A rare renal malakoplakia mimicking renal tumor presenting with deep vein thrombosis and bilateral pulmonary embolism in a 58-year-old woman

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ABSTRACT

Malakoplakia is an uncommon chronic inflammatory disease that appears as soft plaques in various organs and results from defective macrophage function, which tends to affect immunocompromised and debilitated patients. The pseudotumoral form presentation is rare especially with para-neoplastic syndrome. Preoperative diagnosis of renal malakoplakia in appropriate clinical settings can prevent unnecessary surgery. We present a clinical case of renal malakoplakia in a 58 old woman mimicking a malignant locally advanced renal carcinoma with rare presentation of bilateral pulmonary embolism.

Introduction

The term malakoplakia is derived from the Greek words malakos, which means soft, and plakos, which means plaque. It was first described in 1901 by von Hansemann and then were first published by Michaelis and Gutmann in 1902. Malakoplakia has a predilection for the genitourinary tract, but has been known to affect the gastrointestinal tract, bone, lungs, lymph nodes and skin, with a female predominance 4:1. Patients with malakoplakia usually present with fever, flank and/or loin pain, abdominal mass and a history of E. coli urinary tract infection. It can rarely present as renal vein thrombosis. We present a rare clinical case of renal malakoplakia mimicking a malignant renal carcinoma presenting with deep vein thrombosis and bilateral pulmonary embolism in a 58-year-old woman. This case can reinforces the idea that unusual disease entities should be explored to aid in achieving a correct diagnosis and, thus, potentially avoid unnecessary treatment.

Case presentation

A 58-year-old woman presented with a 4 week history of worsening left-sided thoracique and flank abdominal pain, loss of weight and dyspnea. She was admitted through the emergency department with suspicion of pulmonary embolism (PE) which was confirmed by CT angiography (Fig. 1: left side). It also show accidently left renal mass of 9 cm. Doppler ultrasound was also done and show deep vein thrombosis and she has been seen by our hematologist with no hematological pre-disposition to her thrombosis other than Para-neoplastic syndrome related to left renal mass. Two month after anticoagulant treatment and owing to the appearance of the mass on imaging, and after repeated CT scan (Fig. 1: right side), the patient underwent left radical nephrectomy, which is generally indicated for solid renal masses.

The histopathological macroscopic examination had revealed that the lesion extended to the renal capsule and perinephric fat. The immunohistochemical stainings showed strong reactivity for CD 68 (Fig. 2: right side), and the histological examination revealed diffuse cellular infiltration, with eosinophilic macrophages (von Hansemann cells) (Fig. 2: left side) and sheets of Periodic Acid Schiff-positive histiocytes into the renal parenchyma. These cells had granular acidophilic cytoplasm and round concentric layered intracytoplasmic Michaelis-Gutmann bodies (Fig. 3).

The final diagnosis was renal parenchymal malakoplakia. During her first follow-up appointment with urology 1 month later, her respiratory and abdominal symptoms had resolved, and BUN and creatinine were within normal range. Her follow up Doppler ultrasound and CT angiography show disappear of DVT and she advice to stop anticoagulant (Apixaban) after 6 months of treatment. During her most recent visit to urology 1 year later, she was again asymptomatic, and has
not had any further clinical manifestations of malakoplakia in any organ system nor has she presented with any septic episodes or UTI since the nephrectomy.

Discussion

Malakoplakia is an inflammatory condition associated with immunosuppression, infection and systemic illness. Gram negative bacterial infection tends to be associated with malakoplakia and *Escherichia coli* infection tends to be encountered in two thirds of cases of malakoplakia. Kobayashi et al. documented that by using polymerase chain reaction (PCR), they had been able to site-specifically amplify *Escherichia* DNA from malakoplakia kidney biopsy.

Malakoplakia may occur at any age, but most commonly presents in the fifth decade of life. Appleson et al. reported a 13 months baby with renal malakoplakia.

Our patient manifests with pulmonary embolism symptoms which first to be reported in English literature. The manifestation of renal malakoplakia is non-specific including fever, lethargy, loin pain, hematuria and loss of weight plus other non-specific symptoms. Megson et al. reported malakoplakia can presented as complication post radical nephrectomy for malignancy.

The pathogenesis of malakoplakia remains unknown, yet it is generally considered that the lesion is associated with a defect in the ability of macrophages to digest phagocytosed bacteria. This lead to decrease levels of intracellular cyclic guanosine monophosphate and decreased release of β glucuronidase. It is the calcification of these incompletely digested organisms that are thought to produce the Michaelis-Gutmann bodies.

The radiology imaging appearances tend to be non-specific (normal,
mimicking abscess or renal carcinoma or other chronic inflammations of kidney). Parenchymal calcification is rare. A unifocal renal lesion is uncommon and can resemble a necrotic renal cell carcinoma.

The diagnosis of malakoplakia must be kept in mind especially in patient with renal mass and a history of long-term recurrent renal infections or renal failure, immunosuppression, and systemic illness. Fine needle aspiration (FNA) biopsy should be one of the diagnostic options in high suspected patient.

The treatment of malakoplakia depends on the extent of the disease and underlying conditions of the patient. Typically, patients with bilateral or multifocal diseases are cured after using antibiotics, including for example quinolones, rifampin, doxycycline and vancomycin. In addition, a cholinergic agonist, such as bethanechol chloride, is used in combination, with antibiotics to correct lysosomal defects. Surgical excision is the choice treatment of unifocal disease, and nephrectomy is indicated when damage to the kidney is extensive.

In conclusion, the diagnosis of malakoplakia must be kept in mind for patients presenting with classical symptoms and history and should be evaluated for possible malakoplakia with imaging studies plus needle aspiration or biopsy. In order to achieving the correct diagnosis and, potentially avoid delayed or unnecessary treatment; we should increase the awareness of this disease.

Consent

Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

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