CASE REPORT

Thoracic kidney incidentally detected on renal dimercaptosuccinic scintigraphy (DMSA): a rare case report and review of literature

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ABSTRACT

Background: Thoracic kidney with diaphragmatic eventration is an extremely rare finding, and its clinical significance is not well documented. Patients with thoracic kidneys are usually asymptomatic, and the condition is usually discovered incidentally during radiological evaluation for other conditions or during thoracic surgery. Dimercaptosuccinic acid scintigraphy (DMSA) scan is a renal imaging modality that is widely used for the evaluation of renal function and anatomy.

Case Presentation: In this case report, we present a case of a patient with a thoracic kidney incidentally detected on a renal DMSA scan, along with a review of the literature. A 16-year-old female patient was referred to the Nuclear Medicine Department for a renal DMSA scan to evaluate the renal functions, as her right kidney was not visualized on her routine ultrasound examination. A DMSA scan was performed according to standard institutional protocol. Chest X-ray and computed tomography (CT) scan were done later on by the urologist, who also confirmed the kidney position and showed right-sided diaphragmatic eventration.

DMSA scan showed bilateral normal functioning kidneys; however, the right kidney was located ectopically in the thoracic cavity. Chest X-ray and CT scans also confirmed the kidney position and showed right-sided diaphragmatic eventration. Since the patient was asymptomatic, no further intervention was recommended by the urologist, and regular follow-up was suggested.

Conclusion: The renal DMSA scan is a useful imaging modality in the evaluation of the thoracic kidney as it gives us a good idea about the kidney's location, morphology, and function.

Keywords: Case report, diaphragmatic eventration, renal scintigraphy, DMSA scan, thoracic kidney.

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Background

Renal Dimercaptosuccinic acid scintigraphy (DMSA) scan is an imaging modality that is widely used in nuclear medicine to delineate renal morphology, to evaluate the presence of renal scarring, and to detect renal cortical defects. It involves the injection of a radioactive tracer that is taken up by the renal cortex and allows for the visualization of the kidneys' morphology and cortical function. The scan comprises static imaging of the kidneys, 2-4 hours after radioactive injection. A DMSA scan has a sensitivity of 85% and specificity of 97% for detecting renal anomalies. However, a rare finding on renal DMSA scan is the identification of a thoracic kidney, which can either be a congenital anomaly or an acquired medical condition [1,2]. Computed tomography (CT) of the kidney, ureter and bladder (CT KUB), CT urography (CTU), and MR urography (MRU) may also be required for complete evaluation of the different clinical conditions associated with thoracic kidney [3]. The management and clinical significance of the thoracic kidney depend mainly on the underlying etiology. However, in this case, we present a case of a thoracic kidney detected incidentally on a renal DMSA scan and discuss possible etiologies and clinical implications of this rare finding.

Case Presentation

A 16-year-old female patient was referred to the Nuclear Medicine Department, Swat Institute of Nuclear Medicine, Oncology & Radiotherapy (SINOR), Saidu Sharif Swat, KPK, Pakistan, for a renal DMSA scan to evaluate the location and function of her right kidney, which was not visualized on her routine ultrasound examination. The

patient presented with complaints of occasional right flank pain and had a history of pulmonary tuberculosis in childhood. Renal DMSA imaging was performed two hours following 99mTc-DMSA administration using dual-head single-photon emission CT gamma camera with a low-energy high-resolution collimator. Six planar images of 500 Kilo Counts on a 128 × 128 matrix format with adjustable zoom (1.5-3.2) were taken in anterior, posterior, right anterior oblique, left anterior oblique, right posterior oblique, left posterior oblique, left lateral, and right lateral projections. Radioactive markers were placed at anatomical landmarks anteriorly, i.e., umbilical level and Xiphoid-sternal level, to gauge the position of the kidneys. Relative renal function was evaluated on the basis of the posterior image after background correction. The concerned urologist performed a CT scan and chest X-ray and confirmed the presence of a right thoracic kidney with eventration of the right hemidiaphragm.

Results

Anterior, posterior, right anterior oblique, left anterior oblique, right posterior oblique, left posterior oblique, and left lateral and right lateral projections, taken from DMSA, are shown in Figure 1(a); however, radioactive markers are shown in Figure 1(b).

The images showed the left kidney located at the normal anatomical position with a relative function of 54%, as demonstrated in Figure 1(a). The right thoracic kidney was well-formed with relative functioning mass of 46% as shown in Figure 1(a). However, it was not visualized in the right renal bed but was identified in the right thoracic cavity region, as shown in Figure 2. CT images along with chest X-ray recommended by the urologist for further evaluations of the right kidney are shown in Figures 2-4.

The patient was asymptomatic, and the urologist recommended no further intervention. The patient was asymptomatic and her urologist recommended no further intervention. She was advised to have regular follow-up with primary care physician and to have periodic renal function tests.

Discussion

Renal ectopia refers to a kidney situated in any location other than the renal fossa. An ectopic kidney can be in the pelvic, iliac, or abdominal region or can be contralateral, or cross-located. Thoracic ectopic kidney is the rarest type of ectopic kidney with a reported finding of one in every 15,000 autopsies. Wolfromm in 1940 reported the first case of intrathoracic kidney using retrograde pyelography [2,4]. More than 200 cases have been reported since then, and this condition shows male predominance [2]. It is mostly observed on the left side (62%); however, it can also be located on the right side (36%) or bilaterally (2%) [2]. Interestingly, in our case, the thoracic kidney was seen on the right side in a female patient with diaphragmatic eventration. Diaphragmatic eventration with an ectopic intrathoracic kidney itself is a very rare disorder with the incidence of 0.25% of all ectopias as reported by Balela et al. [2,4-6,8]. Clinical significance of thoracic kidney depends on the underlying etiology, which may be either congenital or acquired.

Various etiologies of congenital thoracic kidneys are as follows: excessive renal ascent, congenital diaphragmatic hernia, pulmonary hypoplasia, and congenital neuropathies affecting the diaphragm. It is considered that folic acid or vitamin-A deficiency during the intrauterine period, as well as exposure to teratogenic medicines, chemical substances, ionized rays, infections, or infestations, such as *Schistosoma haematobium* or malaria during the intrauterine period, play a role in the etiopathogenesis of congenital thoracic kidney [4]. Various mechanisms have been thought to be responsible for congenital

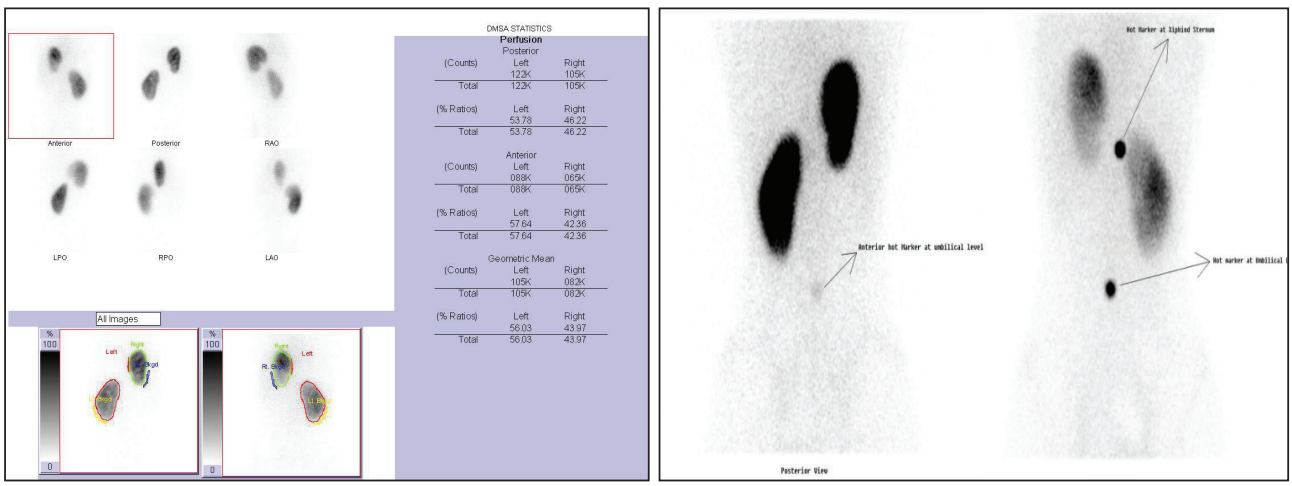

***Figure 1**. (a) Renal DMSA scan with multiple views showing the location and relative function of both kidneys. (b) Renal DMSA scan showing right thoracic kidney, anterior view, and radioactive marker at umbilical and Xiphoid-sternum level.*

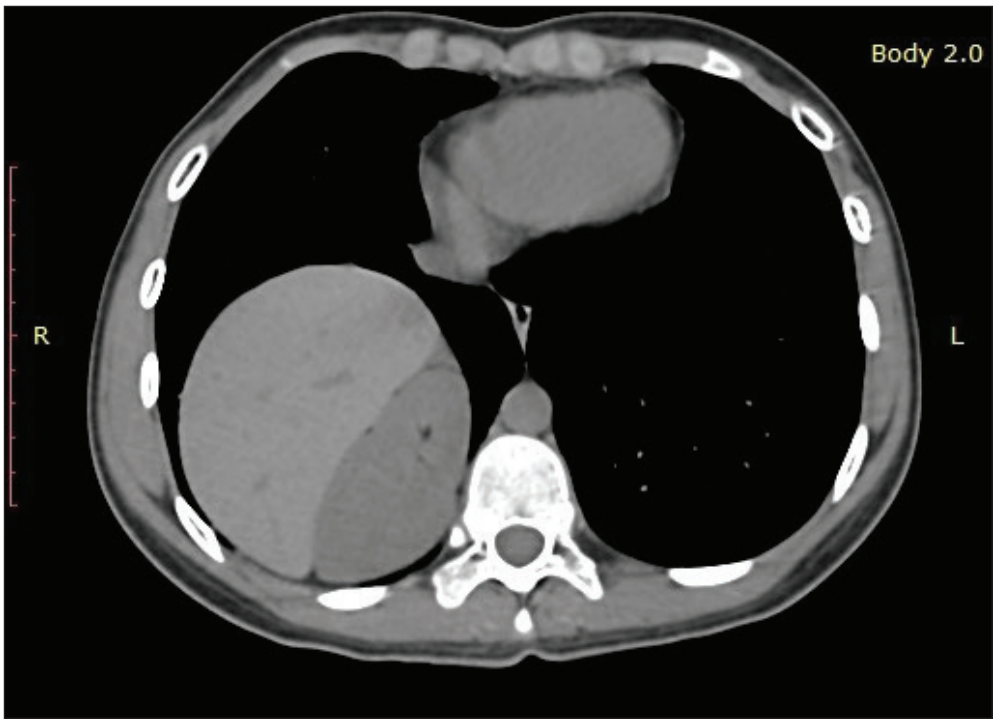

***Figure 2**. CT cross-sectional view showing right thoracic kidney, right lobe of liver, and left lung.*

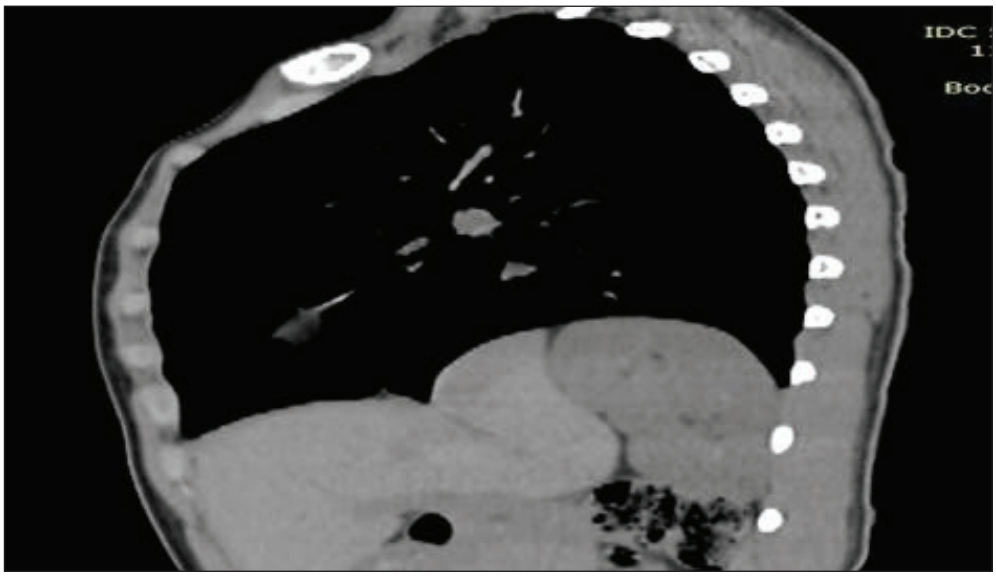

***Figure 3**. CT sagittal view showing right thoracic kidney and eventration of the right hemidiaphragm.*

intrathoracic kidneys, such as accelerated ascent of the kidney, delayed closure or maldevelopment of the pleuroperitoneal membrane (Bochdalek hernia), the effect of the developing liver and adrenal glands, and the persistence of the nephrogenic cord [4-6]. Today, a more widely accepted mechanism for this condition is that the developmental phases of adrenal gland and liver alter the posture of the kidney, and the kidney develops secondary to the cord. Nevertheless, none of these mechanisms is completely ruled out, and it is reported that each can take part in the development of an ectopic kidney [5]. It has been reported that an ectopic kidney could also be found with multiple system anomalies in this case, as it was diaphragmatic eventration. The relationship of the intra-thoracic kidney with diaphragm may vary from case to case. This case was accompanied by the eventration of diaphragm. It is caused by insufficiency of muscle fibers due to paralysis, aplasia, or atrophy in the prenatal period, and it is a very rare condition. As in this case, diaphragm eventration is usually asymptomatic. Mostly, it does not require a specific treatment. In severe cases with symptoms, plication surgery generally relieves the condition. The features of

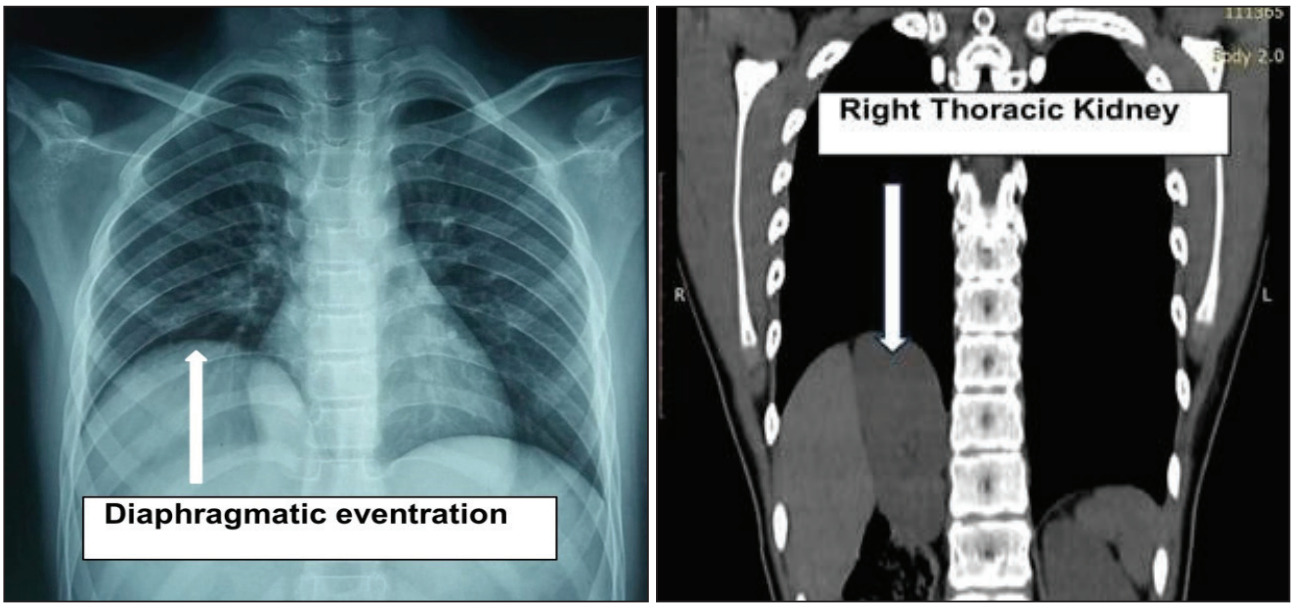

***Figure 4**. Chest X-ray showing right-sided diaphragmatic eventration, and coronal CT scan images showing right thoracic kidney.*

congenital thoracic kidney include rotational anomaly, along the ureter and anomalous high derivation of the renal vessel, and medial deviation of the lower pole of the kidney. In most cases, the adrenal gland occupies its normal position within the abdomen [5-8].

Acquired thoracic kidneys are also rare and may occur secondary to trauma, neurological disease, or space occupying lesion in abdomen. Most commonly, acquired thoracic kidneys are seen secondary to diaphragmatic dysfunction. Any disease that interferes with diaphragmatic innervation, contractile muscle function, or mechanical coupling to the chest wall can cause the diaphragmatic dysfunction. Post-traumatic rupture of diaphragm and diaphragmatic eventration due to a large space occupying lesion in the abdomen, i.e., renal tumor, sub-phrenic abscess, liver abscess, Banti's disease are other common causes of acquired thoracic kidneys [7,9,10]. Thoracic kidney is one of the important differential diagnoses for posterior mediastinal mass and in the evaluation of a non-visualized kidney in the renal fossa.

Several methods have been used to diagnose an intrathoracic kidney. Plain radiographs are often indeterminate and may confuse this condition with other posterior mediastinal lesions such as Bochdalek hernia, pulmonary sequestration, or neurogenic masses. CT and ultrasonography have surpassed IV urography as the modality of choice. Functional imaging using Tc-99m-DMSA and Tc-99m-DTPA scintigraphy still plays a very important role in differentiating an ectopic thoracic kidney from other tissues. Renal scintigraphy must be performed even if CT and IV pyelogram results are normal because it depicts the kidney function more accurately and obviates the need for unnecessary investigation [1,2,5,7,9].

Management of the thoracic kidney depends on the underlying etiology and associated medical condition. Thoracic kidneys can be associated with various other congenital anomalies, such as congenital heart disease and vertebral and genitourinary anomalies. It can be associated with certain acquired medical conditions, such as trauma, surgery, and cancer [8,9,11].

It can also be associated with various clinical manifestations, such as chest pain, dyspnea, cough, and hemoptysis, depending on its location and relation to the surrounding structures, and with urinary tract abnormalities such as vesicoureteral reflux, ureteropelvic junction obstruction, or ureterovesical junction obstruction. In most cases, such as in this case report, the thoracic kidney may be asymptomatic [6,9]. In asymptomatic patients with a functional thoracic kidney as in this case, observation is recommended. In patients with associated diaphragmatic, urinary tract abnormalities, customized surgical correction may be required. In certain associated conditions such as respiratory difficulty, cancer, medical management along with surgical intervention may be necessary [7].

Conclusion

Thoracic kidney is an extremely rare finding, and it can sometimes be a diagnostic challenge for both clinicians and radiologists. In such a scenario, DMSA renal scintigraphy is a very useful imaging modality and usually the first line of investigation in a case of a non-visualized kidney on ultrasonography. Although CT is a preferable imaging modality for accurate diagnosis of the condition, but DMSA renal scintigraphy is still a must-do investigation as it can be used to differentiate an ectopic thoracic kidney from other tissues and also depicts the kidney

function more accurately. Accurate diagnosis of this condition and awareness of all possible underlying etiologies can obviate patient anxiety, unnecessary investigations, and the need for surgical procedures.

List of Abbreviations

CT Computed tomography
CTKUB Computed tomography of kidney, ureter, and bladder.
DMSA Dimercaptosuccinic acid scintigraphy
DTPA Diethylenetriamine penta-acetic acid
MRI Magnetic resonance imaging
Tc-99m Technetium-99 meta-stable state

Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this article.

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Ethical approval

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