CASE REPORT

Spontaneous non-obstructive nephropleural fistula with an autoimmune disorder causing massive urinothorax: a rare association

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SUMMARY

Urinothorax, an unusual and rare cause of pleural effusion, is usually secondary to urinary obstruction and abdominal trauma. We describe an uncommon case of left-sided urinothorax in a 35-year-old man with diabetes and hypothyroidism associated with an autoimmune disorder without obvious obstructive uropathy. Workup revealed pancytopenia, mild proteinuria, positive anti-nuclear and anti-dsDNA antibodies suggestive of probable systemic lupus erythematosus. Contrast-enhanced CT-chest and abdomen showed hepatosplenomegaly with bilateral renal abscesses and a fistulous connection between left superior calyx and left the pleural cavity. Patient was initially managed by intravenous antibiotics, intercostal tube drainage and ipsilateral double-J stent placement. The definitive management in the form of closure of nephropleural fistula was achieved with sclerotherapy using 0.1% povidone-iodine instillation, while oral steroids were started for the probable autoimmune disorder. To the best of our knowledge, this is the first case of spontaneous non-obstructive nephropleural fistula associated with an autoimmune disorder, managed by minimally invasive methods.

BACKGROUND

Urinothorax is a rare clinical entity that is characterised by the presence of urine in pleural space secondary to the nephropleural fistula. The most common occurrence is associated with urosurgical procedures in patients of obstructive uropathy and following renal trauma.¹⁻⁴ Patients usually have minimal to moderate pleural collection and may rarely present with severe respiratory distress. Pleural fluid to serum creatinine ratio of greater than 1 is considered as a gold standard for diagnosis.⁴⁻⁶ To date, there are less than 80 cases reported in the medical literature.⁴ Such unusual, non-iatrogenic urinothorax without any urinary tract obstruction has not been described before.

CASE PRESENTATION

A 35-year-old man with diabetes and hypothyroidism presented with left flank pain of 15 days duration associated with fever and dyspnoea for 2 days. Evaluation at community health centre with ultrasound abdomen and CT-thorax revealed left-sided moderate loculated pleural collection with underlying collapsed lung and mild mediastinal

shift towards the right side with the left perinephric collection ([figure 1A,B](#)). The patient underwent left intercostal tube drainage (ICTD) for empyema and 12-Fr pigtail tube insertion in the left perinephric collection. The pigtail initially drained about 100 mL/24 hours, which progressively decreased. The pigtail was withdrawn after 7 days. The patient presented to us with a complaint of urine leakage through the left flank puncture site (nephro-cutaneous fistula) and intercostal tube draining straw coloured fluid ([figure 2B](#)). Examination revealed multiple petechial haemorrhagic patches ([figure 2A](#)) over the chest wall with urostomy bag applied over the nephro-cutaneous fistula ([figure 2B](#)). Clinically, the patient was pale without jaundice and no lymphadenopathy.

INVESTIGATIONS

Blood investigations showed normocytic normochromic anaemia (haemoglobin—7 g/dL), leucopenia (3200/ μ L), thrombocytopenia (20 000/ μ L) and elevated C reactive protein levels (214 mg/L). Other reports including renal function tests (S.Creatinine—1.0 mg/dL) and blood glucose level (105 mg/dL) were unremarkable. Urine examination revealed mild proteinuria (600 mg/24 hours) and urinary sediment analysis showed no alteration. Blood and urine cultures were analysed and returned negative. Bone marrow biopsy was indicative of secondary myelosuppression. In consultation with a physician, the patient was given 4 units of platelet transfusion and 2 units of packed red blood cell transfusion which improved the platelet count to 60 000/ μ L and haemoglobin level became 8.5 g/dL. Connective tissue disease workup revealed positive antinuclear antibodies (ANA) (1:640) and positive anti-dsDNA antibodies (360 units). Serum complement factor levels were equivocal (C3=84 (88–252 mg/dL), C4=20 (12–72 mg/dL)). X-ray chest showed the ground-glass appearance of left haemithorax suggestive of pleural effusion ([figure 1C](#)). Contrast-enhanced CT (CECT)-abdomen revealed moderate hepatosplenomegaly with delayed films showing contrast leakage from left superior calyx with seepage into the left perinephric space that was communicating with left pleural space. Both kidneys had multiple renal abscesses (right side at mid pole and left side at the inferior pole) with no calculi ([figure 3A-D](#)). Pleural fluid examination revealed low pH (6.9), low sugar

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Figure 1 CT-thorax revealed left-sided moderate loculated pleural collection with underlying collapsed lung and mild mediastinal shift towards the right side (A,B). Chest X-ray showing pleural effusion with a blunted costophrenic angle (C) and repeat normal chest X-ray at 3-month follow-up (D).

(9 mg/dL), protein (0.3 g/dL) and 550 white blood cells/mm³ with high pleural fluid creatinine to serum creatinine ratio (8.6:1), thus confirming the diagnosis of urinothorax. The pleural fluid culture showed insignificant growth of *Escherichia coli*. Examination of this fluid for tuberculosis was negative.

DIFFERENTIAL DIAGNOSIS

A diagnosis of nephro-pleurocutaneous fistula along with probable autoimmune disorder (systemic lupus erythematosus (SLE)) was made.

Figure 2 On presentation, multiple petechial haemorrhages noted over the anterior chest wall (A) with urostomy bag applied over the nephro-cutaneous fistula and the thoracostomy tube in situ (B). Healed nephro-cutaneous fistula (black arrow) and thoracostomy site (white arrow), posturinary diversion (Double-J stenting) and retrograde intracalyceal sclerotherapy (C).

Obstructive uropathy was excluded as both the ultrasound abdomen and CECT abdomen showed no hydronephrosis.

In the presence of myelosuppression and multiple renal abscesses with hepatosplenomegaly, generalised septicæmia was a probable differential diagnosis. Another rare possible mechanism for nephropleural fistula could have been a simultaneous infection of multiple bilateral primary renal cysts getting converted into renal abscesses.

The patient met both the 1997 Modified American College of Rheumatology (ACR) criteria and 2012 Systemic Lupus International Collaborating Clinics (SLICC) criteria for the diagnosis of SLE (fulfilling four or more criteria—at least one clinical and one immunological): the presence of leucopenia, thrombocytopenia, mild proteinuria (>500 mg/24 hours), serositis (pleural effusion) and positive ANA and anti-dsDNA antibodies.⁷

TREATMENT

The patient was started on intravenous antibiotics according to the local hospital guidelines. Under local anaesthesia and antibiotic cover, retrograde pyelogram on the left side revealed leakage of contrast into perinephric space (figure 4) for which a 5 Fr/26 cm double-J stent was placed. Ultrasound-guided aspiration of bilateral renal abscesses was performed. Following this, nephro-cutaneous fistula (at previous pigtail insertion site) became dry and healed completely in 4 days. The patient was started on oral prednisolone (1 mg/kg/day) on the advice of an immunologist, which was gradually tapered over a period of 40 days. After 20 days of double-J stent placement, the daily intercostal tube output remained around 200 mL. Repeat haemogram showed haemoglobin—9 g/dL, white blood cells—6000/μL and platelet—130 000/μL.

The patient was taken up for repeat retrograde pyelographic study that revealed the persistence of communication between the superior calyx and pleura. Based on our experience of treating chyluria with 0.2% povidone-iodine as a sclerosant,⁸ we removed the double-J stent and placed a ureteric catheter in the superior calyx under fluoroscopic control. The patient was subjected to sclerotherapy with intracalyceal 0.1% povidone-iodine instillation through a ureteric catheter-placed retrograde. After completion of three doses of sclerotherapy, the intercostal drain output decreased and was <20 mL/24 hours after 7 days. Internal urinary diversion was done with a Double-J stent. After an observation of about 7 days, left intercostal tube was removed without any adverse effects.

OUTCOME AND FOLLOW-UP

At follow-up visit of 3 months, the patient was doing well and left Double-J stent was removed. Repeat chest X-ray was normal (figure 1D). The patient is due for next follow-up at 6 months.

DISCUSSION

Nephropleural fistula, an abnormal communication between the pelvicalyceal system and pleural cavity causing urinothorax, is a rare clinical entity. Usual causes include complications of urological procedures (percutaneous endoscopic renal procedures, perforation, extracorporeal shock wave lithotripsy), blunt abdominal trauma, malignancy, retroperitoneal inflammation and polycystic renal disorder.^{4 5 8 9} The fluid collected in the pleural space originates from the retroperitoneal collection of urine (retroperitoneal urinoma), following extravasation from the pelvicalyceal system. The possible explanations for the extension of the retroperitoneal collection into the thoracic cavity include either through the

Figure 3 Contrast-enhanced CT abdomen and pelvis, coronal view revealed bilateral renal abscesses with contrast leakage from the left superior calyx (arrow) into the perinephric space (A), axial view showing contrast reaching the posterior flank surface (arrow) through nephro-cutaneous fistula (B), sagittal view showing fistulous connection (multiple arrowheads) between perinephric collection and pleural cavity (C), reconstructed image showing contrast extravasation into perinephric space and extending into the thorax with thoracostomy tube in situ (D).

lymphatic drainage or by dissection through the retroperitoneum at the site where Gerota's fascia is attached to the diaphragm and eventually rupturing into pleural space.^{4 10} Lee *et al* have reported a case of urinothorax associated with VURD syndrome (posterior urethral Valves, Unilateral vesicoureteral Reflux and renal Dysplasia) in an infant.¹¹

In patients of urinothorax, the pleural fluid is mostly transudative in nature with low cell count, low sugar and low protein content. The ratio of fluid creatinine to serum creatinine greater than 1 has been considered as a hallmark of biochemical tests for its diagnosis.⁴⁻⁶ Diagnostic imaging in the form of a chest radiograph, abdominal and thoracic CT with delayed scanning, retrograde pyelography and radionuclide renal scans are useful

in locating the retroperitoneal urinoma and anatomical location of fistula and thus confirming the diagnosis.

Our case of nephro-pleurocutaneous fistula was complicated with associated comorbidities and autoimmune disorder causing secondary myelosuppression. This myelosuppression may be the limiting factor for generalised sepsis causing hepatosplenomegaly and bilateral renal abscesses. We suspect that the renal abscess involving the upper pole of the left kidney might have ruptured into the perinephric space, subsequently finding its way into the left pleural cavity through retroperitoneum causing a large pleural effusion.

In the majority of cases, urinothorax resolves spontaneously, once the underlying cause of obstructive uropathy is corrected. However, in cases of persistent pleural effusion, thoracentesis or thoracostomy and internal urinary diversion (double-J stent) for pelvicalyceal decompression are usually successful. Surgical correction including pleural sclerosis, decortication and nephrectomy may seem necessary for persistent ICTD output or fever despite pelvicalyceal and thoracic decompression.^{4 12 13}

In our case, the nephro-cutaneous fistula resolved with internal urinary diversion but nephropleural fistula persisted with significant ICTD output. Taking inspiration from the fact that povidone-iodine has been used as a sclerosant in chyluria cases for the closure of pyelolymphatic fistula, we used the same for nephropleural fistula.⁸ As our case was complicated by other comorbidities and associated autoimmune disease, we have prophylactically reduced the dose of povidone-iodine to half (0.1%). As povidone-iodine as a sclerosant in reno-pleural fistulas has not been studied in the past, its efficacy and long-term outcome are still unknown. Longer follow-up of our patient is needed for evaluating its outcome. Moreover, for defining the nature and severity of renal involvement in a case of probable SLE, renal biopsy was warranted in our case but was not possible due to the presence of perinephric infection. The patient is due for renal biopsy at his next follow-up at 6 months.

Figure 4 Retrograde pyelography showing contrast extravasation (arrow) from the superior calyx into the perinephric space and extending superiorly into the thoracic cavity.

To the best of knowledge, such unusual case of spontaneous urinothorax without any underlying urinary obstruction has not been reported before.

Learning points

- ▶ Even though urinothorax is usually associated with obstructive uropathy, a high degree of suspicion should be kept in patients of pleural effusion with retroperitoneal urinoma without any obstruction or antecedent surgical intervention.
- ▶ Diagnostic modalities like chest X-ray, Contrast-enhanced CT abdomen with thorax and retrograde pyelography are helpful in defining the fistulous tract and making the diagnosis of urinothorax.
- ▶ Definitive management includes multidisciplinary approach and should also consider patients' clinical status and comorbid factors.
- ▶ Thoracostomy and urinary diversion remain the cornerstone in the management of urinothorax. However, in reluctant cases, retrograde sclerotherapy using 0.1% povidone-iodine may be considered as a minimally invasive modality of treatment. Short-term results in our patient are good.

Contributors All of the authors declare that they have all participated in the design, execution and analysis of the paper, and that they have approved the final version. Individual author contribution is as follows: RA—Concept, design, supervision, processing, writing manuscript and critical analysis. SG—Concept, supervision, processing, writing manuscript and critical analysis. AG—Supervision, processing, writing manuscript and critical analysis. VK—Supervision, writing manuscript and critical analysis.

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