Case Report

Pulmonary lymphangitis carcinomatosa in a patient with carcinoma of urinary bladder

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ABSTRACT

Pulmonary lymphangitis carcinomatosa occurs due to dissemination of cancer cells in pulmonary lymphatics. It is only rarely seen with urological malignancies and portends a grave prognosis. Its presence in patients with urothelial carcinoma of the urinary bladder is uncommon. We present a case of a nested variant of transitional cell carcinoma of the urinary bladder associated with pulmonary lymphangitis carcinomatosa. We also discuss the symptoms and radiological features that might aid in timely diagnosis of this entity.

INTRODUCTION

Pulmonary lymphangitis carcinomatosa is a rapidly progressive pathology, most commonly seen in patients with a known history of adenocarcinoma.[1] It is rarely seen in patients with urothelial carcinoma.[2,3] We describe a case in which pulmonary lymphangitis carcinomatosa was associated with bladder carcinoma.

CASE REPORT

A 57-year-old male had a history of painless hematuria for 2 months. He was evaluated at an outside facility and was found to have a 2 cm ×2 cm bladder tumor on an ultrasound. Transurethral resection of the tumor was done, and the histopathology was suggestive of a nested variant of transitional cell carcinoma (TCC). He was referred to us for further management after 4 weeks of primary surgery. He had a history of recent-onset lower-limb weakness and breathing difficulty with nonproductive cough. On examination, he was pale and had fever (99.5°F), tachycardia, tachypnea, and motor weakness of bilateral lower limbs. His oxygen saturation on room air was 82% which improved to 95% with 6 L/min of oxygen support. The prostate was normal on digital rectal examination with decreased rectal tone. There was suprapubic fullness despite a 22-Fr 3-way Foley catheter *in situ* since the time of surgery. A bladder wash was given, and nearly 200 ml of old clots were removed. The hemoglobin was 7.5 g/dl and total leukocyte count was 14,500 with neutrophilia. He had a creatinine of 1.1 mg/dl, low albumin (2.5 g/dl), and elevated alkaline phosphatase (300 IU/L), while the prostate-specific antigen was 4.2 ng/ml. Urine examination showed plenty of pus cells, red blood cells, and leukocyte esterase positivity, and urine culture was positive for *Pseudomonas aeruginosa*. The contrast-enhanced computed tomography (CT) abdomen revealed diffuse thickening of the bladder wall, a diverticulum adjacent to the right vesicoureteric junction, and air in the lumen (as a result of bladder wash). There was bilateral external iliac and para-aortic lymphadenopathy. There was metastatic involvement of the sacrum, right acetabulum, bilateral iliac bones, fifth lumbar vertebra, fourth and ninth thoracic vertebrae, and eighth costovertebral angle [Figure 1a and b]. Chest CT revealed thickened, nodular interlobular septa and polygonal arcades in bilateral upper lobes and right middle lobe suggestive of pulmonary lymphangitis carcinomatosa [Figure 2a and b].

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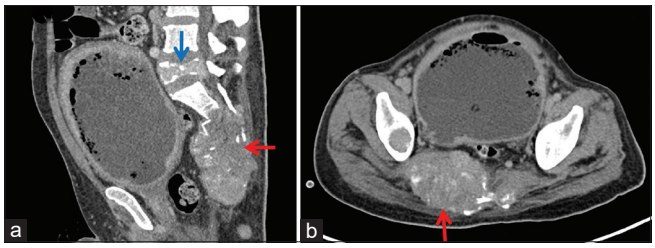

Figure 1: (a) Sagittal and (b) transverse sections of computed tomography abdomen showing the diffusely thickened bladder with a right-sided diverticulum and the metastatic deposits in the sacrum along with the right iliac bone (red arrow) and fifth lumbar vertebra (blue arrow)

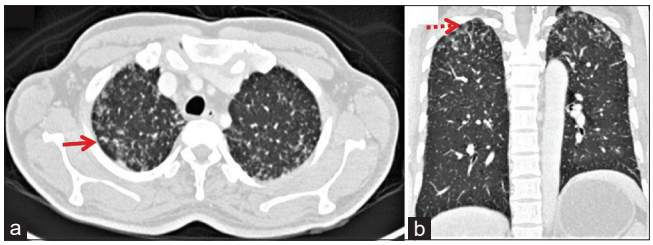

Figure 2: The features of pulmonary lymphangitis carcinomatosa on (a) axial sections of computed tomography chest showing the thickened and nodular interlobular septa (red arrow) without alteration of parenchymal architecture and (b) coronal image showing the characteristic polygons (red broken arrow) and involvement of bilateral upper lobes

There was no significant mediastinal/hilar lymphadenopathy or pleural effusion. The patient did not wish for any active treatment, and hence, only palliative pain care was offered. He succumbed to his disease after 1 month.

DISCUSSION

Pulmonary lymphangitis carcinomatosa occurs due to the dissemination of cancer to the pulmonary lymphatics leading to obstruction of these channels. The condition is often seen in patients with breast, gastrointestinal, prostate, and lung cancers.[1] The blocked lymphatic vessels cause outflow obstruction of the lymph from the pulmonary interstitium leading to a mismatch of ventilation and perfusion. The most common symptoms are dyspnea, dry cough, and hemoptysis in the background of a known malignancy. Clinical differentials include congestive cardiac failure, pulmonary embolism, and pneumonia. The symptoms precede radiological findings and are rapidly progressive and often out of proportion of the findings.[1,4,5] Chest X-rays may show nonspecific findings such as reticular opacities, Kerley A and B lines, and pleural effusion but also can be normal in more than half of the patients. CT scan of the chest is the main diagnostic modality. The classical findings include interlobular septal thickening which can be nodular or smooth, peribronchovascular thickening, and polygonal arcades. The changes can be patchy or diffuse and may involve one or both the lungs with a predilection to involve lower lobes. The parenchymal architecture of the secondary pulmonary lobule is maintained which, along with the presence of polygons, helps in differentiating it from interstitial lung disease and pulmonary fibrosis.[1,5,6] A transbronchial lung biopsy is confirmatory but often not possible due to the clinical condition of the patients. The history of a preexisting malignancy with the aforementioned findings on imaging substantiates the diagnosis of pulmonary lymphangitis carcinomatosa.[1]

TCC of the bladder is a common malignancy with a tendency to metastasize to lymph nodes, followed by hematogenous spread to bones, lungs, and liver. The pulmonary metastasis due to bladder cancer is usually solid, discreet nodular but can rarely be cavitary or endobronchial.[7] The association of bladder cancer and lymphangitis carcinomatosa is rare, and to the best of our knowledge, only two such cases have been reported in the English literature.[2,3] The nested variant of the TCC is an uncommon but aggressive tumor with a tendency to cause early metastasis.[8] The association of nested variant of bladder TCC with pulmonary lymphangitis carcinomatosa has not been previously reported in the literature. The presence of pulmonary lymphangitis carcinomatosa indicates rapid progression and poor prognosis. It has been reported that majority of the patients die within 2 months of being symptomatic or 3 weeks following hospitalization.[1,4] The treatment consists of glucocorticoids, oxygen supplementation, and opioids for symptomatic relief. Chemotherapy, targeted against the primary tumor, may be helpful, but these patients are often too sick to be administered this treatment.[9]

CONCLUSION

We describe a rare case of nested variant of TCC of the urinary bladder associated with pulmonary lymphangitis carcinomatosa with a fatal outcome. The clinicians should be aware of this condition and should have a high index of suspicion, especially in those patients of bladder cancer who present with progressive respiratory distress which is out of proportion to the radiological findings.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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