Incidentally found focal xanthogranulomatous pyelonephritis with extensive venous thrombus

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A 71-year-old woman with history of asthma presented with 2 months history of shortness of breath; on imaging an incidental left renal mass was noted. Subsequent renal protocol CT was obtained that showed a 4.5 cm left upper pole exophytic mass with renal vein thrombus extending into the inferior vena cava to the level of the caudate lobe concerning for renal cell carcinoma. She underwent an open left radical nephrectomy and IVC thrombectomy with subsequent postoperative pathology demonstrating xanthogranulomatous pyelonephritis without renal cell carcinoma.

Key Words: inferior vena cava thrombus, focal xanthogranulomatous pyelonephritis

Introduction

Xanthogranulomatous pyelonephritis (XGP) is a rare, severe and atypical form of chronic pyelonephritis, most commonly in the setting of infected, obstructing renal nephrolithiasis. Patients frequently experience flank pain, fever, malaise, anorexia and weight loss. Lab findings may include anemia, elevated inflammatory markers, pyuria and/or bacteriuria. The most commonly involved organisms are Escherichia coli and Proteus mirabilis but other typical urinary pathogens are also seen.[1] It is best characterized on imaging by CT scan, often with a large, nonfunctioning kidney with multiple low-density areas surrounded by contrast enhancing rims. A well-described radiological finding is known as the "bear paw sign". Frequently nephrolithiasis is found. It can be mistaken as renal cell carcinoma (RCC), especially in the 10% of cases

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where it is a focal finding within a functional kidney.[2] Pathology demonstrates an inflammatory destruction of the kidney with necrotic yellow material and frequently nephrolithiasis. On microscopic review sheets and groups of foamy histiocytes forming xanthogranulomas are seen with associated coarse calcifications and extensive necrosis.[1]

Case report

A 71-year-old woman with history of asthma, gastroesophageal reflux disease, and depression presented with 2 months of shortness of breath and cough. Work up in the emergency department involved a pulmonary embolism (PE) protocol CT angiogram that showed no evidence of PE but did incidentally note a left renal mass. In the visualized portion of the IVC there was no thrombus. Her baseline creatinine was 1.05 at that time. Subsequent renal protocol CT was obtained that showed a 4.5 cm left upper pole exophytic mass and infrahepatic IVC tumor thrombus that also invaded the left gonadal vein, consistent with cT3b RCC, Figure 1. The mass included small areas of calcification that did not appear consistent with nephrolithiasis. Preoperative labs were unremarkable, including negative UA. Systemic anticoagulation was deferred preoperatively to lessen the risk of hemorrhage as her IVC thrombus was not occlusive and there were no embolic events. She was taken to the operative room 7 weeks after initial ED presentation for open nephrectomy and IVC thrombectomy without complication. The thrombus was adherent to the IVC so with some difficulty blunt dissection was used to remove it en bloc leaving the vena cava intact. Following the case she was admitted to the surgical ICU for close observation. Postoperatively she was given prophylactic subcutaneous heparin for venous thromboembolism prevention. Other than acute blood loss anemia not requiring transfusion she had an uncomplicated immediate postoperative course and was discharged on postoperative day (POD) 7. She presented to the ED with a small bowel obstruction on POD12, was readmitted and conservatively managed with resolution of obstruction overnight and was again discharged on POD14. The remainder of her postoperative course was uneventful. Pathology demonstrated XGP without renal cell carcinoma, Figure 2. Sheets and groups of foamy histiocytes, confirmed by staining positive for CD68, formed xanthogranulomas with associated course calcifications and extensive necrosis. Aggregates of foamy epithelioid histocytes with some foci of associated necrosis and calcifications formed a 7.5 cm x 3.0 cm thrombus that extended 4.0 cm x 3.0 cm beyond the renal vein into the vena cava on gross specimen, Figure 2c. There was no note of nephrolithiasis present in the specimen but

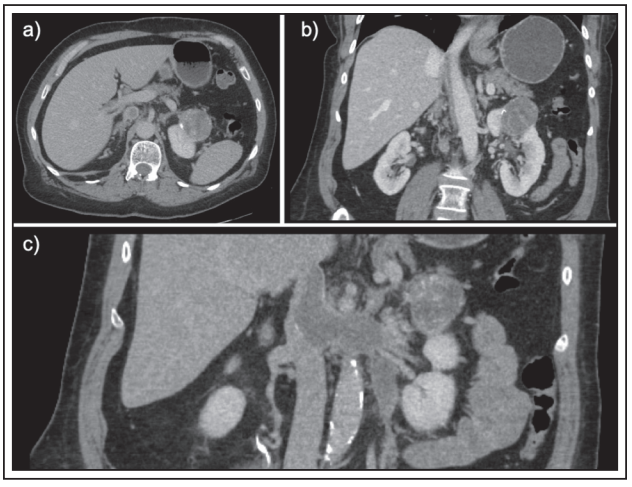

Figure 1. CT renal protocol images demonstrating: **(a, b)** focal heterogenous enhancing left renal mass with **(c)** extensive tumor thrombus extending into the subhepatic IVC and gonadal vein.

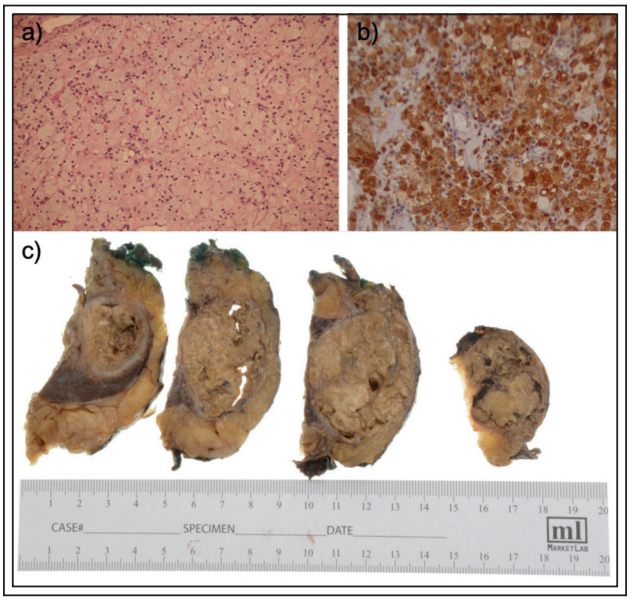

Figure 2. (a) Sheets and groups of foamy histiocytes forming xanthogranulomas with associated coarse calcifications and extensive necrosis. **(b)** Histiocytes stain positive for CD68. **(c)** The xanthogranulomatous process formed a renal vein thrombus that extended 4.0 cm x 3.0 cm beyond the renal vein consisting of foamy epithelioid histiocytes with foci of necrosis.

the mass replaced the superior calyceal system. On postoperative directed questioning she did endorse remote history of two episodes of pyelonephritis, last greater than 10 years prior to presentation. This case was further reviewed in a joint urology/radiology case conference without post-hoc findings that would have indicated a diagnosis of XGP.

Discussion

This case depicts a unique presentation of a focal XGP mass with extensive IVC thrombus in a woman without recurrent urinary tract infections or nephrolithiasis. While it has been well documented that XGP can be mistaken as RCC, it has not been described as a small focal mass with extensive tumor thrombus without concomitant infectious history. A number of rare descriptions of thrombus with XGP have been with a large mass or complete XGP kidneys.[3-5] Additionally, in each of these reports the patient's presentation or history was consistent with urologic infection. There were two case reports of a focal XGP tumor with associated renal vein thrombus found in the literature and they presented differently from our patient: both were found in the setting of active urosepsis.[6,7] A third patient of Rosevear et al was found to have concomitant focal XGP and RCC with IVC thrombus but had no history of urologic infections.[8] The mass showed rapid shrinkage from 7 cm x 5 cm to 4 cm x 3.8 cm without intervention on repeat CT scans prior to nephrectomy and pathology demonstrated a 3.7 cm clear cell RCC in addition to XGP. While somewhat rare, synchronous RCC and XGP has been reported in no less than 16 cases.[9] One theory is the mass may cause focal calyceal obstruction resulting in XGP. The pathologic specimen for Rosevear et al appeared to have the majority of the XGP findings within the thrombus.[8] A unique finding in that case was inability to dissect the thrombus from the IVC, an issue they felt in retrospect to likely be due to the inflammatory nature of the thrombus. They remarked in the future such findings might be an intraoperative clue to the nature of the mass as RCC thrombus is typically described as freely mobile. Similarly we found our patient's inflammatory thrombus to be more difficult to remove than typical. Preoperative identification of bland, or benign, thrombus by imaging would not be a reliable clue as one study using MRI to predict IVC invasion found 16.7% of their patients to have bland thrombus only.[10] With directed questioning during the postoperative visit we discovered a distant history of pyelonephritis, although if discovered preoperatively that alone would not have changed management. This case serves as a reminder of XGP as the "great imitator".

Conclusion

We present the first known case of focal XGP kidney with associated IVC tumor thrombus in an elderly woman without nephrolithiasis, recurrent UTI, or other risk factors for XGP. This highlights the continued difficulty in preoperatively discriminating between RCC and focal XGP. □

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