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Open Splenectomy of Giant Spleen Secondary to Splenic Infarction in a Patient with Philadelphia Chromosome-Positive Acute Lymphoblastic Leukemia: A Case Report

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INTRODUCTION

Acute lymphoblastic leukemia (ALL) is a malignant neoplasm of immature lymphoid progenitor cells, characterized by an uncontrolled and excessive proliferation of these cells in the bone marrow and other hematopoietic organs. This disease primarily affects children and young adults, and is characterized by the presence of abnormal immature lymphoblastic cells in the blood and bone marrow.1

Splenic infarction, on the other hand, is a rare but potentially serious complication associated with various medical conditions. It refers to necrosis or death of splenic tissue due to interruption of adequate blood supply through the splenic arteries.2

In the context of ALL, an association between this disease and splenic infarction has been observed, although it is important to note that this relationship is not very common. It is postulated that splenic infarction in patients with ALL may be the result of multiple factors.

It is also important to mention that the Philadelphia chromosome is a reciprocal translocation between chromosomes 9 and 22, a chromosomal break occurs in two specific regions of chromosome 9 and 22, exchanging their positions and resulting in an altered chromosome 9 and a chromosome 22 also altered but characterized by the fusion of their genes (BCR-ABL), which encode an altered protein. The ABL gene in its normal situation expresses a protein tyrosine kinase, when the fusion with the BCR gene occurs, this tyrosine kinase activity is maintained, the effect of which generates a protein that remains continuously active, without regulation, which leads to an uncontrolled alteration of proteins and enzymes that cause alterations in the cell division cycle and consequently inhibit DNA repair, causing genome instability, giving rise to the disorganized creation of cancer cells, this anomaly aggravates the prognosis of the disease.3

One of the factors that could contribute to this relationship is the presence of leukemic cells in the blood circulation. In some cases of ALL, leukemic cells can infiltrate and obstruct blood vessels in the spleen, leading to decreased blood flow and ultimately splenic infarction.4

In addition, it is believed that thrombosis may play a role in the development of splenic infarction in patients with ALL. Abnormal clotting may occur due to the release of procoagulant substances by leukemic cells or as a result of alterations in the body's natural clotting mechanisms. These clots can block blood vessels in the spleen and lead to necrosis of splenic tissue.5

In addition, leukemic infiltration of the spleen can provoke a local inflammatory response, which contributes to blood vessel obstruction and subsequent infarction.5

It is important to note that splenic infarction in patients with ALL may present asymptomatically or with nonspecific symptoms, such as abdominal pain or malaise. In some cases, imaging studies, such as ultrasound or CT scan, may be necessary to confirm the diagnosis.6

EPIDEMIOLOGY

Regarding the epidemiology of ALL, it is estimated that about 3-4 new cases occur per 100,000 children under 15 years of age each year. The incidence of ALL shows a peak in preschool age, decreases in middle childhood, and then gradually increases during adolescence. The disease is slightly more common in boys than in girls.5

Regarding splenic infarction, it is important to mention that it is a rare complication associated with various medical conditions, including ALL. The precise incidence of splenic infarction in patients with ALL is not well established due to its rarity. However, it has been reported in the medical literature to occur in a small percentage of ALL cases, usually in advanced stages of the disease.6

PATHOPHYSIOLOGY

The pathophysiology of acute lymphoblastic leukemia (ALL) and splenic infarction involves a series of complex processes occurring at the cellular and tissue level.

In the case of ALL, a genetic alteration originates in immature lymphoid progenitor cells, leading to their uncontrolled proliferation and the accumulation of leukemic cells in the bone marrow and other hematopoietic organs. This abnormal proliferation affects the production of normal blood cells, causing a decrease in the amount of red blood cells, white blood cells and platelets in the blood circulation.

Leukemic infiltration of the spleen is a common finding in patients with ALL. Leukemic cells can infiltrate the splenic tissue, leading to obstruction of the splenic blood vessels. This obstruction can lead to decreased blood flow and, consequently, a reduction in the supply of oxygen and nutrients to the spleen. As a result, localized hypoxia occurs in the splenic tissue and a number of pathological responses are triggered.

Hypoxia and altered nutrient supply can lead to cellular dysfunction and ultimately tissue necrosis. In addition, leukemic infiltration into the blood vessels of the spleen may promote the formation of blood clots (thrombi) due to the interaction of leukemic cells with clotting components in the blood.

Thrombus formation may further obstruct blood vessels, aggravating the decrease in blood flow and contributing to the development of splenic infarction. These clots can generate a positive feedback loop, as thrombus-induced vascular obstruction further promotes hypoxia and splenic tissue necrosis.

In addition, leukemic infiltration of the spleen can trigger a localized inflammatory response. This inflammatory response involves the release of cytokines and other inflammatory molecules that recruit inflammatory cells and promote the migration of leukemic cells into splenic tissue. This localized inflammation may contribute to vascular obstruction and further tissue damage, exacerbating the onset of splenic infarction.

The pathophysiology of acute lymphoblastic leukemia and splenic infarction involves abnormal proliferation of leukemic cells in the bone marrow, leukemic infiltration of the spleen, vascular obstruction due to thrombus formation, tissue hypoxia, cell necrosis, and localized inflammatory response. These complex processes contribute to the clinical manifestation of splenic infarction in patients with ALL.

CASE PRESENTATION

19-year-old female with no pathological history of interest, without previous exposure to myelotoxic agents. She began her illness in September 2021 with asthenia, adynamia, profuse epistaxis and intermittent diffuse abdominal pain, with the presence of postprandial fullness, and later fever was added. She went to the emergency department where paraclinical tests were performed, highlighting leukemoid reaction, with pancytopenia of the rest of the cell lines, bone marrow aspirate and biopsy was performed, including acute lymphoblastic leukemia with the presence of chromosome 9:22.

He began the induction phase of chemotherapy with CVAD, reported persistent abdominal pain and fever, so a CT scan was performed, which reported severe splenomegaly extending to the pelvis and displacing adjacent structures, areas of infarction, retroperitoneal lymph node activity and hepatomegaly (Fig. 1, 2).

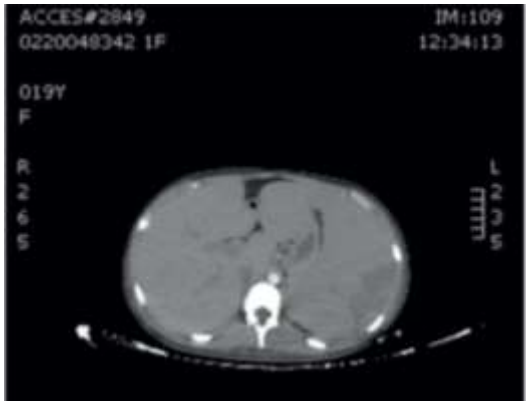

Fig.1 Computed axial tomography scan with contrast where the spleen is visualized giant with infarction in the periphery.

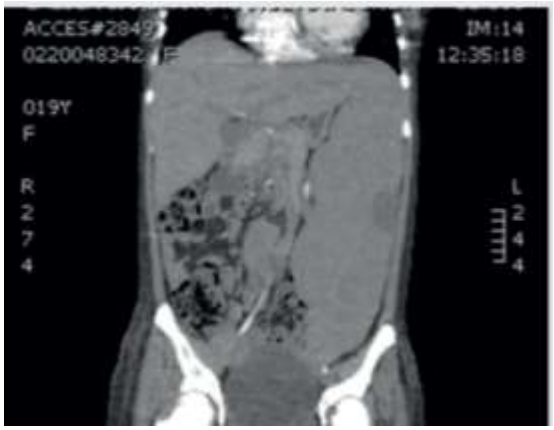

Figure 2. Coronal tomography with contrast where a giant spleen is observed extends to pelvis

Splenectomy was decided due to splenomegaly grade 4 according to the Boyd scale with presence of splenic infarction, which implies a high risk of infection and splenic sequestration. A midline approach was performed through a supra-infraumbilical incision, dissection of the supporting ligaments, ligation of the splenic bed with 1-0 silk,

verification of hemostasis, closure by planes and the surgical procedure was concluded (Fig. 3,4).

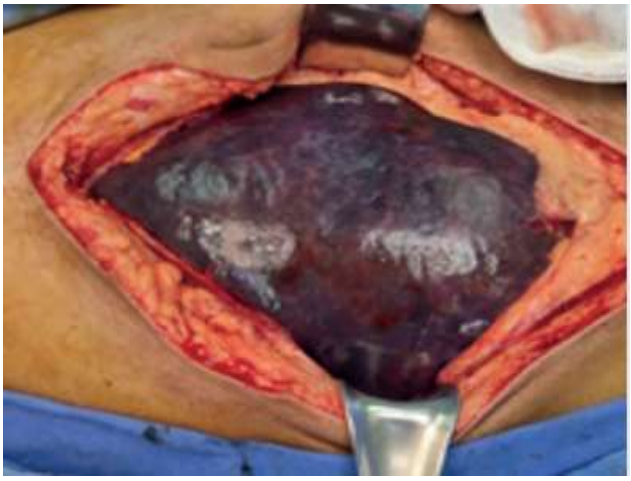

Fig.3 Midline approach showing the giant spleen.

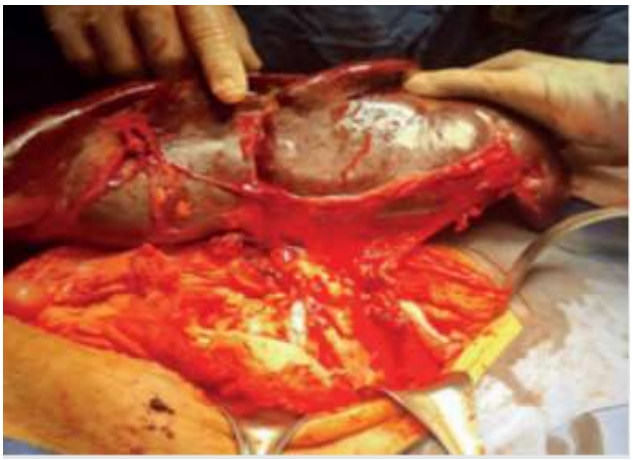

Fig. 4 Removal of spleen from the abdominal cavity.

Transoperative findings: ascitic fluid approximately 200 ml, enlarged spleen of approximately 50 x 20 cm, with areas of infarction of approximately 10 cm in diameter in parietal face, accessory spleens in number of 7, 3 adjacent to the tail and body of pancreas, 3 hilar and one in omentum.

2.Amount of bleeding during surgery: 400 ml.

Preoperative and transoperative preparation: Fasting 8 hrs before surgery, 6 pg and 1 platelet transfusion, prophylactic measures for thrombosis (bandage of pelvic limbs) and during surgery 3 pg and recombinant factor VII were requested.

Laboratories before surgery

Pre-surgical labs Leukocytes 21.6, neutrophils 1.2, lymph 79.8, hb 9.4, platelets 51, tp 15.4, tpt 29.1, inr 1.35

Post-surgical labs leukocytes 58.8 neutral 0.8, lymph 97, hb 10.7, platelets 57

In the postoperative period she was managed with Penrose type drainage to derivation, antibiotic, analgesia and transfusion of blood products, without presenting mediate complications, it was decided to discharge her home to continue with chemotherapy, however, the patient died 2 weeks later due to tumor activity and lack of response to chemotherapy.

RESULT

The relationship between acute lymphoblastic leukemia and splenic infarction may be related to leukemic infiltration of the spleen, blood clot formation and local inflammatory response. However, it is important to keep in mind that this complication is rare and may require specific evaluation and management by the specialized medical team.

Splenic infarction is associated with various predisposing factors such as oncologic, hematologic or infectious diseases. It is usually asymptomatic or symptomatic with abdominal pain, fever and splenomegaly. There is no consensus on which is the best treatment option, in most cases conservative or surgical treatment with splenectomy is decided according to the individual characteristics of the patient.

CONCLUSION

We present a clinical case of splenectomy with midline approach in a patient with giant splenomegaly who presented splenic infarction, in a context of acute lymphoblastic leukemia with poor prognosis, with the aim of preventing further splenic infarction and latent risk of infection or abscesses, as well as correction of cytopenias and improvement of symptoms secondary to massive size of this organ.

Acute lymphoblastic leukemia (ALL) may be infrequently associated with the occurrence of splenic infarction, a potentially serious complication. This relationship may be explained by several complex and multifactorial pathophysiological mechanisms.

The presence of leukemic cells in the blood circulation can lead to obstruction of splenic blood vessels, resulting in decreased blood flow and eventual necrosis of splenic tissue. Leukemic infiltration of the spleen can lead to the formation of blood clots and altered clotting mechanisms, promoting thrombosis and subsequent disruption of adequate blood supply to the spleen.

In addition, leukemic infiltration of the splenic tissue may induce a respiration of the splenic tissue.

Improvement in symptoms secondary to organomegaly was concluded, and improvement of cytopenias was documented, however, the patient died 2 weeks after the surgical procedure due to tumor activity and lack of response to chemotherapy.

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