Case Report

Unilateral duplicated collecting system and ureter with severe hydroureteronephrosis and ectopic ureter insertion of upper pole moiety: A case report

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ABSTRACT

Introduction: Incidence of duplicated urinary system is 0.7–4% of population, mostly are females and often diagnosed in childhood. Various symptoms meet difficulties to be diagnosed.

Case presentation: A 20-years-old woman admitted to hospital with fever, fatigue, nausea, loss of appetite, colic epigastric pain and right flank pain since 5 days before admission, normal pattern of urination with dribbling and recurrent urinary tract infection. Abdominal ultrasonography showed complicated cystic tumor upper pole of right kidney. Abdominal computed tomography with contrast showed enlargement right kidney with duplicated collecting system and duplicated ureter obstruction (severe hydroureteronephrosis right upper pole moiety) right kidney and ectopic ureter insertion of upper pole moiety between urethra and anterior vagina, no insertion to bladder, seemed to be dead end.

Discussion: Various symptoms of duplicated collecting system are asymptomatic, flank pain, abdominal pain, urinary incontinence, and recurrent UTI, often accompanied by abnormality of upper pole or lower pole or both. Abnormality of upper renal moiety usually has ectopic ureter as in Weigert-Meyer principle. Insertion into infrasphincter usually manifests as urinary dribbling or urinary incontinence or normal urination with few volume leakage or spotting incontinence, whereas suprasphincter usually manifests as recurrent UTI without incontinence. There are imaging modalities for diagnosing these anomalies, including USG and abdominal CT with contrast.

Conclusion: Understanding embryology, symptoms, imaging modality, and complications are necessary to consider this diagnosis for early detection. Ultrasonography and abdominal CT with contrast can be used to diagnose the urinary tract anomalies, especially duplicated urinary systems with ectopic ureter insertion.

1. Introduction

Duplicated collecting system and duplicated ureter are congenital urinary tract anomalies and can be accompanied by ectopic insertion of ureter [1–3]. The symptoms are various from asymptomatic, hematuria, flank pain, abdominal pain, incontinence, and recurrent urinary tract infection (UTI) [1,2]. The incidence of duplicated urinary system is 0.7–4% of population, mostly are females, and often diagnosed when childhood [2]. Assessment and diagnosis can be done with some modalities, including ultrasonography (USG) and computed tomography (CT). This case presented a 20-years-old with duplicated collecting system and duplicated ureter with right kidney obstruction and ectopic ureter insertion of upper pole moiety. Although it is a congenital anomaly, various symptoms meet difficulties to be diagnosed since childhood.

2. Case report

A 20-years-old woman admitted to hospital with fever, fatigue, nausea, loss of appetite, colic epigastric pain and right flank pain since 5 days before admission. Pattern of urination was normal with dribbling. There was no urinary incontinence or increasing urinary frequency. No urine came out from vagina or anus. There were history of recurrent epigastric pain as well as right flank pain 2 years ago and recurrent UTI.
since childhood.

Physical examinations were febrile at 37.9 °C, blood pressure was 127/72 mmHg, heart rate was 82 times per minute, respiratory rate was 20 times per minute, oxygen saturation was 99%. Abdominal examination was normal bowel sounds, tympanic in percussion, epigastric pain in palpation, positive costovertebral angle tenderness, and no Mc Burney tenderness. Laboratory results of urea and creatinine were normal, Salmonella titer was negative, SARS COV-2 antigen was negative. Thorax X-ray was normal.

Abdominal ultrasonography showed in Fig. 1 with differential diagnosis such as right kidney hemorrhagic cyst, right adrenal cystic tumor. No stone was found in kidney. Liver, pancreas, spleen and gall bladder was seen normal by ultrasound and there was no wall thickening or stone in bladder.

Abdominal CT with contrast showed in Fig. 2. It showed bulging upper pole and right kidney enlargement around 15 cm with normal location and excretion function, thinning upper pole parenchyma density, normal mid and lower poles parenchyma density. There was duplicated collecting system with severe dilated upper pole (slightly high density water attenuation). Neither dilatation of lower pole collecting system nor stone of upper and lower poles was seen. There was duplicated right ureter, diffuse dilated upper ureter from proximal to distal, ectopic ureter insertion between urethra and anterior vagina, no insertion to bladder, seemed to be dead end.

Conservative management was done by giving antibiotics and symptomatic therapy. The patient discharged with improvement of the symptoms. This case report has been reported in line with the SCARE Criteria [4].

3. Discussion

Duplicated collecting system is a congenital anomaly where two collecting systems of kidney drain to each ureter at upper pole and lower pole. In normal embryology, ureteric bud that develops from Wolfian duct as primitive mesonephric, migrates and fuses with metanephros as kidney precursor then forms nephron as kidney parenchyma functional unit. Metanephros induces bifurcation of ureteric bud to form renal collecting system, including calyx and renal pelvis. Failure in this process will cause anomalies in urinary system [2,5,6]. This case reported a complete duplicated collecting system and duplicated ureter. The process occurred might be metanephros induced two ureteric buds to bifurcate into two separated collecting systems [2].

The symptoms are various from asymptomatic, hematuria, flank pain, abdominal pain, urinary incontinence, and recurrent UTI [1,2]. Complications of these anomalies also can manifest in symptoms, such as obstruction, reflux, ureterocele, and infection, usually seen in complete duplicated urinary systems [7]. In this case, the patient felt fever, fatigue, nausea, loss of appetite, colic epigastric pain and right flank pain, normal pattern of urination with dribbling and no urinary incontinence or increasing urinary frequency. There were history of recurrent epigastric pain as well as right flank pain 2 years ago and recurrent UTI since childhood.

Ultrasonography as initial imaging modality can be used to detect major abnormalities like hydronephrosis or ureterocele [8]. In this case, abdominal ultrasonography showed complicated cystic tumor upper pole of right kidney. Ultrasonography findings of kidney with duplicated collecting system are asymmetrical renal length, abnormal parenchymal contour and asymmetrically dilated pelvicuretecal of upper pole and lower pole [2]. However, minor abnormalities can be missed, particularly the complete tract and the insertion of ectopic ureter [6].

Hence, abdominal CT with contrast was chosen in this case to evaluate another anatomical abnormalities and complications. Computed tomography can detect duplicated urinary systems, especially in case of nonfunctioning moiety, anomaly with duplications such as ectopic ureter, and ureterocele [8]. In this case report, there was right kidney enlargement with duplicated collecting system and duplicated ureter obstruction (severe hydroureretonephrosis of right upper pole moiety) right kidney and ectopic insertion of the upper pole moiety between urethra and anterior vagina, no insertion to bladder, seemed to be dead end.

Duplicated collecting system is often accompanied by abnormalities of upper pole or lower pole or both [2]. In Weigert-Meyer principle, upper renal moiety ureter has ectopic insertion medially and inferiorly to lower renal moiety ureter, and ureterocele. Whereas lower renal moiety ureter has orthotopic insertion laterally and superiorly to upper renal moiety ureter and vesicoureteral reflux (VUR). Stephen’s postulate breaks this principle about ectopic pathway that ectopic ureter is not only drained distally to normal ureter insertion but can be medially and superiorly to normal insertion [1–3]. Duplicated collecting system with duplicated ureter and ectopic insertion of ureter can cause several complications. The farther insertion of ectopic ureter from the orthotopic location, more dysplasia are found at upper pole parenchyma. Dysplasia of the parenchyma or scarring due to VUR or uropathy obstruction, particularly in upper pole ectopic ureter, causes dilatation of collecting system (hydroureteronephrosis) and contributes to lithiasis, ureterocele, as well as decreased upper pole function [2,5,9]. In this case report, there was severe hydroureretonephrosis of right upper pole moiety.

In 80–85% cases of ectopic ureter are related to duplicated collecting system. Locations of ectopic ureter in female are bladder neck, upper urethra (33%), vaginal vestibule between urethra and vaginal opening (33%), vagina (25%), and cervix or uterus (<5%) [10,11]. In patients with insertion of ectopic ureter into below external urethral sphincter (infrasphincter), they often feel urinary dribbling or urinary incontinence or normal urinary pattern with few volume leakage or spotting incontinence [3]. Whereas in patients with insertion into or above bladder neck and upper urethra (suprasphincter) usually got recurrent UTI and no incontinence [3]. In this patient, the insertion was between urethra and anterior vagina, it seemed to be dead end. The patient felt normal urinary pattern with dribbling and recurrent UTI since childhood without incontinence.

Other abnormalities such as renal dysplasia and non-genitourinary anomalies (congenital heart disease, spinal cord malformations, anorectal malformations, etc.) can be related to ectopic ureter [3], but these were not found in this case.

Management of duplicated collecting system, duplicated ureter and ectopic ureter is based on how the symptoms affect patient’s life, effects to kidney function and age. Conservative management is one of choice in adulthood with less troublesome symptoms, but not reducing risk of complications. Curative treatment such as surgery is considered when the symptoms interfere quality of life. In patients with upper pole moiety and poor kidney function, related to ectopic ureter, upper pole
heminephrectomy can be done. Otherwise ureter reimplantation is considered when the upper pole function is preserved. The other alternatives are uretero-ureterostomy or uretero-pyelostomy to drain upper pole system to lower pole [12].

4. Conclusion

Duplicated collecting system and ureter symptoms can be various from asymptomatic to symptomatic due to complications such as obstruction, reflux, ureterocele, and infection. Understanding embryology, symptoms, imaging modality, and complications are necessary to consider this diagnosis for early detection. Ultrasonography and abdominal CT with contrast can help physicians to diagnose the urinary tract anomalies, especially duplicated urinary systems with ectopic ureter.

Patient perspective

The patient did not present his point of view.

Provenance and peer review

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Ethical approval

This article type (case report) does not require a formal ethical committee approval. Access to data was approved by the head of the department.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this
journal on request.

**Author contribution**

Melissa Angela Chionardes: Data collection, reference collection, drafting and editing the manuscript, Aldrich Kurniawan Liemarto: Data collection, reference collection and editing the manuscript, Siska Liana Gunardi: Data collection, final editing of the manuscript.

**Registration of research studies**

This is not an original research project involving human participants in an interventional or an observational study but a case report. This registration was not required.

**Guarantor**

Aldrich Kurniawan Liemarto.

**Declaration of competing interest**

No Conflicts of Interest.

**Appendix A. Supplementary data**

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2022.103255.

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