Atypical Presentation of Perforated Sigmoid Diverticulitis in a Kidney Transplant Recipient with Autosomal Dominant Polycystic Kidney Disease

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Abstract

Perforated sigmoid diverticulitis, a complication of colonic diverticulosis commonly associated with autosomal dominant polycystic kidney disease (ADPKD), can be life-threatening in allogeneic kidney transplant recipients in the postoperative period. Immunosuppressive medications not only place the patient at risk for intestinal perforation, but also mask classic clinical symptoms and signs of acute abdomen, and subsequently lead to delayed diagnosis and treatment. We report a case of an ADPKD patient post kidney transplantation presenting with nausea, vomiting, and abdominal pain without signs of peritonitis. Chest x-ray revealed free air under the diaphragm consistent with intestinal perforation. Post kidney transplant recipients with ADPKD presenting with abdominal pain should prompt a search for possible perforated colonic diverticulitis in order to diagnose and treat this life-threatening condition early.

Keywords

autosomal dominant polycystic kidney disease (ADPKD), diverticulitis, diverticulosis, kidney transplant

Introduction

Autosomal dominant polycystic kidney disease (ADPKD) is the most common inherited kidney disease, and is a systemic disease presenting with extrarenal manifestations.[1] Diverticulosis is one of the extrarenal manifestations of ADPKD. It can lead to perforated diverticulitis, a life-threatening complication especially in post-kidney transplant recipients under immunosuppression. Generally, classic clinical symptoms and signs of peritonitis can lead to diagnosis; however, post renal transplant recipients with ADPKD may not have these classic clinical manifestations of diverticulosis and perforated diverticulitis. A high index of suspicion for early diagnosis and timely surgical treatment are warranted to decrease morbidity and mortality of this life-threatening condition.

Case Report

The patient is a 46-year-old Caucasian man with a history of ADPKD, hypertension, and gout. He underwent deceased donor kidney transplantation 1 month prior to admission. Operation was uneventful, and he continued immunosuppressive medications including tacrolimus 3 mg twice a day, mycophenolate mofetil 1,000 mg twice a day, and prednisone 5 mg daily. He had several episodes of acute polyarticular gouty attacks which were treated with oral prednisone 30 mg per day tapered off in 1 week. The last attack was 3 weeks prior to admission. Three days prior to admission, he presented with sudden onset of sharp lower abdominal pain, nausea, vomiting, and one loose stool. He also had a low-grade fever, decreased appetite, and constant abdominal pain. His wife also had similar symptoms. He went to see his doctor as a regular appointment. He was afebrile and vital signs were normal. Abdomen was soft with no tenderness, guarding, or rigidity. Because he felt unwell, he was admitted on that day. CBC showed leukocytosis with a white blood cell count of 14,800 /mm^3. Serum creatinine was 1.3 mg/dl (his baseline serum creatinine after kidney transplantation ranged 1.2 to 1.4 mg/dl), and serum electrolytes were normal. Tacrolimus level was 29.9 ng/ml (desired target level range of 8–12 ng/ml). Chest x-ray showed free air under the diaphragm and no pulmonary infiltration (Figure 1). As a result, he underwent emergent exploratory laparotomy. Operative finding showed diverticulitis with small perforation in the sigmoid colon. Partial sigmoidectomy and end colostomy were performed. Tacrolimus was held and it was resumed after the level went down to 5.7 ng/ml (below the target level). Prednisone was also held. Postoperatively, he developed intestinal ileus which was resolved by nasogastric tube suction. He also had acute polyarticular gouty arthritis which responded well to a short course of intravenous methylprednisolone and oral colchicine. He was doing well and was discharged home on postoperative day 11. His renal graft function improved with serum creatinine before discharged of 0.9 mg/dl.

Discussion

Mortality rate of perforated colon in adults treated with staged operations of primary resection is high, occurring in up to 45% of patients.[2] In post kidney transplant recipients receiving immunosuppressive therapy, colonic perforation causes higher mortality when compared to patients with normal immunity. One study reported the mortality rate of perforated diverticulitis in post renal transplant recipients with ADPKD was 100%.[3]

The incidence of diverticulosis in non-ADPKD with chronic renal failure is similar to that in the general population (32% and 38% respectively).[4] However, ADPKD is commonly associated with colonic diverticulosis[3] which is the leading cause of perforated colon in this population.[5] The prevalence of ADPKD had been reported from 2.8% to 4.1%,[3,6] and prevalence of colonic diverticulosis in ADPKD patients varies from 53.5% to 83%[3,4,6] The onset of intestinal perforation likely occurs in the first 3 weeks post kidney transplantation.[7]

The most common site of diverticulosis in ADPKD is the sigmoid colon.[3] It does not correlate with the location of transplanted kidney.[5] It is thought that a congenital colonic defect in ADPKD contributes to colonic diverticulosis or perforation.[4] Even though, colonic diverticulosis is commonly found in ADPKD patients, perforation is uncommon.[5] Perforated colon in ADPKD patients had been reported from 2% to 4%.[5,8] In one study, colonic diverticulitis was the cause of colonic perforation in 6 out of 13 post kidney transplant recipients (46%);[5] how-

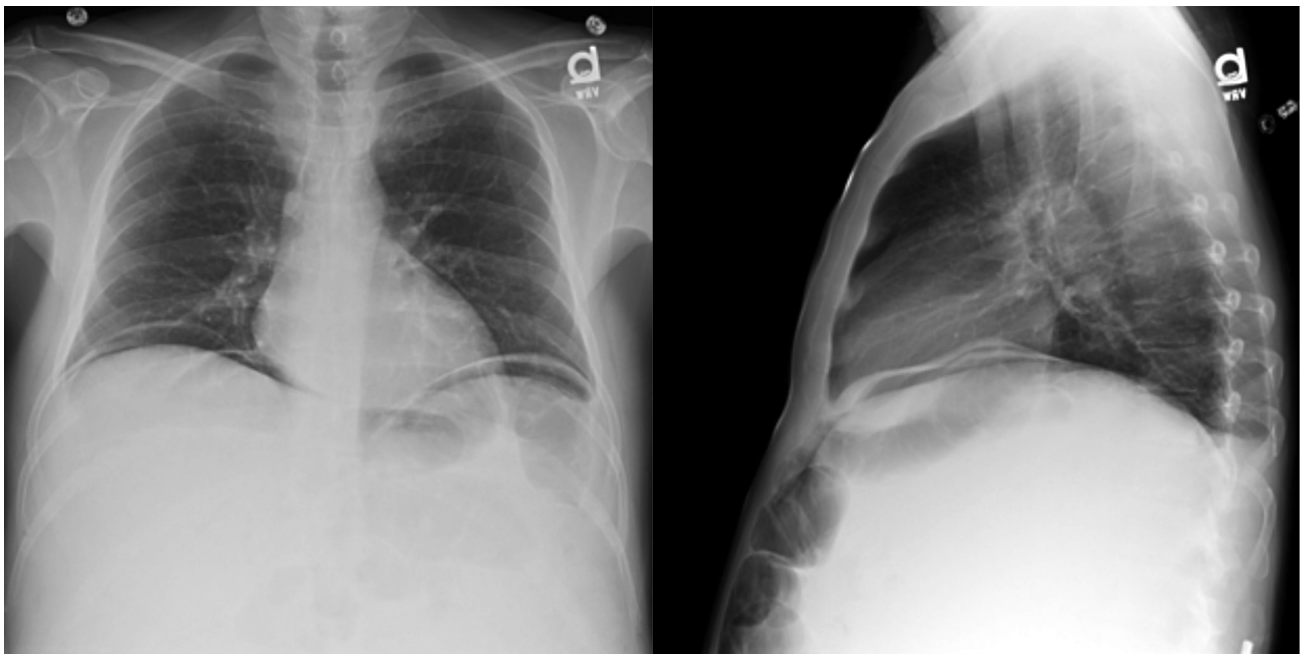

Figure 1. Chest x-ray upright in PA and lateral views on the first day of admission showed free air under diaphragm.

ever, a more recent study reported only one case of perforated colonic diverticulitis out of 46 post kidney transplant patients with ADPKD (2.2%).[6] The decreased incidence of perforated colonic diverticulitis from 46% to 2.2% could be due to changes in immunosuppressive regimens especially reduction in corticosteroid doses.[6] Being a well-known cause of gastroduodenal ulcers, corticosteroid use is also associated with colonic ulceration.[9-12] Its inhibitory effect on collagen synthesis facilitates colonic perforation. In addition, it inhibits inflammatory cells, some pyrogenic interleukins and pain-inducing prostaglandins, masking the normal inflammatory response.[13,14] As a result, symptoms and classic signs of acute abdomen may not occur in intra-abdominal visceral perforations under immunosuppression, and this leads to a diagnostic challenge. Apart from extrarenal manifestation such diverticulitis, abdominal pain in ADPKD patients may result from polycystic kidneys. Enlarged kidneys in ADPKD patients can cause chronic abdominal pain and their complications such as ruptured cysts may present with acute abdominal pain which should be one of the differential diagnoses of acute abdomen in ADPKD patients.

The most common cause of death in perforated colonic diverticulitis in post kidney transplant recipients is sepsis, and one of the most important factors determining the survival outcome after perforation is the duration from the onset of symptoms until surgical treatment. Higher survival rate occurs in the patients undergoing surgery within 24 hours after the onset of symptoms. In addition, renal graft function during the first week after perforation predicts the survival. Patients whose postoperative serum creatinine is less than 2.5 mg/dl have a higher survival rate than those with a serum creatinine more than 2.5 mg/dl.[5] Many studies have reported a high mortality rate of colonic perforation in post kidney transplant recipients receiving corticosteroids;[10,11] however, the dose of prednisone the patients received before perforation was not different between surviving and non-surviving groups.[5]

There is association between ADPKD and colonic diverticulosis, but complications of diverticulosis including diverticulitis and perforation are not common. However, these fatal complications in post kidney transplant recipients have raised the need to consider aggressive prevention including workup for colonic diverticulosis and further elective colectomy in ADPKD patients who will undergo kidney transplantation. Thus far there is still no conclusion in this matter.[3] It is reasonable to have this workup for patients who have previous history of symptomatic diverticulosis.[5] However, it is crucial to recognize and diagnose early for possible perforated colonic diverticulitis in post kidney transplant recipients with ADPKD presenting with abdominal pain as well as to initiate appropriate antibiotics, decrease immunosuppressive medication, and promptly initiate surgical treatment, as all of these lead to improved survival.[15]

This patient presented with vague, non-specific abdominal pain, and benign abdominal signs which were out of proportion to the underlying perforated sigmoid diverticulitis. Immunosuppressive medications, especially glucocorticoids, play an important role in not only causing perforated sigmoid diverticulitis, but also masking classic symptoms and signs of acute abdomen. As a result, perforated diverticulitis should be one of the differential diagnoses of abdominal pain in post renal transplant recipients;[4] even though signs of peritonitis may be absent. Early recognition and prompt treatment are crucial and decrease morbidity and mortality.

Conclusion

ADPKD is associated with colonic diverticulosis. Post allogeneic kidney transplant recipients with ADPKD may develop perforated diverticulitis which is a life-threatening condition. A high index of suspicion is needed to recognize and promptly treat this complication early, as typical presentations may not occur in the patients who are immunosuppressed.

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Conflict of interest

The authors report no conflict of interest.

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