Case Report

Spontaneous splenic rupture due to uremic coagulopathy and mortal sepsis after splenectomy

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ABSTRACT

Nontraumatic spontaneous splenic rupture (NSSR) has been encountered much more rarely compared with the traumatic splenic rupture. Although NSSR generally emerges in dialysis patients on account of such causes as the use of heparin during hemodialysis, uremic coagulopathy, infections, and secondary amyloidosis. Herein, we aimed to present a case of spontaneous splenic rupture which had developed soon after the inclusion of the case suffering from end-stage renal disease in routine hemodialysis program in the absence of any trauma or other prespecified risk factors for splenic rupture. A 55-year-old male patient was admitted to our hospital to have the ureteral double J stent removed. The operation was completed without any complication. Complaining an abdominal pain more prominent in the left upper abdominal quadrant in the first postoperative day, the patient underwent a through physical examination which disclosed abdominal distension, widespread tenderness, and rebound and defense positivity. The abdominal tomography depicted 122 × 114 × 95 mm lesion compatible with a hematoma. On the basis of these findings, an emergency exploratory operation was decided to be performed. Following clearance of the retroperitoneal hematoma, splenectomy was implemented. Experiencing progressive deterioration in his clinical status despite antibiotic therapy, the patient unfortunately died of sepsis with multiorgan failure on the 25th postoperative day. In conclusion, NSSR is such an entity that may be missed out, can pursue variable clinical courses, and requires emergency therapy upon definitive diagnosis. The possibility of spontaneous bleedings should be kept in mind in any case with the history of hyperuricemia even in the absence of overt trauma, no matter if they are included in routine hemodialysis or not.

Key Words: Coagulopathy, splenectomy, splenic rupture, spontaneous, uremia

INTRODUCTION

Rupture of the spleen is generally seen following a blunt abdominal trauma and may prove to be a life-threatening entity.[1] Nontraumatic spontaneous splenic rupture (NSSR) has been encountered much more rarely compared with the traumatic splenic rupture.[2] Among some of the risk factors mentioned in the literature are malignancies, extramedullary hematopoiesis,[3] splenic infarcts,[4] thrombocytopenia[5] and other coagulopathies,[3] anticoagulant therapy, amyloidosis[6] portal hypertension[7] soft tissue diseases,[8] intrasplenic venous thrombosis,[9] malarial infestation,[10] aortic valve replacement due to bacterial endocarditis,[11] and focal splenic lesions.[12]

Although NSR generally emerges in dialysis patients on account of such causes as the use of heparin during hemodialysis, uremic coagulopathy, infections, secondary amyloidosis, and pancreatic or splenic infarctions,[13,14] true etiology often remains to be elucidated.[1]

Herein, we aimed to present a case of spontaneous splenic rupture which had developed soon after the inclusion of the case suffering from end-stage renal disease (ESRD) in routine hemodialysis program in the absence of any trauma or other prespecified risk factors for splenic rupture.
CASE REPORT

A 55-year-old male patient was admitted to our hospital to have the ureteral double J stent removed, which had been implanted during endoscopic treatment of left ureteric calculus 1 month previously. His medical history revealed no specific disease condition or surgical intervention other than being under a follow-up program for compensated chronic renal failure for 1 year, a daily diuresis of 2500 cc volume and the ureteroscopic intervention 1 month previously. In the laboratory analysis, blood glucose, urea, creatinin, uric acid, sodium and potassium levels were measured to be 110 mg/dL, 259 mg/dL, 8.8 mg/dL, 7.4 mg/dL, 138 mmol/L, 4.93 mmol/L, respectively. Moreover, the liver function tests were within normal range; hemoglobin level, hematocrit, white blood cell (WBC) count, platelet count were detected to be 7.7 gr/dL, 24 (%), 6000/uL, 250.000/uL, respectively, together with normal prothrombin (PT) and activated partial thromboplastin times (aPTT). Urinary system ultrasonography reported bilateral reduced kidney sizes and increased parenchymal echogenicity compatible with ESRD. With respective creatinin clearance and protein level detected to be 7.9 ml/min and 2364 mg/day in 24-hour urine test, the patient was then included in routine hemodialysis program after having been consulted to the department of nephrology. The ureteric catheter was removed through cystoscopy under sedation in the operating theater 1 day after the second hemodialysis session. The operation was completed in a total of 10 min without any complication. Complaining an abdominal pain more prominent in the left upper abdominal quadrant in the 1st postoperative day, the patient underwent a thorough physical examination which disclosed abdominal distension, widespread tenderness, and rebound and defense positivity. As for his vital signs, the body temperature was 36.5°C; pulse rate was 105/min; respiratory rate was 20/min; and, blood pressure was 85/50 mm Hg. Laboratory analysis revealed the following findings: Urea 119 mg/dL; creatinin 4.2 mg/dL; sodium140 mmol/L; potassium 4.16 mmol/L; hemoglobin 6.7 gr/dL; hematokrit 19.9 (%); WBC count 8000/uL; platelet count 57.000/uL; PT 21.7 s; aPTT 110 s; and, INR 1.91. The abdominal tomography depicted a high-density (75 HU) lesion of 122 × 95 mm dimension compatible with a hematoma which was localized in retroperitoneal region and extending from left upper renal pole, where it possessed an indistinguishable border into the subdiaphragmatic area [Figure 1]. On the basis of these findings, an emergency exploratory operation was decided to be performed on the patient. During the exploration, free fluid with hemorrhagic character was observed in the abdomen, together with a number of capsular laceration sites and accompanying hematomas. Following clearance of the retroperitoneal hematoma, splenectomy was implemented. Fresh whole blood, fresh frozen plasma, and thrombocyte suspension were transfused intraoperatively, four units for each. Suffering from persistent thrombocytopenia and high INR level despite stable vital signs and hemoglobin and hematocrit levels, the patient was then consulted to the department of hematology. Following a definitive diagnosis of uremic coagulopathy by the department of hematology, the patient was put under close monitoring together with platelet and fresh frozen plasma replacements. On the postoperative 3rd day, all the blood parameters became stabilized, rendering further transfusions unnecessary. Furthermore, complying with the recommendations by the department of infectious diseases, a set of pneumococcal, Haemophilus influenza type B (Hib), and meningococcal vaccines were administered postoperatively to the patient. On the 10th postoperative day, the patient was consulted to the chest physician due to emergence of respiratory distress and high fever. Suspected to have incurred a pulmonary thromboembolism, the patient underwent a pulmonary computed tomography angiography, which depicted no thromboembolism. On the strength of both sputum and blood culture positivity for the growth of Acinetobacter, intravenous colimycin therapy was commenced on the recommendations of the infectious disease physician. Thereafter, he was transferred to the intensive care unit for more stringent monitoring due to persistent high fever, deteriorating respiratory distress, progressive hypotension, and emerging need for further oxygen therapy. Experiencing progressive deterioration in his clinical status despite antibiotherapy, the patient unfortunately died of sepsis with multiorgan failure on the 25th postoperative day.

DISCUSSION

The first case report ever known regarding NSSR in the literature was presented by Atkinson in 1874.[15] NSSRs constitute approximately 1% of all cases of splenic rupture.[16] Despite their rarity, they are such entities
that are likely to pursue a mortal course upon diagnostic delay. While it is of much more ease to establish the diagnosis of splenic rupture in trauma admissions, the diagnosis of spontaneous splenic rupture in atraumatic cases may prove rather challenging. Although the exact mechanism by which NSSR occurs has still yet to be thoroughly resolved, three mechanisms see collective acceptance. These are physical activity-related increase in the intraabdominal pressure and compression applied by vigorously contracting abdominal muscles, increase in the intrasplenic pressure resulting from the cellular congestion, and promoted endothelial fragility by such events as thromboses and infarctions within the spleen.\[17,18\]

The hyperuricemia evident in ESRD patients induces dysfunctional platelet adhesion and aggregation.\[19\] Uremic platelet dysfunction has been characterized, in most of the cases, with prolonged bleeding time.\[20\] The levels of such substances playing active role in the mechanism of hemostasis as Thromboxane A2, collagen, thrombin, and arachidonic acid appear to have decreased in patients with ESRD.\[21,22\] Some experimental studies reported loss of the functions of the platelets which had been transferred from a normouricemic plasma into a hyperuricemic one,\[23\] which was ascribed to the possibility that the toxins residing in the hyperuricemic plasma may disrupt the structure of the enzymes involved in the haemostatic cascade through creation of a metabolic imbalance.\[24\]

Thanks to the process of hemodialysis, toxic substances that disrupt the platelet functions, such as urea, creatinin, phenol and phenolic acid, are removed from the body.\[25\] Accordingly, one can assume that hemodialysis may improve the platelet functions. However, studies revealed no correlation between the bleeding time and the plasma levels of these toxins. Moreover, it is also another fact that the heparin administered during the process of hemodialysis fosters a further inclination to bleeding. It is, therefore, debated how exactly the hemodialysis affects the bleeding time and hemostasis in patients with ESRD.\[26\] In this regard, the inclination of the patients with ESRD towards bleeding generated by chronic exposure to the toxins which disrupt the platelet functions should always be taken into consideration, no matter if the patients undergo routine hemodialysis. Moreover, hypertension which is a common accompaniment of ESRD constitutes another risk factor for spontaneous bleeding in these patients.

Our patient was such a case that carried no risk factor other than suffering from ESRD for 1 year and malignant hypertension. Left upper quadrant pain, the most commonly seen symptom in NSSR, was also the first and the most conspicuous symptom in our case. In addition to this symptom, the findings that can emerge due to peritoneal irritation carve out another indication for a surgical intervention. Our patient also experienced such peritoneal irritation findings as widespread abdominal tenderness and defense and rebound positivity.

Splenectomy cannot possibly be performed in all cases with NSSR. While medical therapy and follow-up in conjunction with ultrasonography (USG) controls is recommended for the patient with stable hemodynamic status, it is surgery that is recommended for those under unstable hemodynamic condition, those with the findings related to peritoneal irritation, those in whom radiological investigations depicted an intraperitoneal hematoma, those with enlarging subcapsular hematomas, and those in whom an intraparenchymal bleeding has been detected.\[27\] One alternative among the spleen-preserving approaches has been the method comprising a constellation of splenic arterial embolization, subsegmental resection, and transposition of the spleen. With unstable hemodynamic status and the presence of peritoneal irritation findings constituting an ultimate indication for a surgical exploration, our patient underwent an emergency splenectomy.

Some infection-related findings that are likely to vary on the basis of patient age may emerge in the patients in the postsplenectomy period. A mortal sepsis may pursue especially in patients in whom splenectomy was performed in the childhood period.\[28\] In an attempt to minimize the infections, vaccines especially against the capsular bacteria should be injected in the immediate postsplenectomy period. Vaccination and close follow-up are of paramount importance for the purpose of the fight against infectious diseases after splenectomy especially in the patient groups with ESRD who are well-known for their inclination to the infections. Accordingly, our patient died of sepsis despite being in his adulthood, relevant vaccinations administered, and close follow-up within the hospital.

**CONCLUSION**

NSSR is such an entity that may be missed out, can pursue variable clinical courses, and requires emergency therapy upon definitive diagnosis. The possibility of spontaneous bleedings should be kept in mind in any case with the history of hyperuricemia even in the absence of overt trauma, no matter if they are included in routine hemodialysis or not. Furthermore, NSSR should be taken into consideration in ESRD patient with abdominal tenderness and especially pain in the upper left abdominal quadrant. The choice between spleen-preserving managements or other surgical methods should be dictated by the degree of the rupture. Spleen-preserving surgery should be preferred whenever the spleen is feasible to be protected, whereas splenectomy should be reserved only for the aforementioned indications,
keeping in mind that the immune system is likely to deteriorate partially after splenectomy.

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Cite this article as: Gazel E, Açikgöz G, Kasap Y, Yigman M, Gunes ZE. Spontaneous splenic rupture due to uremic coagulopathy and mortal sepsis after splenectomy. Int J Crit Illn Inj Sci 2015;5:119-22.

Source of Support: Nil, Conflict of Interest: None declared.