



Bilaterally-Complicated Horseshoe Kidney Associated to Situs Inversus with Levocardia: A Case Report

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ABSTRACT

Aim: To present a very rare case of bilaterally-complicated horseshoe kidney by nephrolithiasis in a patient with situs inversus with levocardia and skeletal anomalies.

Case Report: A 41-year-old female presented by right loin pain. Abdominal ultrasound showed bilateral kidney stones with malrotation, right mild hydronephrosis and transposition of the abdominal organs. Chest X-ray showed the heart normal in the left side. Also, computed tomography revealed a horseshoe kidney with bilateral stones. After counseling the patient about the stones treatment options, she preferred extracorporeal shock-wave lithotripsy which was tried on the right side firstly, but it failed. So, percutaneous nephrolithotripsy was scheduled.

Discussion: Situs inversus is a spectrum of congenital anomalies. Situs inversus totalis is the commonest variety and was reported with renal congenital anomalies like horseshoe kidney, acquired renal diseases like renal cell carcinoma, and in living kidney donation. However, situs inversus with levocardia is a rare variety with a known association to the congenital heart defects. Its association to a complicated horseshoe kidney and scoliosis could not be found in the English literature before the current case. Moreover, absence of congenital heart defects is extremely rare.

Conclusion: Association of situs inversus with levocardia to skeletal anomalies and complicated horseshoe kidney is a unique situation. It indicates meticulous evaluation and management.

Key Words: Horseshoe kidney, Levocardia, Nephrolithiasis, Situs inversus

INTRODUCTION

Situs inversus anomaly is the mirror image transposition of the thoracic and abdominal organs around the midline with multiple variants[1]. Situs inversus totalis includes dextrocardia representing the complete extreme variant of the spectrum[1, 2]. This spectrum could be associated to renal anomalies like horseshoe kidney which is the commonest renal fusion anomaly with an isthmus connecting the lower poles. However, situs inversus with levocardia or isolated levocardia is an extremely rare variant [1, 3]. The heart is seen in its normal levo position in the chest, while the abdominal viscera are in the dextro position [1, 4, 5]. It is mostly associated to congenital heart defects [1]. Few cases without major heart anomalies lived long [6, 7]. So, the diagnosis of the combination of situs inversus anomalies to horseshoe kidney and management of its overt cases are challenging[8]. This article describes a rare case of horseshoe kidney complicated

by bilateral stones in a patient with situs inversus with levocardia and skeletal anomalies.

CASE REPORT

A 41-year-old female patient presented by right loin pain of many months duration. She was single with no previous significant medical or urological history. On examination, she was tall, and generally looked healthy. Abdominal ultrasound revealed liver and spleen transposition, bilateral kidney stones, mildly dilated right kidney pelvicalyceal system and malrotation. Also, Kidney-Ureter-Bladder imaging films showed bilateral dense kidney stones with right lumbar scoliosis and right-sided small sacral bone defect (Figs. 1 and 3). Chest X-ray showed the heart in the normal left-sided position and so its apex direction (Fig. 2). Transverse cuts of the non-contrast computed tomography of the abdomen

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confirmed the diagnosis of situs inversus of the abdominal organs, horseshoe kidney with its isthmus and mildly dilated right pelvicalyceal system, tilted vertebrae of scoliosis, and nephrolithiasis (Fig. 4). Both kidney moieties were low in position and the right one was relatively higher and larger than the left (Figs. 3 and 4). In the right moiety, computed tomography described two stones; pelvic (16 mm x 14.6 mm) and upper calyx (3 mm x 2 mm). Also, there were two left stones; pelvic (11.4 mm x 9.7 mm) and lower calyceal (3 mm x 3 mm) stones (Fig. 3 and 4). The average Hounsfield Unit values of the right and left kidney stones were 604 and 610, respectively. Examination revealed no upper respiratory tract disorders. Also, she was virgin and the gynecological evaluation was irrelevant. Cardiac evaluation by electrocardiogram and echocardiography revealed normal findings.

The patient was counseled for the anomalies and the possible lines for stones treatment which were extracorporeal shock wave lithotripsy and percutaneous nephrolithotripsy. She preferred shock wave lithotripsy to percutaneous nephrolithotripsy. Considering the anatomical challenges of situs inversus, scoliosis and the mild degree of hydronephrosis, I agreed to this preference as an initial option. Unfortunately, two sessions of shock wave lithotripsy failed to affect right side stone topography. So, percutaneous nephrolithotripsy was scheduled. However, the patient seemed to be unconvinced with this decision and she did not fit for the date of operation.

DISCUSSION

Etiology of situs inversus anomalies was attributed to the biological expression of a certain protein that affects the heart looping during embryogenesis. Mostly, it has an autosomal recessive inheritance pattern [2]. Organ situs anomalies represent a spectrum and classified into situs solitus (the normal variant), situs inversus totalis (the commonest), situs inversus with levocardia, isolated dextrocardia and situs ambiguus [1].

Horseshoe kidney is a fusion anomaly with a reported incidence of 0.25% [9]. It is attributed to the abnormal migration of the posterior nephrogenic cells. However, mechanical forces may have a role in the anatomical orientations of the horseshoe kidney including the relatively low position and malrotation [10]. The typical anatomical findings were described and illustrated in the present case.

The urologist may meet situs inversus anomalies as an association to one of the following genitourinary disorder categories; congenital disorders only [10, 11], acquired disorders only [8], combination of congenital and acquired disorders [12] or without any disorders like in living kidney donation [13].

The present case belonged to the combined disorders category. Coincident combination of the multiple unrelated rare anomalies is very rare medical situation [10]. Therefore, further congenital or acquired disease combinations usually attribute to extremely rare situations. This could be applied to the current combination of situs inversus with levocardia, skeletal anomalies, and horseshoe kidney which was complicated by nephrolithiasis and hydronephrosis. Moreover, an extremely rare feature in this case of situs inversus with levocardia is the absence of the congenital heart defects which were reported up to 100% of cases [1]. Similarly, Imamura et al. [3] and Jo et al. [5] reported single cases of this variant without congenital heart defects in adults. Only, few cases of situs inversus with levocardia were reported with urological disorders including adrenal adenoma [7] and micropenis [3]. However, similar presentations to the current case included cases of horseshoe kidney and the common variant situs inversus totalis combined to cilia defects syndromes like Kartagener syndrome [12], or combined to Klippel-Feil syndrome. These syndromes consist of the heart anomalies, skeletal anomalies including scoliosis and renal anomalies including the horseshoe kidneys [14].

Association of situs inversus to nephrolithiasis is a rare finding [15]. Moreover, association of horseshoe kidney/situs inversus combination to nephrolithiasis could be detected very rarely in publications [2]. Accordingly and again, addition of a common congenital or acquired disorder to a rare anomaly, solely or in combinations, will create a rare situation too. The evidence evolves from the literature, where nephrolithiasis is common with horseshoe kidney reaching up to 60% [9], but it is rarely reported in combinations of situs inversus and horseshoe kidney [2].

Anatomical and metabolic abnormalities of horseshoe kidney predispose to nephrolithiasis [9]. In such cases, shock wave lithotripsy is the preferred treatment method for small stones (≤ 15 mm), while percutaneous nephrolithotripsy is the standard line of treatment for larger stones or after failed shock wave lithotripsy. However, anatomical challenges are considered in decision-making [9, 11]. Accordingly, relatively hard stones with multiple rare anatomical anomalies may indicate the non-invasive lines of treatment. So, in the current case, shock wave lithotripsy was tried firstly, although it failed, mostly, due to stone impaction or localization issues. Then, shift to percutaneous lithotripsy was the logic following step. However, missing of the patient at the scheduled dates might make the situation incomplete.

To the best of my knowledge, the current case is the first one to be reported with this constellation of congenital anomalies and presented by urological symptoms; situs inversus with levocardia (without congenital heart defects), skeletal deformities and horseshoe kidney complicated by nephrolithiasis and hydronephrosis.

CONCLUSION:

Association of situs inversus with levocardia (without congenital heart defects) and skeletal anomalies to a bilaterally nephrolithiasis-complicated horseshoe kidney is very rare entity and novel urological situation. Meticulous evaluation is indicated to exclude the high possibilities of congenital heart defects co-existence and choose the best line for treatment of complications.

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Ethical approval

Ethical committee of the Faculty of Medicine in Assiut University approved this case study. Informed consent was taken from the patient.

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Figure 1: Plain Kidney-Ureter-Bladder: Bilateral kidney stones and stomach gaseous shadow on the right side above the colonic shadow.



Figure 2: Chest X-ray: Levocardia with left-sided directed heart apex.



Figure 3: Computed tomography KUB image: Bilateral stone distribution refers to low kidneys position (note the higher levels of the right stones). Small sacral bone defect.

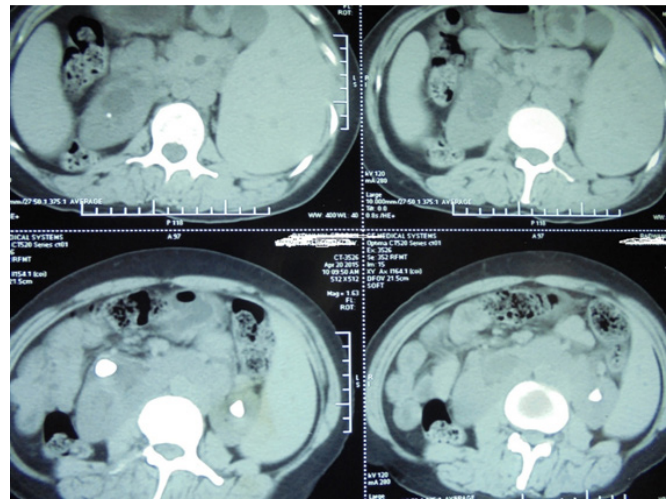


Figure 4: Computed tomography transverse cut images: Abdominal situs inversus; horseshoe kidney with bilateral stones, right hydronephrosis and lower poles fusion. Note the relatively larger and higher right kidney moiety and the tilted vertebrae of scoliosis. (Scratches were done for obscuring patient's name)