CASE REPORT

Atraumatic splenic rupture secondary to Epstein-Barr virus infection

Jessica Barnwell,¹ Paramjeet Singh Deol²¹Chelsea and Westminster Hospital NHS Foundation Trust, London, UK²Chelsea and Westminster Hospital, London, UK**Correspondence to**Dr Paramjeet Singh Deol,
paramjeet.deol@chelwest.nhs.uk

Accepted 7 January 2017

SUMMARY

We present a case report of atraumatic splenic rupture secondary to Epstein-Barr virus (EBV) infection. A woman aged 36 years presented to a London teaching hospital's Accident and Emergency department with severe abdominal pain following a 6-day history of diarrhoea and vomiting, which had been under review by her GP. A CT scan demonstrated free intraperitoneal fluid and abnormal appearance of her spleen. Blood tests demonstrated EBV infection with positive serology and leucocytosis. She underwent a laparoscopic washout, which confirmed a subcapsular splenic haematoma that was initially managed conservatively. However, she subsequently re-presented with increasing pain and required an elective splenectomy. This case demonstrates the risk of splenic rupture following EBV infection, even in the absence of trauma, and highlights the importance of prompt diagnosis and appropriate counselling in patients with infectious mononucleosis.

BACKGROUND

This case demonstrates an atypical presentation of Epstein-Barr virus (EBV) infection and demonstrates one of the more serious complications of EBV infection, atraumatic splenic injury. It shows both approaches to management, conservative and surgical, and the importance of counselling and follow-up.

CASE PRESENTATION

A woman aged 36 years was brought in by ambulance to Accident and Emergency (A&E) following a 1-day history of sudden-onset severe abdominal pain radiating to her left shoulder tip, with reported light-headedness. She described a preceding 6-day history of diarrhoea and vomiting and a lump in her groin. She had also been suffering from intermittent fevers but was afebrile on admission. She had seen her GP for this and had been prescribed a course of oral antibiotics to treat a presumed infected subcutaneous cyst. There were no other symptoms on systemic enquiry, no infectious contacts, no abnormal foods, no significant travel history and denied pregnancy.

On admission to the emergency department, she was tachycardic at 137 bpm, normotensive BP 135/83 and afebrile. On examination, her abdomen was tense and tender. She described a pain severity of 10/10 despite 10 mg intravenous morphine with the ambulance service.

Her medical history included polycystic ovary disease with no significant family history of note.

INVESTIGATIONS

Blood tests demonstrated elevated liver function tests: ALT 186 IU/L, alkaline phosphatase 180 IU/L and white cell count of $10 \times 10^9/L$ with elevated lymphocyte count at $4.6 \times 10^9/L$. Urine β HCG was negative. Point-of-care ultrasound scan (USS) in the A&E department demonstrated free fluid within the abdomen. She went on to have a CT scan with contrast of the abdomen and pelvis.

DIFFERENTIAL DIAGNOSIS

We determined that a ruptured appendix or ovarian cyst was most likely in this age group.

TREATMENT

The patient was assessed by the A&E team and received analgesia and intravenous fluids. She was admitted under the surgical team with differential diagnoses at this point of a ruptured appendix or ovarian cyst. A CT scan of her abdomen and pelvis demonstrated high density-free fluid, likely consistent with haemoperitoneum, peritoneal and pelvic lymphadenopathy, and on consultant review of the images, an abnormal appearance of the spleen was noted, reported as soft tissue mass that could be a splenunculus (figure 1). She was taken for laparoscopy, which demonstrated blood in all four quadrants and a subcapsular splenic haematoma. They performed a washout but did not dissolve the splenic haematoma in case it was occluding a tear or bleed, and left a drain in situ.

Figure 1 Initial CT scan shows haemoperitoneum and perisplenic haematoma.

CrossMark

To cite: Barnwell J, Deol PS. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-218405

She was discussed with the haematology team, given that the CT scan demonstrated lymphadenopathy and there was no history of trauma as a precipitant for the injury. A full work up was performed for an underlying haematological or infectious cause. A CT chest and neck was performed which revealed multilevel subcentimetre cervical lymph nodes, but no intrathoracic lymphadenopathy. Blood film showed pleomorphic lymphocytosis, with a few atypical lymphocytes with large basophilic cytoplasm. EBV IgG and IgM were positive, CMV, HIV and hepatitis serology were negative. Autoimmune screens were also negative. She was diagnosed with EBV-associated infectious mononucleosis (IM) with secondary atraumatic splenic injury.

She was treated conservatively with analgesia and had an uncomplicated inpatient stay.

OUTCOME AND FOLLOW-UP

She was discharged when mobilising safely with analgesia. She re-presented 3 weeks later with recurrence of left upper quadrant and shoulder tip pain. She had been afebrile during this period and reported no other symptoms. Liver function tests and lymphocyte count were all within normal limits now. CT scan demonstrated an enlargement of her intracapsular bleed with no reported lymphadenopathy; an MRI demonstrated 35 mm of old haematoma, and 15 mm of new bleed. This was conservatively managed and she was discharged with outpatient follow-up under the surgical team. On review in clinic, her pain persisted and was limiting her movement and activities of daily life. She had repeated USS, which showed an increasing anechoic region around the spleen from 3.5 to 5 cm (figure 2). Given the increasing size of the haematoma and persistent symptoms, she was admitted for an elective open splenectomy and washout. Of interest, she was noted to have a small accessory spleen, which was preserved. There were no intraoperative complications and she was discharged with appropriate postsplenectomy antibiotic prophylaxis and vaccination regime with routine outpatient surgical follow-up. Twelve months on, the patient reports she has ongoing pain and that she is currently undergoing investigations for this.

Figure 2 Significantly increased subcapsular perisplenic haematoma.

DISCUSSION

Infectious mononucleosis is a common disease within the UK, classically presenting with a triad of lymphadenopathy, fever and pharyngitis. The most common causative agent is EBV accounting for 90% of cases; a herpes virus transmitted via pharyngeal secretions.¹ Most cases are self-limiting, managed in primary care with appropriate symptomatic advice. It can be associated with hepatosplenomegaly, jaundice and temporary derangement of liver function tests. In uncomplicated cases, a clinical diagnosis may be sufficient for confirmation of infection; the Hoagland criteria state that in clinically suspected infection with 50% lymphocytosis with 10% atypical, the diagnosis can be confirmed with the heterophile antibody test.² On serological testing, a positive anti-EBV viral capsid IgM and elevated IgG but negative viral nuclear antibodies would indicate active infection. Steroid therapy is not recommended, and while treatment with antiviral therapy such as aciclovir reduces viral shedding; there is no evidence to support its use for symptomatic control in immunocompetent patients.¹

Splenic injury, most commonly atraumatic, is a recognised complication of IM occurring in <0.5% and more commonly in men under 30 years of age.³ It classically presents with left upper quadrant pain radiating to the left shoulder tip, which can be elicited on palpation of the spleen, known as Kehr's sign.^{4 5} They may also present with haemodynamic compromise and anaemia. It is most common within 3 weeks of infection, although can occur later in the disease, and a recent review of published case reports described a mortality rate of 9%.³

Patient's perspective

- I was rushed into hospital after having severe pain for a few hours at home. It was a bizarre situation as at first no one quite knew what was wrong with me and after an ultrasound, it showed there was blood in my stomach. After being admitted a couple of days later, I had surgery to remove the blood and work out where it was coming from. It turned out from the spleen, which again caused confusion as I hadn't had an accident or trauma of any kind. After a week and loads of tests, it was discovered I had glandular fever. It is apparently very rare that the spleen ruptures with glandular fever but the doctors were happy that I could go home to recover and all would be OK. After just over a week at home, I went back to hospital with the pain again and after tests, it showed that the spleen was still bleeding. Rather than take out the spleen, the doctors decided to leave it to heal again and see what happened. After an ultrasound check-up, the haematoma had grown again. This is when I decided to get a special opinion and they advised me that I should have a splenectomy as soon as possible and spoke to my consultant. This was then scheduled straight away and the spleen and haematoma were removed.
- As a patient it was a very frightening and painful experience. For a long time, I was in a lot of pain and felt extremely ill. I appreciate the doctors wanting to keep my spleen for health reasons but feel that they waited maybe a little too long to take it out. A year on I am still getting some extreme pain in that area and am undergoing tests at the moment to try and find out the cause of this.

Learning points

- ▶ Infectious mononucleosis (IM) does not always present with the classical triad of pharyngitis, cervical lymphadenopathy and fever.
- ▶ The majority of cases of splenic injury are atraumatic, and so care should be taken to consider IM as a differential with patients presenting with left upper quadrant pain.
- ▶ In haemodynamically stable patients, non-operative management should be considered, including endovascular techniques where available.
- ▶ Appropriate counselling of those patients with suspected or confirmed IM is needed to reduce the risk of splenic injury.

Diagnosis requires appropriate imaging with contrast CT allowing staging of the injury. Staging can be performed using the American Association for the Surgery of Trauma (AAST) splenic injury scale; however, in unstable patients, focused USS could be used with prompt surgical intervention.^{4–6} It can be managed conservatively and surgically. Non-operative management is suggested in haemodynamically stable patients with low splenic injury scale grade, and there is increasing use of endovascular techniques such as splenic artery angiography with embolisation to allow preservation of the spleen.^{4–7–8} However, over half of them require surgical management and splenectomy. If a splenectomy is performed, appropriate advice, prophylactic antibiotic and immunisation regimes should be given.

It is important that patients with a suspected or confirmed diagnosis of IM are counselled to reduce the risk of splenic injury. While there is no international formal consensus on when patients are safe to return to normal activities, splenic injury has been reported up to 8 weeks postinfection. The American Association of Family Physicians (AAFP) and current NICE guidelines for management of IM recommend advising

patients to abstain from contact sports for at least 1 month following onset of infection, and until asymptomatic.^{9–10} They should be counselled to seek urgent medical attention if they develop abdominal pain or become systemically unwell. If they have demonstrated splenomegaly and wish to return to high impact sports, it has been suggested that an USS should be performed to ensure resolution prior to returning to sporting activities.³

Twitter Follow Paramjeet Deol @paramydeol

Contributors The conception and design of study was by JB. Acquisition of data was by JB. Analysis and interpretation were performed by PSD and JB. Draft of script was by JB and revision of script was by PSD.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Lennon P, Crotty M, Fenton JE. Infectious mononucleosis. *BMJ* 2015;350:h1825.
- 2 Hoagland RJ. Infectious mononucleosis. *Prim Care* 1975;2:295–307.
- 3 Bartlett A, Williams R, Hilton M. Splenic rupture in infectious mononucleosis: a systematic review of published case reports. *Injury* 2016;47:531–8.
- 4 Sergeant SR, Johnson SM, Ashurst J, *et al.* Epstein-Barr virus-associated atraumatic spleen laceration presenting with neck and shoulder pain. *Am J Case Rep* 2015;16:774–7.
- 5 Lowenfels AB. Kehr's sign—a neglected aid in rupture of the spleen. *N Engl J Med* 1966;274:1019.
- 6 AAST splenic injury scoring scale, USA. <http://www.aast.org/library/traumatools/injuryscoringscales.aspx#spleen> (accessed 30 Aug 2016).
- 7 Brichkov I, Cummings L, Fazylov R, *et al.* Nonoperative management of spontaneous splenic rupture in infectious mononucleosis: the role for emerging diagnostic and treatment modalities. *Am Surg* 2006;72:401–4.
- 8 Tinkoff G, Esposito TJ, Reed J, *et al.* American Association for the Surgery of Trauma Organ Injury Scale I: spleen, liver, and kidney, validation based on The National Trauma Data Bank. *J Am Coll Surg* 2008;207:646–55.
- 9 NICE guidelines for management of infectious mononucleosis, UK. <http://cks.nice.org.uk/glandular-fever-infectious-monomucleosis> (accessed 30 Aug 2016).
- 10 Ebell MH. Epstein-Barr virus infectious mononucleosis. *Am Fam Physician* 2004;70:1279–87.

Copyright 2017 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow