ABSTRACT

Introduction: Infectious mononucleosis (IM) is a disease caused by the Epstein-Barr virus, characterized by the triad: fever, pharyngitis and adenopathy, and may be accompanied by splenomegaly and hepatitis. Spontaneous splenic rupture is a rare complication of IM and may be a cause of death.

Case report: The authors present two case reports of a 26 and a 44 year-old males, previously healthy, with suspicious and posteriorly confirmed infectious mononucleosis, who developed an acute abdomen with hemodynamic instability, requiring surgical intervention. The authors present treatment and the patients' evolution.

Discussion/Conclusion: Splenic rupture is a rare complication of IM, occurring in 0.1-0.5% of patients. The patients usually have an acute abdomen where its early detection and treatment are crucial.

Keywords: infectious mononucleosis, hemoperitoneum, spontaneous splenic rupture, splenic hematoma, Epstein Barr Virus.

RESUMO

Introdução: A mononucleose infeciosa é uma doença causada pelo vírus de Epstein-Barr, classificada pela tríade: febre, faringite e adenopatias, podendo ser acompanhada de esplenomegalia e hepatite. A rotura esplênica espontânea, apesar de ser uma complicação muito rara, é a principal causa de morte em doentes com mononucleose infeciosa.

Caso clínico: Os autores apresentam dois casos clínicos, ambos do gênero masculino, 26 e 44 anos, previamente saudáveis, com quadro sugestivo e confirmado de mononucleose infeciosa que desenvolveram um quadro de abdômen agudo com instabilidade hemodinâmica, necessitando de intervenção cirúrgica. Os autores apresentam o tratamento e evolução.

Discussão/Conclusão: A rotura esplênica é uma complicação rara desta doença, podendo ocorrer em 0.1-0.5% dos doentes. Cursa, geralmente, com um quadro de abdômen agudo e a sua detecção precoce e tratamento atempado são cruciais.

Palavras-Chave: mononucleose infeciosa, hemoperitoneu, ruptura esplênica espontânea, hematoma esplênico, vírus de Epstein Barr.
INTRODUÇÃO
Introduction
Infectious Mononucleosis (IM) is a disease caused by the Epstein-Barr Virus. It affects mainly young adults and teenagers with an incidence of 345-671 cases per 100,000 population per year [3].

IM is characterized by the triad of fever, pharyngitis and lymphadenopathy as well as an increase proportion of atypical lymphocytes [2]. Most cases are self-limited with pharyngitis subsiding within 7-10 days, fever 7-14 days and lymphadenopathy up to 3 weeks [2]. On physical examination splenomegaly was detected in 50 percent of the cases whereas by ultrasound it could be detected in almost 100 percent of the patients [2]. Ninety percent of the patients may have hepatitis with mild elevation in serum transaminases [2] and the level of bilirubin may also be increased due to hepatitis or haemolytic anaemia [8].

In spite of the disease having a benign clinical process, IM may cause haematological or neurological complications, splenic rupture, upper airway obstruction, acute myocarditis and mucosal oedema (which can increase the risk of bleeding) [10]. Splenic rupture is the most common potentially fatal complication of IM. It has been estimated to occur in 0.1-0.5% of all cases [2,11], mostly between the 7th and the 28th day of illness [11] and occurring almost exclusively in males [1].

Case Report 1
A 44-year-old male painter who smokes and had unprotected sexual intercourse with an HIV and tuberculosis infected partner, was medicated the previous month with penicillin due to odynophagia.

He was admitted to the emergency department with a 3-day odynophagia and fever. He was observed by an otolaryngologist and was medicated with ceftriaxone, paracetamol and ibuprofen. Three days later he returned to the emergency department complaining of fever, malaise, diffuse abdominal pain, thoracic pain, lipothymia and sweating. He denied a previous history of abdominal trauma.

On examination, he was febrile (temperature=39°C), sweaty and pale, his blood pressure was 82/40 mmHg with a heart rate of 75 beats per minute. His abdomen was generally painful to palpation, especially in the left upper quadrant with tenderness and peritoneal sign.

Laboratory tests revealed a hemoglobin of 12.4 g/dl, white blood cell count of 14 x 10³/µl with lymphocytosis of 70% and an increase proportion of atypical lymphocytes, platelet count of
166x103/µl and a protein C reactive of 3,7mg/dl. An increase of the liver enzymes was noted with an aspartate aminotransferase (AST) level of 279U/l, an alanine aminotransferase (ALT) level of 271U/l, alkaline phosphatase level of 214 U/l, gamma-glutamyl transferase (GGT) level of 323U/l, total bilirubin level of 2.1mg/dl and a lactate dehydrogenase level (LDH) of 736mg/dl. The HIV serology was negative. Monospot test obtained at the time of admission was later confirmed as positive for infectious mononucleosis.

An abdominal computed tomography (CT) scan showed a moderate amount of free liquid in the abdominal cavity and poor definition of spleen contours (probably a hemoperitoneum due to splenic subcapsular hematoma rupture).

Due to hemodynamic instability and a hemoglobin drop of 3g/dl, he was subjected to an emergency laparotomy. Intra-operatively, there was a hemoperitoneum and a spontaneous splenic rupture involving the splenic hilum. A splenectomy was performed.

The postoperative period was uneventful, with a clinical and laboratory improvement. The serology for B and C Hepatitis Virus and Cytomegalovirus was negative. He had a positive IgM and a negative IgG for Epstein Barr Virus. The discharge was on the 7th postoperative day and he did the vaccine for the capsulated microorganism after a complete recovery from the infectious disease.
Case Report 2

A 26-year-old male smoker with sporadic alcoholic habits was admitted to the emergency department with a 10 day history of headaches and myalgia and odynophagia started on the day of admission. He was medicated with clarithromycin and anti-inflammatory. Three days later he returned to the emergency department complaining of malaise and epigastric abdominal pain. He was treated symptomatically. Five days later he turned up in the emergency department with the same complaints accompanied with worsening abdominal pain, nausea, vomiting, jaundice, diarrhea and eructation.

On examination, he was afebrile and pale. His blood pressure was 91/60 mmHg with a heart rate of 137 beats per minute. His tonsils were hyperemic and his abdomen was distended and painful to deep palpation in the epigastric and left upper quadrant.

Laboratory tests revealed a hemoglobin of 14.2 g/dl, white blood cell count of 24.2x 10^3/µl with lymphocytosis of 53%, platelet count of 208x10^3/µl and normal renal function. An increase of the liver enzymes was detected with an aspartate aminotransferase (AST) level of 213U/l, an alanine aminotransferase (ALT) level of 492U/l, alkaline phosphatase level of 214 U/l, gamma-glutamyl transferase (GGT) level of 112U/l, total bilirubin level of 2.1mg/dl and a lactate dehydrogenase level of 524mg/dl.

An abdominal ultrasound was performed which revealed a splenomegaly with 15.3cm and a heterogeneous parenchyma with a hyperechoic area. Under observation in the emergency department, there was a hemoglobin drop to 8.6g/dl with hemodynamic instability. An emergency laparotomy was made. Intra-operatively, there was hemoperitoneum and a spontaneous splenic rupture involving the splenic hilum. A splenectomy was performed and 4 units of red blood cells were transfused.

The postoperative period was uneventful, with a clinical and laboratory improvement. The serology for Cytomegalovirus was negative. He had a positive Monotest for the Epstein Barr virus.
Virus. The discharge was on the 7th postoperative day and he did the vaccine for the capsulated microorganism after a complete recovery from the infectious disease. The histologic specimen revealed hyperplasia of red pulp of the spleen with histiocytic cells which suggested an infectious process.

Discussion

Spontaneous splenic rupture (SSR) due to IM is rare. Patients usually complain of abdominal pain and the presence of Kehr’s sign may be present in 50 percent of the cases of SSR. Signs of hypovolemia, such as tachycardia, hypotension, oliguria and pallor, may also be present [2, 12]. Splenic involvement in IM is common. Histologically, Epstein-Barr infection leads to lymphocytic and mononuclear infiltration of splenic parenchyma, causing distortion of normal tissue anatomy [11] which may end up in splenomegaly and fragility. In splenomegaly the spleen loses some protection through the rib cage [13] and this infiltration and fragility might contribute to splenic capsule fragmentation [11].

The mechanisms that may induce SSR include an acute increase in portal venous pressure from a Valsava-like manoeuvre, contributing to engorgement of the already enlarged and fragile spleen; capsule rupture due to sudden compression from diaphragmatic or abdominal wall contraction [11]; and anticoagulants, fibrinolytics and anti-platelet agents [5,9,10]. Some authors have suggested that the splenic architecture takes time to be altered, for this reason the splenic rupture is more frequent during the third week of disease [13], as occurred in the second case report.

However, Asgari et al. describe the time of highest vulnerability between 10-21 days [2]. Early rupture is unusual, but has been reported as early as 3 days following the onset of symptoms [2], as seen in our first case report.

The standard treatment was splenectomy. In hemodynamically unstable patients, emergency splenectomy continues to be the gold standard. In case of patients who are stable hemodynamically and have minimal transfusion requirements, non-operative management are gaining popularity and have been described successfully [2,4], such as splenic embolization [7, 12], vigilance or somatostatin infusion [11]. The advantages of non-operative management are the avoidance of post-splenectomy sepsis and the avoidance of postoperative complications [2].

In spite of the advantages, the non-operative management may carry some risks which include acute and ongoing (or delayed) haemorrhage, blood transfusions, extended hospitalization and long-term activity restriction [11].
Conclusion

Spontaneous splenic rupture is a rare and life-threatening complication of IM and it occurs mainly between day 10 to 21. The diagnosis may the challenging due to a variety of clinical manifestations and when it happens early, within the onset of disease. It can be manifested as an acute abdomen in hypotensive and tachycardic patients with previous history compatible with IM. An early diagnosis and treatment are crucial.

References: