Implications of Erdheim-Chester disease and percutaneous renal access

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Erdheim-Chester disease (ECD) is a rare systemic histiocytosis with urologic manifestations in a majority of affected patients. An important manifestation is a pronounced retroperitoneal fibrosis with reported dense inflammatory rind surrounding the kidneys. We report a case of a patient with large stone burden necessitating percutaneous nephrolithotomy and the implications related to his Erdheim-Chester-related retroperitoneal fibrotic changes. Foreknowledge of these implications may inform perioperative counseling and surgical planning to maximize opportunity for successful outcomes.

Key Words: nephrolithiasis, retroperitoneal fibrosis, Erdheim-Chester disease, histiocytosis

Introduction

Erdheim-Chester disease (ECD) is a rare disease and part of a larger group of conditions known as systemic histiocytosis or histiocytic neoplasms which includes ECD, Langerhans cell histiocytosis, and Destombes-Rosai-Dorfman disease.[1] There have been an estimated 1000 cases reported globally.[2] This disease most often affects middle-aged men and may involve multiple organ systems, distinguishing this disease from the other histiocytic conditions.[2] The classic urologic manifestation of ECD is retroperitoneal involvement with inflammation and perinephric fat infiltration that may result in retroperitoneal fibrosis and a classic radiographic finding of 'hairy kidneys' on CT scan.[3] This infiltration may extend to the adrenal glands and appear as adrenal masses.[1,2] Although dense perirenal encasement has been described, percutaneous renal surgery in the setting of ECD has not been previously reported. Unique challenges encountered and treatment implications are herein described.

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Case report

A-67 year-old man was referred to our urology clinic with branched, greater than 3 cm calcyceal, renal pelvic,

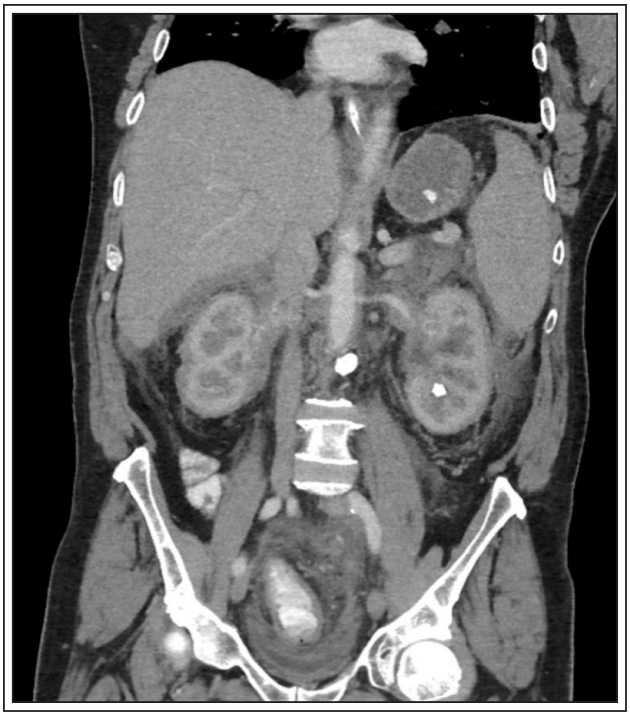

Figure 1. CT abdomen/pelvis from 2012 preceding his diagnosis of ECD. Note the classic radiographic finding of 'hairy' kidneys.

and proximal ureteral stone in an enlarged right kidney without hydronephrosis. A large nonobstructing upper pole contralateral left renal stone was noted, in a similarly enlarged left kidney. He has a history of ulcerative colitis requiring a total colectomy and known ECD.

His ECD was diagnosed 7 years prior during evaluation for abdominal pain. A CT scan was obtained, Figure 1, showing extensive retroperitoneal inflammation and renal encasement with an inflammatory "rind." Subsequent molecular analysis of retroperitoneal biopsy confirmed the diagnosis. Asymptomatic nephrolithiasis was identified, but no intervention had been pursued. Seven years later, he developed Klebsiella urosepsis, and repeat abdominal films revealed increased bilateral renal stone burden, Figure 2, which prompted his referral to our specialty clinic. Based on volume of stone burden, he elected to proceed with right percutaneous nephrolithotomy (PCNL) to address his branched right renal stone.

Multiplanar fluoroscopy was employed to obtain percutaneous renal access into an upper pole calyx via an 18 gauge needle. It was noted immediately that there was dense fibrotic tissue causing increased resistance to needle passage. Safety and working wires were placed without incident. However, tract dilation presented particular challenges. A standard 8Fr/10Fr dilator would not pass through the retroperitoneum to the kidney, assuming an accordion configuration with attempt at passage. We switched to a rigid Amplatz dilator set to serially dilate the tract starting at 6F

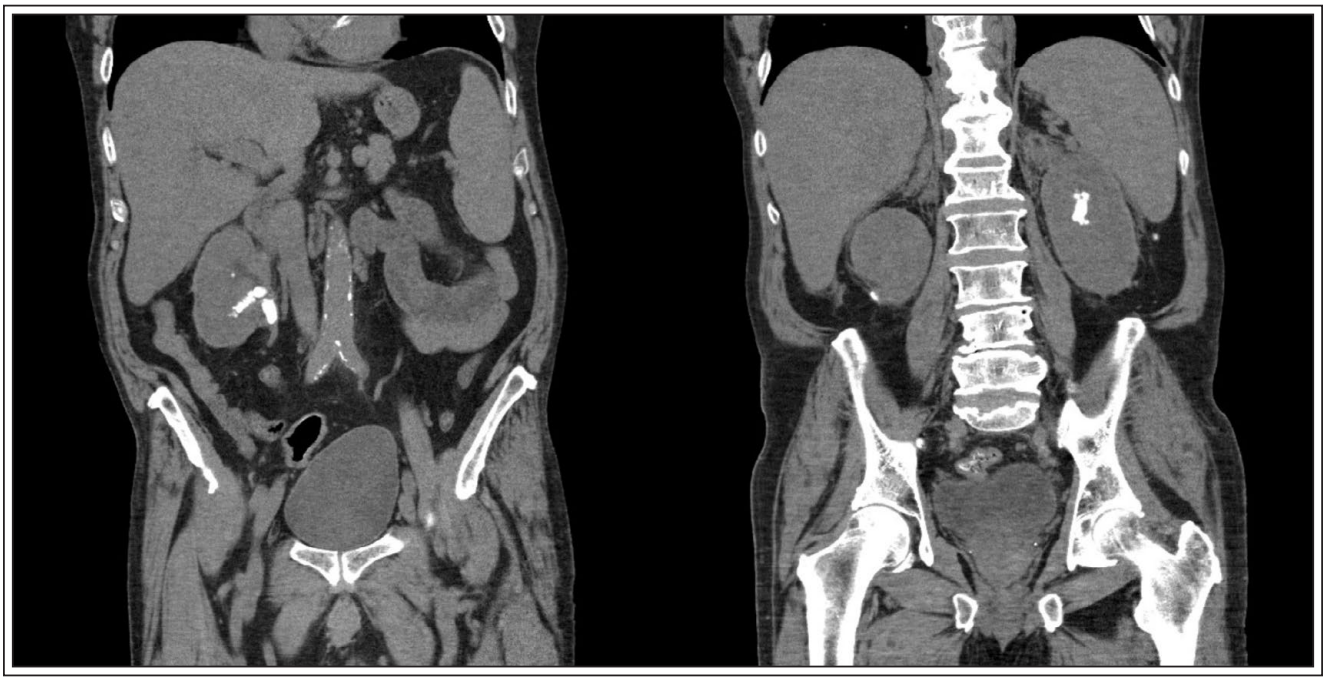

Figure 2. Bilateral partial staghorn calculi.

and working to 10F. We were then able to pass the 8/10 dilator. Balloon dilation with a 30Fr Nephromax (Boston Scientific, Marlboro, MA, USA) was attempted but could not be accomplished due to failure of the balloon to expand to full diameter despite inflation to full recommended inflation pressure. We aborted the balloon dilation and instead chose to continue with the Amplatz dilator set which we used up to 30F. This was technically challenging given the density of the encasing scar tissue, but ultimately successful. Due to a narrow upper pole infundibulum and the fixed, immobility of the kidney reducing navigability, a second access was required. Similar challenges with tract dilation were encountered. At completion, two nephrostomy tubes, a stent, and a foley catheter were placed. He was observed overnight without any complications. Postoperative CT scan the next morning revealed approximately 5 mm of remaining stone fragments and he presented for second look ureteroscopy on the right side 2 weeks later. All residual fragments were removed ureteroscopically. Stone analysis revealed 100% uric acid.

Discussion

Nephrolithiasis has not been previously reported in the setting of ECD. The challenges encountered in the surgical management of stones in this setting prompted this report to inform those who may subsequently encounter this constellation. ECD is a rare inflammatory condition which may manifest retroperitoneal fibrosis. Once diagnosed, median survival is 162 months with a 25% mortality rate for disease related complications.[2] Radiographically, this diagnosis may be suggested by the finding of 'hairy kidneys.'[3]

Prior reports of urologic manifestations of ECD have revealed several key points. In the largest published series to describe these patients, Yelfimov et al report that 79% of ECD patients exhibited urologic involvement with almost half reporting voiding symptoms and a third requiring a urologic procedure.[4] In their case report, Wimpissinger et al describe a patient with progressive renal failure due to bilateral renal compression from retroperitoneal fibrosis. The patient had been medically managed for several years on steroids and other immune-modulators but continued to progress towards renal failure. Given the failed medical management, this group performed open surgical decompression of his right kidney. Over time, the renal parenchyma expanded and the patient avoided dialysis. Similar to our experience, this group described the rind of tissue around the kidney as densely adherent and fibrotic - in their report a 'block of concrete,' such that a subcapsular dissection was required.[5]

Our patient was a gentleman in his mid-60s who had a diagnosis of ECD in addition to ulcerative colitis. He was referred to our specialty clinic for management of bilateral partial staghorn calculi in the setting of retroperitoneal fibrosis due to ECD. The patient required a second look ureteroscopy to complete his stone extraction on the right side and his analysis revealed uric acid stones. This metabolic stone is likely due to his ulcerative colitis and not his ECD and he may be able to be managed medically with urinary alkalinization therapy with avoidance of future percutaneous procedures.

Conclusion

Our case describes to our knowledge the first report of PCNL in the setting of ECD. A diagnosis of ECD or radiologic findings of enlarged, "hairy" kidneys should alert the surgeon to the potential for a dense inflammatory rind surrounding the kidney. Such fibrotic changes may prevent standard balloon dilation as in our case. Thus, those treating stones in patients with diagnosed or suspected ECD should be prepared to employ additional techniques of dilation. Nevertheless, with these precautions, successful stone removal may be accomplished. □

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