Título
Manejo de rotura esplénica espontánea en paciente con síndrome mielodisplásico: reporte de caso

Titulo inglés
Spontaneous Splenic Rupture in a Patient with Myelodysplastic Syndrome: A Case Report

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Resumen

Introducción: El síndrome mielodisplásico (SMD) es un trastorno hematopoyético clonal con riesgo de progresar a leucemia mieloide aguda. Este informe de caso discute una rara instancia de ruptura esplénica espontánea en un paciente con SMD.

Presentación del caso: Un paciente varón de 76 años con antecedentes de SMD presentó dolor abdominal agudo y se descubrió que tenía una ruptura esplénica. A pesar de rechazar la quimioterapia, el paciente se sometió a una esplenectomía de emergencia después de que una tomografía computarizada revelara la presencia de hemoperitoneo. Después de la intervención, el paciente experimentó shock hipovolémico; por lo que requirió una segunda operación.

Discusión: La ruptura esplénica en SMD es poco común y su patogénesis no se comprende completamente. El manejo del SMD es complejo, a menudo requiere un enfoque multidisciplinario. Este caso enfatiza la importancia de considerar las preferencias del paciente en las decisiones de tratamiento y destaca el papel de la secuenciación de nueva generación en la comprensión de la patogénesis del SMD.

Conclusión: El caso subraya la necesidad de vigilancia en el manejo del SMD debido a posibles complicaciones como la ruptura esplénica. También ilustra la importancia de la atención personalizada y los avances en el perfil genético para predecir el pronóstico y guiar el tratamiento en el SMD.
Abstract

Introduction: Myelodysplastic syndrome (MDS) is a clonal hematopoietic disorder with a risk of progression to acute myeloid leukaemia. This case report discusses a rare instance of spontaneous splenic rupture in a patient with MDS.

Case Report: A 76-year-old male with a history of MDS presented with acute abdominal pain and was found to have a splenic rupture. Despite declining chemotherapy, the patient underwent an emergency splenectomy after a CT scan revealed hemoperitoneum. Post-surgery, the patient experienced hypovolemic shock, necessitating a second operation. Discussion: Splenic rupture in MDS is uncommon and its pathogenesis is not fully understood. Management of MDS is complex, often requiring a multidisciplinary approach. This case emphasizes the importance of considering patient preferences in treatment decisions and highlights the role of next-generation sequencing in understanding MDS pathogenesis. Conclusion: The case underscores the need for vigilance in managing MDS due to potential complications like splenic rupture. It also illustrates the importance of personalized care and advances in genetic profiling in predicting prognosis and guiding treatment in MDS.

Introduction

Myelodysplastic syndrome (MDS) is a heterogeneous group of haematological neoplasms that affect hematopoietic stem cells, with a risk of evolving into acute myeloid leukaemia (1–4).

This diverse group of clonal disorders of the stem cells, characterized by ineffective hematopoiesis, peripheral blood cytopenias, and bone marrow dysplasia, presents a diagnosis based on specific criteria. The diagnosis includes hematological findings bone marrow (BM) findings, and the presence of certain genetic mutations (5).

Next-generation sequencing has identified recurrent somatic mutations in genes involved in various cellular splicing, DNA damage response, transcriptional regulation, and signal transduction (6). These mutations have provided valuable insights into the pathogenesis of MDS and have potential implications for prognosis and therapeutic strategies. The prognosis of MDS is conventionally assessed using the Revised International Prognostic Scoring System (IPSS-R), with additional adverse prognostic impact associated with specific genetic mutations, such as ASXL1, EZH2, or TP53 (7,8).

The clinical manifestation of MDS is highly variable and can include symptoms such as fatigue, weakness, pallor, fever, infections, bleeding, and easy bruising (9,10). These symptoms result from the cytopenias (red, white, and platelet series) that characterize this entity.

In the presented clinical case, the 76-year-old patient with a history of MDS presents with acute abdominal pain in the left hypochondrium, mucocutaneous pallor, abdominal distension with positive peritoneal irritation signs, and hypotension. In addition, the blood count reveals...
decreased haemoglobin values and thrombocytopenia. These findings are consistent with the clinical manifestations of MDS.

One of the serious complications of MDS is spleen rupture. Indeed, splenomegaly can occur in some cases of MDS due to extramedullary hematopoiesis (blood cell production outside the BM) or leukemic cell infiltration. Likewise, spleen rupture is an infrequent but potentially fatal complication. While the etiopathogenic mechanism of rupture is not well known, it is thought to involve multiple factors such as its size increase, splenic infarcts, or minor traumas (11).

Case Report

We present the case of a 76-year-old male patient with a history of myelodysplastic syndrome who attended to the emergency department of the Príncipe de Asturias University Hospital with acute abdominal pain in the left hypochondrium. The patient had declined chemotherapy following his diagnosis six months prior. He had no significant previous illnesses or relevant family history. On physical examination, the patient exhibited mucocutaneous pallor. Abdominal examination revealed abdominal distension with positive signs of peritoneal irritation and hypotension (92/75 mmHg). His body temperature was 309.65 K, heart rate was 80 beats per minute, and respiratory rate was 20 breaths per minute. The accompanying family member denied any traumatic history in the anatomical region.

In addition to the standard tests, a complete blood count was performed, which revealed low hemoglobin levels (10.2 g/dL), normal levels of mean corpuscular volume (10 fl), and a platelet count of 49,000 platelets/mm³. Leukocyte count was 2.12 10⁹/L. Neutrophils were 0.78 10⁹/μL. The rest of the parameters were within the normal range. The initial approach included chest radiography and urinalysis, both of which were normal.

An urgent abdominal CT scan was executed, disclosing bleeding within the cavity and hemoperitoneum. Given the patient’s deteriorating condition and the imminent risk of hypovolemic shock, immediate surgical intervention was considered for total splenectomy.

Upon entering the operating room, the patient was tachycardic, tachypneic, and exhibited mucocutaneous pallor.

Surgical findings included 1.8 L of hemoperitoneum with active bleeding, numerous clots, and splenic segments. To perform the splenectomy, the patient was pneumoperitoneum and the spleen was exposed. The patient was placed in a lateral decubitus position at a 40 degrees of inclination, using fixation supports.

The procedure followed this protocol: Clipping of the splenocolic ligament.
1. Ligation and sectioning of the splenic hilum.
2. Identification of possible supernumerary spleens.
3. Extraction of the spleen.

4. Four hours after the surgery, the patient suffered a hypovolemic shock and undergo a repeat procedure.

5. The reopening was performed by removing the staples and placing an Alexis surgical ring with the help of laparostats (Figure 1). The intervention consisted of aspiration (Figure 2), removal of clots (Figure 3) and supporting the patient's hemostasis with platelet transfusion.
Figure 2. Aspiration of bleeding liver cell

During the reopening of the splenectomy performed in the previous operation, the patient required a transfusion of 3 bags of erythrocytes, two bags of platelets, and one of plasma to achieve stabilization. The patient was initially transferred to the intensive care unit for monitoring. His postoperative course was satisfactory, specific treatment for myelodysplastic syndrome was initiated, and he was discharged 14 days after the surgical procedure.

Discussion

Myelodysplastic syndrome (MDS) is a complex and heterogeneous clinical entity characterized by ineffective hematopoiesis and a variable risk of progression to acute myeloid leukaemia. The clinical presentation can be highly variable, and associated complications, such as splenic rupture, are rare but can be potentially fatal (12).

In the case presented, a splenic rupture in a 76-year-old patient with MDS underscores the severity of complications that can arise during the disease.

Splenomegaly, which may be secondary to extramedullary hematopoiesis or leukemic cell infiltration, is a known manifestation of MDS. However, splenic rupture is an infrequent complication, and its pathogenesis is not fully understood, though factors such as increased splenic size, splenic infarcts, or minor traumas may contribute.

The management of MDS is complex and requires a multidisciplinary approach. While bone marrow transplantation offers a potential cure, it carries significant risks and is not a viable option for all patients (13). In this case, the patient had declined chemotherapy, highlighting the importance of considering patient preferences and potential...
outcomes in therapeutic decision-making. Current scientific literature reflects that the prognosis of MDS is affected by various genetic mutations, and next-generation sequencing has enabled a better understanding of the disease's pathogenesis. The Revised International Prognostic Scoring System (IPSS-R) is a conventional tool for assessing prognosis, and certain genetic mutations have been observed to have an adverse prognostic impact (14).

The case presented is particularly interesting due to the complication of splenic rupture, which required immediate surgical intervention and a second operation due to hypovolemic shock. This event emphasizes the need for careful surveillance and proactive management of complications in patients with MDS.

Conclusion

In conclusion, this clinical case highlights the complexity and heterogeneity of myelodysplastic syndrome (MDS), a disease that can present with serious and potentially fatal complications, such as splenic rupture. Although this complication is rare, its occurrence in a 76-year-old patient with MDS underlines the importance of careful surveillance and proactive management of complications in these patients. The management of MDS is challenging and requires a multidisciplinary approach and consideration of patient preferences in therapeutic decision-making. This case also underscores the importance of next-generation sequencing in understanding the pathogenesis of MDS and in evaluating prognosis.

In summary, this case provides a unique and valuable insight into the management of a rare but serious complication of MDS, offering substantial perspective in the management of hematopoietic disorders.

Competing Interests

The author declares that there are no competing interests.

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