr value—within 48-72 hours after intravenous contrast administration.[9][10]

We report a patient with MRKH syndrome, presenting with contrast-induced nephropathy, pyelonephritis, and pelvic ureteric junction obstruction(PUJO) accompanied by dengue fever with thrombocytopenia and anemia.

Case report

A 34-year-old female patient who is a known case of Mullerian agenesis type 2 with solitary kidney presented to the Emergency department of a hospital with chief complaints of fever for 2 weeks, dyspnea, and diarrhea for 3 days. The patient had previously visited her local hospital for the complaints of fever, nausea and vomiting. She was admitted and baseline blood investigations were carried out in that hospital which included complete blood count, renal function test (RFT) and serum electrolytes to identify the cause of persistent fever. Ultrasonography and CT Abdomen with pelvis-contrast were ordered to determine the extent of renal comorbidities. Thrombocytopenia was noted. Ultrasound revealed she had a solitary ectopic kidney. CT contrast of the abdomen was ordered for detailed visualization. Contrast Computer tomography (CT) of her abdomen showed mild hepatosplenomegaly, right large ureteric calculus with moderate obstructive uropathy, the right kidney was grossly enlarged in size but normal in shape, malrotated ectopically placed in right iliac fossa with pelvis directed superiorly and gross hydronephrosis with non-excretion of contrast in right collecting system and significant perinephric fat stranding – pyelonephritis, diffuse tumefactive gall bladder sludge, early umbilical hernia, displaced malunited fracture in bilateral inferior pubic rami. Uterus and ovaries were not visualized. The patient was admitted for a detailed work up of fever of unknown origin. A fever testing for dengue antigen turned out to be positive. After 24hours of taking CT abdomen, the RFT and CBC was repeated, which showed a remarkable increase in the total count, blood urea, and serum creatinine levels as compared to the previous day report (table1.1). In view of serum creatine being 7.8mg/dl, patient was shifted to our centre for further management.

The patient had a temperature of 100°C, respiratory rate - 26bpm, pulse-103 bpm, BP- 130/90mmHg, and SPO2 of 93% on room air. She weighed 52 kg. The patient was conscious and oriented, pale and had bilateral pedal edema. She also had characteristic shortness of breath. Her bowel and bladder habits had no changes. She did not complain of abdominal pain and her VAS score was 0/10. Laboratory test results (Table 1) showed random blood sugar 115 mg/dl, Anti CCR-13.0 U/ml, ANA-10.5 U/ml. Blood levels of potassium and liver function tests were normal.Thus on the basis of the clinical findings, the patient was diagnosed with Mullerian agenesis type II, right kidney pyelonephritis and contrast-induced nephropathy, dengue fever with thrombocytopenia, anduteropelvic obstruction.

A multidisciplinary approach was initiated in the treatment plan for the management of infection, correction of renal impairment, and anemia. On the second day of hospital stay, urologist opinion was obtained who suggested a plan for percutaneous nephrostomy(PCN) followed by right ureteroscopic DJ stenting. The patient was seen by an interventional radiologist and right PCN was performed using aseptic precautions under CT guidance.

Nephrologist opinion was also taken who suggested for 3 sittings of heparin free hemodialysis to minimize further kidney injury and bring down serum creatinine as well as BUN levels to a normal range. In view of dialysis, a right IJV double lumen catheter was inserted under aseptic precautions. The drug treatment plan consisted of Inj. Meropenem 1gm TDS and Inj.N-Acetylcysteine 1200mg bd for containment of infection and treatment of contrast-induced nephropathy. The dialysis regimen for three days consisted of Day1 - 2.5hrs, Day 2&3 for 4hrs each which the patient withstood and there was a notable change in the renal parameters when monitored (table1.1). Pulmonologist opinion was taken with regards to tachypnoea who included Tab. Theophylline/Ambroxol OD and O2 with face mask 6 at L/min. ABG was done for the patient which showed metabolic acidosis, that prompted the start ofsodium bicarbonate infusion.

As a pre-operative workup, echocardiogram was taken which showed tiny vegetations of about 6mm situated in the aortic valve. To further assess for abnormalities, the patient underwent transesophageal echocardiogram for evaluation of vegetation on the following day, which showed tiny vegetations in the non-coronary cusp of the aortic valve (2-3mm) and no other abnormalities were noted.She was started on anticoagulation with regular monitoring of PT/INR. After obtaining anesthetic fitness, the patient was scheduled for right DJ stenting with ureteroscopy under moderate risk. The patient withstood the procedure well and check USG KUB (Figure 1&2)was taken to find out the placement of the stent. On postoperative day 1,patient had hypokalemia and anemia (serum potassium was 2.8 mEq/l and Hb was 7.2 mg/dl). Hence 1 unit of PCV was transfused and 40mEq/l of potassium was infused for potassium correction. The check USG KUB showed a single kidney with normal renal parenchymal structure, the stent was in position and the pigtail catheter was in situ.

On the second postoperative day, the patient developed two episodes of fever spikes ranging from 99-100°C, 2episodes of loose stools for which antipyretic medication and antibiotic was started. Her other vital parameters were stable with a pulse rate of 82bpm, BP-120/80mmHg.Another unit of PCV was transfused as her Hb was still seen at 8.2 mg/dl. The right PCN catheter was removed and the patient was self-voiding from postoperative day 2. The patient got discharged on the 3rd postoperative day as her vital parameters were stable with no fever spike in the last 24hrs.

The patient got readmitted after 10 days with the chief complaints of decreased urine output, nausea, and vomiting along with abdominal distension; the patient was provisionally posted for stent removal. On general examination the vital parameters were stable, the right PCN catheter was in situ, and the Foleys catheter was also in situ with no signs of infection. Baseline investigations such as RFT and USG KUB showed remarkable improvement in renal parameters. The PCN catheter was clamped, and it was eventually removed on 2nd day of admission. The patient tolerated the procedure and was advised discharge the following day with foley's catheter. The patient was scheduled for a follow-up visit after 5 days from the date of discharge. During follow up, the patient was stable and did not have any symptoms.

Discussion

Mayer-Rokitansky-Kuster-Hauser syndrome is a rare congenital syndrome of the female reproductive tract with aplasia or hypoplasia of structures of the Müllerian ducts.[11]. The principal presentation of this patient is primary amenorrhea, normal ex-

Table 1

Lab Investigation	Heamoglobin (g/ dl)	Total count (cells/cumm)	Platelets (laks/cumm)	Urea (mg/dl)	Creatinine (mg/dl)	Sodium (mmol/L)	Potassium (mmol/L)
Outside hospital before contrast	7.8	24,900	-	24	0.8	138	-
Outside hospital after contrast	7.8	29100	0.68	210	7.8	130	-
Day 1	7.4	33200	0.73	162	8.1	109	3.7
Day 2	-	-	1.09	162	8.2	115	3.9
Day 3	-	20000	1.25	133	7.0	128	-
Day 4	-	12000	1.78	59	3.1	132	-
Day 5	-	8900	-	40	1.5	140	-
Day 6	-	-	-	38	1.4	-	-
Day 7	-	6100	-	36	1.6	142	-
Day 8	-	-	-	35	1.7	-	-
Day 9	7.2	-	-	36	1.3	145	2.8
Day 10	-	-	-	24	1.2	-	3.2
Day 11	8.2	3700	-	18	1.0	-	3.2

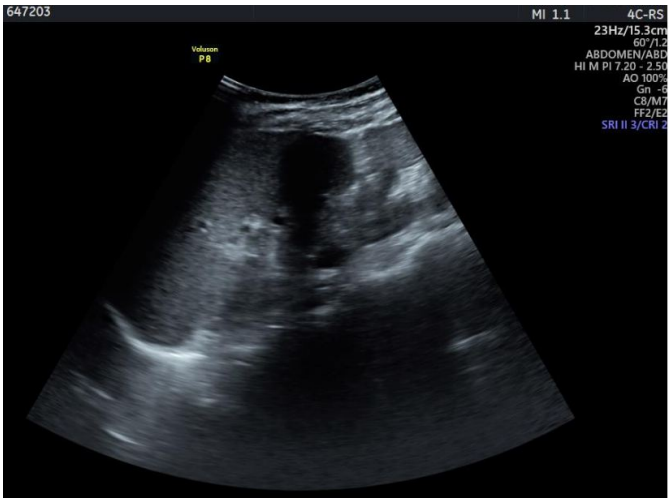

Figure 1: Single pelvic kidney measures 10.1*4.9 cm with normal renal parenchymal echoes and maintained.

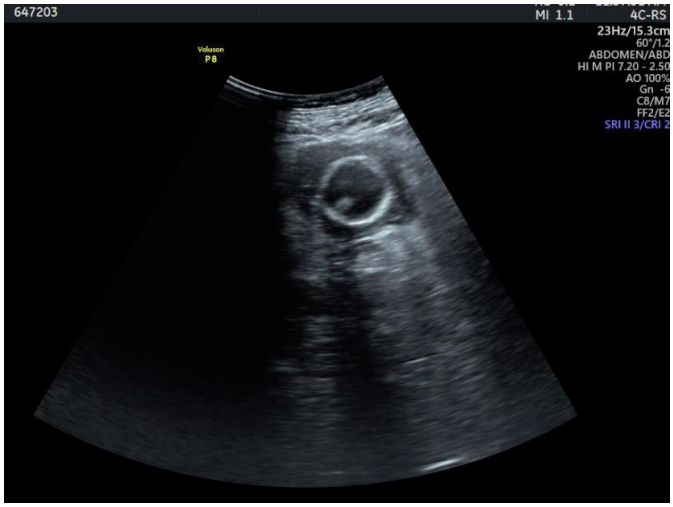

Figure 2: Cortico medullary differentiation is observed with pigtail catheter in situ.

ternal genitalia with a congenital shallow vaginal pouch, abnormalities in internal genitalia, and normal secondary sexual characteristics with pubic hair and breast development.[12][13]The diagnosis of such patients is usually identified at puberty due to other health problems, very rarely diagnosed at birth.[14]. The diagnosis is confirmed by the radiological investigation.[15]

In 2006 Anup Kumar et.al. reported a case on management of an unusual case of Mayer–Rokitansky–Kuster–Hauser syndrome, with unilateral gonadal agenesis, solitary ectopic pelvic kidney, and pelvic ureteric junction obstruction. The treatment consisted of an open pyelovesicostomy. In 2007 Pankaj Wadhwa et.al. reported a case of contrast-induced obstructive anuria in ureteropelvic junction obstruction secondary to a crossing vessel in a solitary kidney in which patient underwent an intravenous urogram (IVU) with iodinated contrast media post-procedure. This patient had reported symptoms of increased flank pain, anuria, azotemia, mild metabolic acidosis, and fluid overload. In 2018, Saleh Bubishateet.al. reported a case on MRKH syndrome with a solitary ectopic pelvic kidney and UPJ obstruction managed with open pyeloplasty.

On the basis of the types, the patient report presented here belongs to, type II MRKH or MURC syndrome wherein along with hypoplasia of Mullerian duct, she also had involvement of the renal system.[10].The patient had a single kidney with features suggestive pyelonephritis and contrast-induced nephropathy. She was treated by placement of a PCN catheter to drain out the excess contrast medium followed by surgical procedure. She also underwent 3 sessions of hemodialysis.

Conclusion

The case report along with the recent studies suggest that doctors who treat a patient with a single kidney associated with MRKH syndrome, should always be cautious in using any contrast medium for diagnostic purposes, to prevent further damage of renal function. A periodic review of renal parameters must be done during the treatment regimen.

Conflict of interest

There are no conflicts of interest to declare by any of the authors of this study.

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