Case Reports in Oncology

Case Rep Oncol 2022;15:277–284
DOI: 10.1159/000522663
Received: January 9, 2022
Accepted: February 10, 2022
Published online: March 21, 2022

Case Report

Atraumatic Splenic Rupture due to Chronic Myelomonocytic Leukemia Treated with Partial Splenic Artery Embolization

Yunan Nie[a] Andrew Kent[b] Minh Do[c] Maria Amaya[b, d]
Catherine Klein[d] Christiane Thienelt[d]

[a]Department of Internal Medicine, University of Colorado, Anschutz Medical Campus, Aurora, CO, USA; [b]Department of Hematology and Oncology, University of Colorado, Anschutz Medical Campus, Aurora, CO, USA; [c]University of Colorado School of Medicine, Anschutz Medical Campus, Aurora, CO, USA; [d]Department of Hematology/Oncology, Rocky Mountain Regional VA, Aurora, CO, USA

Keywords
Chronic myelomonocytic leukemia · Atraumatic splenic rupture · Splenic embolization

Abstract
Splenic rupture can be categorized into two groups: traumatic and atraumatic. Traumatic rupture is frequently associated with blunt abdominal trauma, while atraumatic splenic rupture (ASR) is more uncommon and has been associated with both benign and malignant hematological disorders. In general, most cases of splenic rupture are managed with splenectomy, which carries significant mortality and morbidity; more recently, splenic artery embolization (SAE) has become a mainstay of management particularly after traumatic rupture. We describe a patient with chronic myelomonocytic leukemia (CMML) who presented to the emergency department for acute abdominal pain and was found to have an ASR. He underwent partial SAE, with postoperative complications of leukocytosis and tumor lysis syndrome (TLS) requiring rasburicase and allopurinol. On follow-up in clinic 2 months post-discharge, the patient was doing well on hydroxyurea, without need for further intervention at that time. In patients with hematologic malignancies presenting with abdominal pain and splenomegaly, it is important to consider ASR as a rare, but possible complication. To our knowledge, this is the only reported patient treated with SAE in the context of ASR from CMML, demonstrating that SAE can be an effective nonoperative strategy for treatment of CMML-associated ASR. This case report also highlights postoperative complications and management in this patient

Correspondence to:
Minh Do, minh.do@cuanschutz.edu

Case Rep Oncol 2022;15:277–284
DOI: 10.1159/000522663

population, specifically a profound leukocytosis and TLS, for which close monitoring should be performed.

Background

Atraumatic splenic rupture (ASR) is a rare but life-threatening cause of acute abdominal pain in patients without traumatic history and comprises 0.1–0.5% of cases of splenic rupture [1]. Splenic rupture is frequently a clinical diagnosis; although computed tomography (CT) imaging or ultrasonography (such as identification of free fluid on a FAST exam) can be helpful in confirming the diagnosis, maintaining ASR on the differential for acute abdominal pain is essential. Timely diagnosis can be especially difficult as the classic triad of left hemidiaphragm elevation, lower lobe atelectasis, and pleural effusion is frequently absent [2].

Most patients with ASR have baseline splenic abnormalities, such as splenomegaly [3]. Common causes are malignant hematological disorders (16.4%), viral infections (14.8%), and local inflammation/neoplasm (10.9%) [3]. Timely diagnosis of splenic rupture is essential as early intervention with angiography is associated with splenic salvage and improved outcomes, and splenectomy can be associated with a mortality rate as high as 2% [4, 5].

Patients with hematological conditions are at risk for splenic abnormalities, making the diagnosis of ASR an important consideration when evaluating acute abdominal pain. The mechanism by which splenic rupture occurs in these patients has been hypothesized to be due to several etiologies [6, 7]. The most frequent pathology is infiltration of spleen by the malignant cells, causing rupture of the relatively nondistensible splenic capsule through mechanical pressure. ASR can also be caused by splenic hemorrhage related to thrombocytopenia. In addition, splenic infarcts may cause architectural changes and altered vascular resistance that predispose to rupture. It is quite likely that all three of these pathways are involved in splenic rupture in these patients.

Guidelines for management of ASR are primarily based on data from traumatic splenic rupture cases and frequently involve splenectomy in hemodynamically unstable patients, although nonoperative management (NOM) is now considered the gold standard in hemodynamically stable patients with minor to moderate lesions [8]. Here we present what we believe to be the first reported case of ASR due to chronic myelomonocytic leukemia (CMML) treated with partial splenic artery embolization (SAE).

Case Description

DO was a 74-year-old man with history of papillary thyroid cancer in remission and post-surgical hypothyroidism. He was previously diagnosed with myelodysplastic syndrome, with mild peripheral cytopenias and bone marrow biopsy with normal karyotype and <5% blasts. He was initially treated with decitabine and subsequently observed by primary care. He was re-referred to hematology at our institution 13 years later for rising white blood cell count (~30,000/μL), monocyte predominant with 1–2% circulating blasts, and thrombocytopenia (40–70,000/μL). Repeat bone marrow biopsy showed persistent disease with stable blasts, and a normal karyotype with SRSF2, ASXL1, TET2, and KRAS mutations identified on next-generation sequencing. Given his persistent monocytosis and mutations, he was diagnosed with CMML in proliferative stage, with plan to start hydroxyurea.

Case Rep Oncol 2022;15:277–284
DOI: 10.1159/000522663

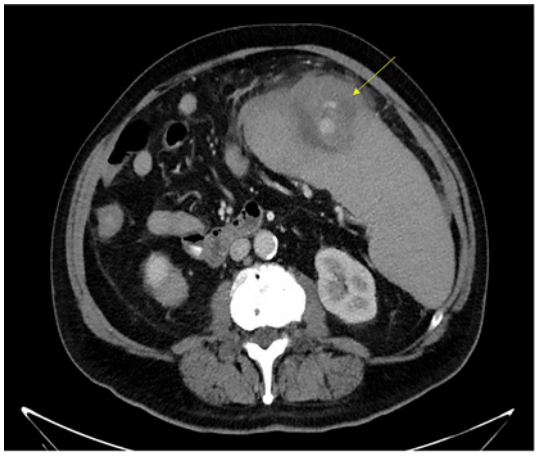

Fig. 1. CT A/P with contrast on admission demonstrating marked splenomegaly, an ill-defined 5 × 6 cm mass within the spleen (of mixed densities some or all of which may be hemorrhagic), and moderate volume-free fluid in the pelvis, small perisplenic, and trace perihepatic.

However, prior to initiating hydroxyurea, he presented to the emergency department with acute left upper abdominal pain with chills, malaise, and lightheadedness, without history of trauma. His vitals were significant for tachycardia with a heart rate of 99 and hypotension to 103/63, with an exam with dry mucous membranes, abdominal tenderness worst in his left upper quadrant with rebound and guarding, and splenomegaly. His initial WBC was 79.9, increased from 47.2 4 days prior, hemoglobin 13.2 g/dL, platelets 52,000/μL. A CT scan demonstrated marked splenomegaly (25 cm in greatest dimension) with a 5 × 6 cm mass of mixed densities suggestive of a hemorrhagic lesion with associated splenic capsular disruption and moderate volume free fluid in the pelvic, perisplenic, and perihepatic regions concerning for hemoperitoneum (shown in Fig. 1).

He was evaluated by surgery and interventional radiology, and ultimately taken for partial SAE. During the procedure, active extravasation was noted from the interior spleen which was successfully stopped with selective embolization. The following day, his hemoglobin had downtrended to 10.9 g/dL, after transfusion of 2 units of blood. His WBC rose to a peak of 118,000/μL (1–2% blasts), for which hydroxyurea was initiated, and his platelets increased to 100,000/μL (shown in Fig. 2).

Hospital course was complicated by acute kidney injury, hyperkalemia, hyperphosphatemia, elevated uric acid, and elevated lactate dehydrogenase, concerning for "tumor" lysis. He received one dose of rasburicase and started allopurinol (shown in Fig. 2). All parameters improved and he was discharged on day 8.

Imaging post-discharge demonstrated a decreased splenic size, with a large cystic component measuring 22 × 13 × 21 cm, likely related to post-embolization liquefaction (shown in Fig. 3). At his 2-month follow-up, the patient was doing well on hydroxyurea, without further intervention.

Discussion

Overall ASR related mortality rate is 12.2%, with lower rates of mortality in patients with NOM [3]. In one review, 85.3% underwent surgery with 7.4% mortality, while 14.7% had nonsurgical conservative treatment with 4.4% mortality [3]. Seven in the conservative group underwent SAE with 0% mortality. Compared to traumatic splenic rupture, ASR patients have lower rates of organ preserving surgery or nonsurgical management, likely related to older patient age and baseline splenic abnormalities, as in our patient. In our patient, diagnosis was confirmed preoperatively, and he was appropriately evaluated for splenectomy versus SAE,

Case Rep Oncol 2022;15:277–284
DOI: 10.1159/000522663

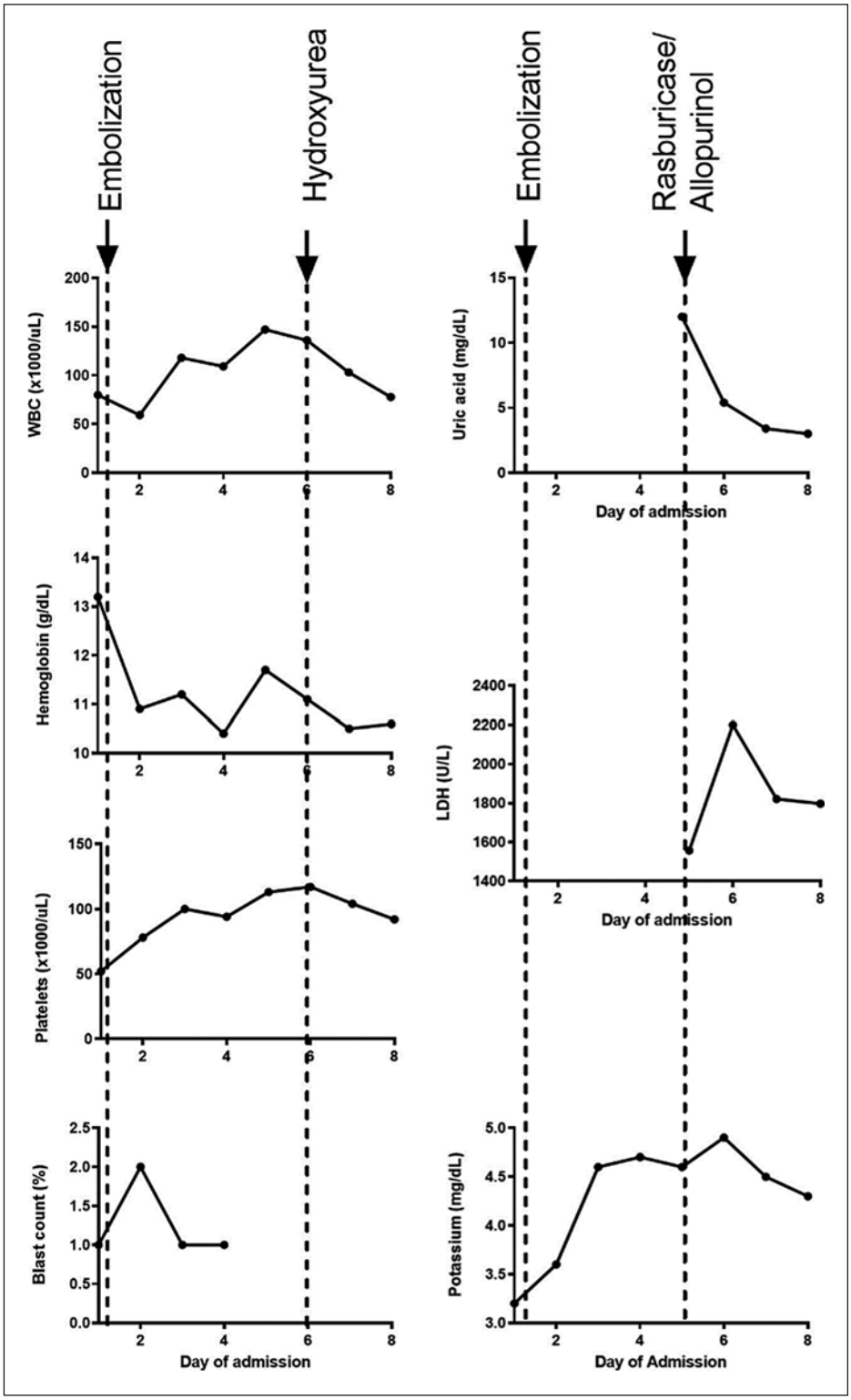

Fig. 2. Pertinent laboratories during hospitalization. Splenic embolization was performed on day of admission. Rasburicase/allopurinol was administered on hospital day 5, and hydroxyurea was administered on day 6.

Case Rep Oncol 2022;15:277–284
DOI: 10.1159/000522663

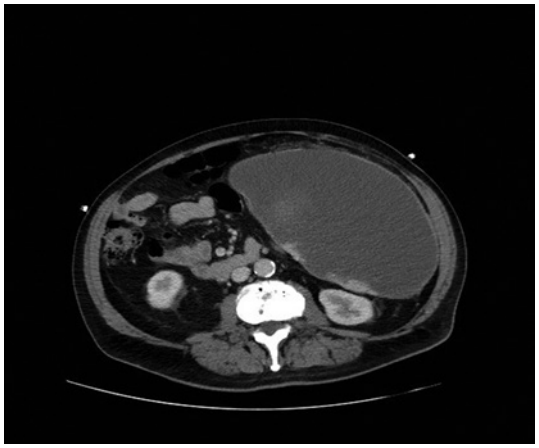

Fig. 3. CT A/P with contrast at 1-month follow-up demonstrating post-embolization liquefaction, overall splenic size is slightly decreased since the previous CT, measuring up to 22 cm craniocaudal versus 24 cm previously. Now large cystic component measuring 22 × 13 × 21 cm, likely relating to post embolization liquefaction. No significant perisplenic inflammatory fat stranding to suggest superimposed infection.

a minimally invasive procedure that allows for splenic salvage. Early recognition of ASR as a cause of acute abdominal pain in patients with CMML is therefore essential.

To our knowledge, this is the only case of ASR in the setting of CMML that has been treated with SAE. In general, SAE is well tolerated, with one study finding 73.1% of patients had a positive outcome with shorter hospital stays, fewer intensive care unit days, and fewer transfusions [9]. Although 8% of patients developed delayed complications (hemorrhage, splenic abscesses, and splenic pseudocysts), most patients did well [10]. In addition, there is evidence that immunological function is preserved after SAE [11]. In the most recent World Society of Emergency Surgery (WSES) guidelines, most hemodynamically stable patients without severe lesions can by monitored or undergo angiography rather than laparotomy, assuming early CT is obtained to triage severity of lesions [8]. A laparoscopic approach is an alternative to laparotomy and SAE. While laparoscopic splenectomy is preferred for benign or planned splenectomies, there are relatively few reported cases of patients with splenic rupture who undergo this technique [12, 13]. Of those patients, most were hemodynamically stable and had failed NOM, with generally positive outcomes. While it appears that this is a safe alternative to a laparotomy, patient selection is key and not every patient is eligible. At this time, there are no consensus guidelines for ASR.

On review of the literature, only 7 prior cases of splenic rupture in CMML have been reported, all treated with splenectomy (Table 1) [14]. Of these 7 patients, outcomes were usually positive or not reported; however, splenectomy in general is often complicated by bleeding and infectious complications, especially in patients with abnormal spleens. Prophylactic splenectomy has also been considered in CMML; in one study, 3/12 patients who underwent splenectomy died as a direct result of the surgery while an additional 4 patients suffered significant post-op complications [15]. There is clearly significant mortality and morbidity associated with splenectomy in CMML patients and SAE may provide an alternative.

Post-procedure leukocytosis is common and raises concerns for CMML progression. Three patients described by Steensma et al. [15] who had prophylactic splenectomies for CMML had postoperative leukocytosis >100,000/μL requiring hydroxyurea. Tracking blast percentage therefore can be reassuring against progression or transformation of hematologic disorder. In our patient, although his leukocytosis peaked at 118,000/μL, his blast percent remained between 1 and 2% with appropriate response to hydroxyurea.

Complications in our patient included elevated uric acid, potassium, and phosphorus, raising concern for a "tumor" lysis syndrome (TLS) and were treated as such with rasburicase and allopurinol. Although the patient did have an AKI, it was thought to be in the setting of hypovolemia/hypotension, rather than directly related to TLS. It could therefore be important to monitor TLS labs (potassium, phosphorus, calcium, uric acid, and creatinine) more frequently in these patients.

Case Rep Oncol 2022;15:277–284
DOI: 10.1159/000522663

Table 1. Seven prior cases of splenic rupture in CMML, all treated with splenectomy

Patient	Reference	Age	Sex	Clinical scenario	Intervention	Outcome
1	Abbasi AM, Adil S, Moiz B. Spontaneous splenic rupture - An uncommon complication of chronic myelomonocytic leukemia. Leuk Res Rep. 2020; 14:100205. doi:10.1016/j.lrr.2020.100205 [9]	50	M	Presented to ED with dizziness, loose stools, and severe abdominal pain, with hypotension and tachycardia, anemia with Hgb 6.2	Ex lap and splenectomy	Leukocytosis to 80 10^9, discharged to outpatient follow-up
2	Goddard SL, Chesney AE, Reis MD, Ghorab Z, Brzozowski M, Wright FC, et al. Pathological splenic rupture: a rare complication of chronic myelomonocytic leukemia. Am J Hematol. 2007;82(5):405-408. doi:10.1002/ajh.20812 [7]	56	M	Presented to ED with abdominal pain, generalized weakness, presyncope, and nausea, with tachycardia and Hgb 8.3	Ex lap and splenectomy	No complications
3	Elliott MA, Mesa RA, Tefferi A. Adverse events after imatinib mesylate therapy. N Engl J Med. 2002;346(9):712–713	61	F	Pain in the left upper quadrant with CT demonstrating splenic rupture	Splenectomy	Not reported
4	Elliott MA, Mesa RA, Tefferi A. Adverse events after imatinib mesylate therapy. N Engl J Med. 2002;346(9):712–713	71	F	Increasing splenomegaly and constitional symptoms	Splenectomy	Not reported
5	Diebold J, Audouin J. Peliosis of the spleen. Report of a case associated with chronic myelomonocytic leukemia, presenting with spontaneous splenic rupture. Am J Surg Pathol. 1983;7(2):197–204	62	M	"Sudden spontaneous rupture of spleen"	Splenectomy	Not reported
6	Jimenez Herraez MC, Larrocha Rabanal C, Fernandez de Castro M, Viloria Vicente A. Pathological rupture of the spleen in a case of chronic myelomonocytic leukemia. Sangre. 1991;36(2):168	Unknown	Unknown	Unknown	Splenectomy	Unknown
7	Steensma DP, Tefferi A, Li CY. Splenic histopathological patterns in chronic myelomonocytic leukemia with clinical correlations: reinforcement of the heterogeneity of the syndrome. Leuk Res. 2003;27(9): 775–782 [10]	65	F	Spontaneous rupture	Splenectomy	Not reported

Overall, this case demonstrates several key points. First, a nonsurgical splenic sparing approach like SAE is feasible for patients with CMML who experience ASR. ASR is important to consider the differential for patients with hematological malignancies presenting with abdominal pain, as early diagnosis is essential in deciding splenectomy versus splenic sparing interventions. Profound leukocytosis can be common post-procedurally in patients with CMML and can be differentiated from AML transformation through monitoring of blast percentage. Finally, monitoring of TLS laboratories post-procedurally may be recommended in such patients.

Statement of Ethics

Ethics approval was not required in accordance with the local or national guidelines. Written informed consent was obtained from the patient for publication of the details of their medical case and any accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Funding Sources

No funding was acquired.

Author Contributions

Yunan Nie wrote and edited the manuscript and assembled tables and figures. Andrew Kent assisted with writing and editing the manuscript and assembled tables and figures. Minh Do assisted with writing and editing of the manuscript. Maria Amaya, Catherine Klein, and Christiane Thienelt provided feedback and edits on the manuscript.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

References

1 Gedik E, Girgin S, Aldemir M, Keles C, Tuncer MC, Aktas A. Non-traumatic splenic rupture: report of seven cases and review of the literature. World J Gastroenterol. 2008 Nov 21;14(43):6711–6.
2 Freiwald S. Late-presenting complications after splenic trauma. Perm J. 2010;14(2):41–4.
3 Renzulli P, Hostettler A, Schoepfer AM, Gloor B, Candinas D. Systematic review of atraumatic splenic rupture. Br J Surg. 2009 Oct;96(10):1114–21.
4 Qu Y, Ren S, Li C, Qian S, Liu P. Management of postoperative complications following splenectomy. Int Surg. 2013 Mar;98(1):55–60.
5 Zarzaur BL, Savage SA, Croce MA, Fabian TC. Trauma center angiography use in high-grade blunt splenic injuries: timing is everything. J Trauma Acute Care Surg. 2014 Nov;77(5):666–73.
6 Bauer TW, Haskins GE, Armitage JO. Splenic rupture in patients with hematologic malignancies. Cancer. 1981 Dec 15;48(12):2729–33.

Case Rep Oncol 2022;15:277–284
DOI: 10.1159/000522663

7 Goddard SL, Chesney AE, Reis MD, Ghorab Z, Brzozowski M, Wright FC, et al. Pathological splenic rupture: a rare complication of chronic myelomonocytic leukemia. Am J Hematol. 2007 May;82(5):405–8.
8 Coccolini F, Montori G, Catena F, Kluger Y, Biffl W, Reva V, et al. Splenic trauma: WSES classification and guidelines for adult and pediatric patients. World J Emerg Surg. 2017;12:40.
9 Sekikawa Z, Takebayashi S, Kurihara H, Lee J, Niwa T, Kawamoto M, et al. Factors affecting clinical outcome of patients who undergo transcatheter arterial embolisation in splenic injury. Br J Radiol. 2004 Apr;77(916):308–11.
10 Cocanour CS, Moore FA, Ware DN, Marvin RG, Clark JM, Duke JH. Delayed complications of nonoperative management of blunt adult splenic trauma. Arch Surg. 1998 Jun;133(6):619–5; discussion 624.
11 Tominaga GT, Simon FJ, Dandan IS, Schaffer KB, Kraus JF, Kan M, et al. Immunologic function after splenic embolization, is there a difference? J Trauma. 2009 Aug;67(2):289–95.
12 Nasr WI, Collins CL, Kelly JJ. Feasibility of laparoscopic splenectomy in stable blunt trauma: a case series. J Trauma. 2004 Oct;57(4):887–9.
13 Thapar PM, Philip R, Masurkar VG, Khadse PL, Randive NU. Laparoscopic splenectomy for spontaneous rupture of the spleen. J Minim Access Surg. 2016 Mar;12(1):75–8.
14 Abbasi AM, Adil S, Moiz B. Spontaneous splenic rupture: an uncommon complication of chronic myelomonocytic leukemia. Leuk Res Rep. 2020 May 29;14:100205.
15 Steensma DP, Tefferi A, Li CY. Splenic histopathological patterns in chronic myelomonocytic leukemia with clinical correlations: reinforcement of the heterogeneity of the syndrome. Leuk Res. 2003 Sep;27(9):775–82.