Case report

Unusual solution to abdominal pain with contrast enhanced ultrasound

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SUMMARY

Splenic infarction might be the symptom onset of an important underlying disease. The possibility of splenic infarction must be inserted into differential diagnosis in all those patients who have pain in the upper left quadrant and/or on the left flank. When faced with a case of splenic infarction in a patient who has flown or climbed to high altitudes, it is appropriate to consider the possibility of an haemoglobinopathy. The diagnosis is far from being obvious for emergency physicians. For these reasons, it is very important to proceed as a multidisciplinary team with appropriate diagnostic examinations. The European Guidelines for non-hepatic applications of contrast enhanced ultrasound suggest the usage of this tool for investigation of suspected ischaemic lesions of the spleen.

BACKGROUND

Abdominal pain is a common cause of emergency department admissions, representing 5%–10% of the cases. Acute abdominal pain can be the symptom of a large spectrum of conditions, from benign and self-limited diseases to surgical emergencies. Evaluating abdominal pain requires an approach that relies on the likelihood of disease, patient history, physical examination, laboratory tests and medical imaging. The location of pain is a useful starting point as it can guide further evaluation and diagnostic route.

CASE PRESENTATION

A 34-year-old man was admitted to the emergency department (ED) after a transcontinental flight from New York to Rome. During the flight he had developed left upper quadrant abdominal pain with nausea and vomiting. The patient had never experienced a similar episode before.

INVESTIGATIONS

An initial abdomen physical examination carried out in our ED revealed tenderness on palpation in the mid-epigastric quadrant and in the upper left quadrant without visceromegaly. Laboratory tests showed an increase of leucocytes, C reactive protein (CRP) and lactate dehydrogenase, with signs of haemolytic anaemia. There were no pathologies or permanent medications in his medical history; there were no reported allergies either.

The patient immediately underwent abdominal ultrasound. B-mode sonography showed suspect areas in the hilar side of the spleen with

inhomogeneous texture (figures 1 and 2). Considering the previous abdominal examination, we decided to perform a Splenic contrast enhanced ultrasound (CEUS)¹ with sulfur hexafluoride-based microbubble contrast. CEUS confirmed the presence of hypoechoic lesions with a well-demarcated, wedge-shaped border; such lesions were extended to splenic capsule (figures 3 and 4). These CEUS images seemed to be compatible with splenic infarction.

DIFFERENTIAL DIAGNOSIS

There are many causes of splenic infarction, among which the most important are: cardiac embolism, pancreatitis, splenic torsion, infections, portal hypertension, haemoglobinopathies (including sickle cell disease), chronic myelogenous leukemia (CML) and myelofibrosis. Actually, the likelihood of each of such causes depends mainly on the patient's age: in the first five decades of life, splenic infarction is often due to haematological problems; passed that age, its most frequent cause is cardiac embolism.² In addition to this, it is important to consider, in every phase of life, the role of infective diseases, in particular malaria and mononucleosis.³ In order to identify the cause of the splenic infarction of our patient, it was essential to collect both family and personal history in detail, in addition to completing diagnostic and laboratory tests. According to the clinical presentation we hypothesised that the main cause of splenic infarction could be haemoglobinopathy. Blood examinations were positive for the presence of pathological haemoglobin and we also found the presence of sickle cells (38%) in a peripheral scratch blood. These findings allowed us to diagnose the heterozygosity sickle-cell anaemia.

TREATMENT

The treatment of splenic infarction is mainly based on causative agent. Altitude-related episodes in patients with sickle cell disease are generally treated in a conservative way. Surgery is indicated only in the presence of relevant complications like bleeding or massive and multiple abscess. The keystones of non-operative therapy are hydration and analgesia with narcotics or non-steroidal anti-inflammatory drugs. Due to the peculiarity of this disorder, the role of antibiotics and antiplatelet agents have not been established yet.⁴ Hospital admission should be suggested to the patient in order to provide

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Figure 1 B-mode ultrasound shows suspected areas in the hilar side of the spleen.

monitoring and supportive treatment, in addition to correcting possible hypoxia and acidosis.

From the first day, our patient was treated with analgesics and hydration with normal saline. During hospitalisation no medical or surgical complication emerged. In a few days the symptoms regressed, and we could discharge the patient.

OUTCOME AND FOLLOW-UP

Abdominal pain in uncomplicated splenic infarction heals in 7–14 days.⁵ Our patient was discharged home with improving clinical symptoms and close follow-up. In these years no further recurrences of disease emerged. We recommended him ‘adequate hydration and in-flight oxygen supplementation’ as Gotlieb *et al* suggest in a 2012 paper for sickle cell trait (SCT) patient taking airplane.⁶ However, further studies are needed to strengthen this recommendation.

Discussion

In 1950, Sullivan documented the first case of splenic enlargement following an airplane flight in a patient with sicklaemia trait. However, it was only in 1954, when Cooley published a

Figure 2 Colour-doppler ultrasound of the spleen.

Figure 3 Difference between B-mode ultrasound and contrast enhanced ultrasound (CEUS). CEUS obviously improved baseline ultrasound performance.

report of six additional cases of splenic diseases, that a relationship was found between high altitude exposure and sickle cell trait.⁷ Later, many other cases of splenic infarction following altitude exposure in patients with undetected SCT were listed in scientific literature.^{8–10} From these empirical observations, we can argue that a quick recognition of this haematological condition plays an important role in the management of acute splenic infarction, for example, by avoiding surgery.¹¹

Sickle cell disease is an autosomal recessive disorder affecting 300 million people all over the world, with highest concentration in Africa and in the Mediterranean region due to improved resistance to malaria.¹² In Europe, haemoglobinopathies are growing in number due to migration flow and mobility.¹³ Moreover, statistical studies suggest that sickle cell disease is an increasing global health problem,¹⁴ also because of the world-wide development of transportations. This evidence should be brought to the attention of emergency physicians and radiologists.

Sickle cell disease is associated with multiple pathological complications including splenic syndrome.¹⁵ The term ‘splenic syndrome’ describes symptoms related to sequestration of red cells and vaso-occlusion by sickled cells. It has been theorised that

Figure 4 Contrast enhanced ultrasound shows a splenic parenchymal hypoechoic area and normal arterial circulation.

the quantity of haemoglobin S (HbS) predisposes SCT patients to splenic syndrome.¹⁶ The higher the HbS rate is, the higher the risk of splenic infarction becomes, especially if the patient is exposed to triggering factors like the low dose of oxygen tension that occurs with a change in altitude (mountain climbing or air flight). Indeed, dehydration and a low dose of oxygen tension increase red cells' rigidity and deformity, and these alterations predispose to microvascular occlusion.¹⁷

In the setting of acute abdomen, spleen diseases are highly underestimated. It is essential to know the advantages of applying different imaging techniques to reach a definitive diagnosis. Indeed, a late diagnosis of splenic emergencies may result in an increase of mortality in patients.¹⁸

As it is stated in a 2006 paper by Görg *et al*: "The poor correlation between B-mode sonographic appearances of the spleen and a specific pathologic feature causes difficulty in clinical diagnosis and often requires additional imaging procedures".¹⁹ In this sense, the introduction of sonographic contrast agents and the birth of CEUS have considerably increased diagnostic accuracy of B-mode ultrasound.

The European Guidelines for non-hepatic applications of CEUS suggest the use of this tool for investigation of suspected ischaemic lesions of the spleen. In particular, it can be used to better evaluate splenic parenchymal microcirculation because of

the spleen's tendency to retain contrast microbubbles.²⁰ Furthermore, CEUS is a cheap and easily repeatable examination, that can be executed quickly at the bedside and that does not expose the patient to ionising radiations. Due to its high diagnostic value for infarcts, future studies are required in order to demonstrate whether it is possible to avoid CT scan if CEUS has detected a splenic infarction.

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Patient's perspective

I had never experienced a similar abdominal pain before, and I was very afraid. It was only my second transcontinental flight.

Some relatives of mine have sickle cell trait, but I had never investigated this disease because I had never had health problems. I understood that it was essential for physicians to collect my family and personal history in detail. Thanks to this, they could understand my health problem.

Learning points

- Sickle cell disease is a disorder affecting 300 million people. Statistical studies suggest that sickle cell disease is an increasing global health problem,¹⁴ also because of the world-wide development of transportations.
- It is appropriate to consider the possibility of a haemoglobinopathy in case of abdominal pain due to splenic infarction in patients who have flown or climbed to high altitudes.
- It is very important to proceed as a multidisciplinary team with appropriate diagnostic examinations. Contrast enhanced ultrasound (CEUS) helps the investigation of suspected ischaemic lesions of the spleen.

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