Successful multidisciplinary management of an asymptomatic gastrosplenic fistula in a diffuse large B cell lymphoma patient

Sarah Mansouri , Chadey Assane , Francois-Charles Malo

General Surgery, University of Sherbrooke Faculty of Medicine and Health Sciences, Sherbrooke, Quebec, Canada

Correspondence to
Dr Francois-Charles Malo; francois-charles.malo@usherbrooke.ca

Accepted 17 February 2022

SUMMARY

Gastrosplenic fistula is a rare and potentially fatal clinical entity unknown to most healthcare providers. Its diagnosis and management are challenging; and addressing it too late can have devastating consequences for patients. To increase awareness about this pathology, we hereby present a case of asymptomatic gastrosplenic fistula arising from a diffuse large B cell lymphoma in a 60-year-old Caucasian man with no significant medical history. The patient was successfully treated with open *en-bloc* splenectomy and partial gastrectomy. The patient was discharged from the hospital 3 days after the surgery. At 1-month postoperatively, the patient was asymptomatic and presented no complication of the surgery. He went on to finish six cycles of chemotherapy (R-EPOCH, rituximab, etoposide phosphate, prednisone, vincristine sulfate, cyclophosphamide, doxorubicin hydrochloride) and achieved complete metabolic response. At 2 years after the surgery, the patient remains asymptomatic and presents no sign of disease recurrence.

BACKGROUND

Gastrosplenic fistula (GSF) is a rare clinical entity. In the last 37 years, only 30 cases have been reported in the medical literature.[1] Most cases occur in the context of diffuse large B cell lymphoma (DLBCL), either at initial diagnosis or following chemotherapy. Most patients present with symptoms of unspecific abdominal pain and/or gastrointestinal bleeding.[2] This highly morbid and potentially fatal complication of DLBCL is frequently misdiagnosed and can lead to fatal outcomes. Therefore, we report here a rare case of asymptomatic GSF successfully managed surgically. To our knowledge, only one other case of asymptomatic GSF was described in the medical literature. The patient did not undergo surgical intervention because of personal preferences.[3]

This case report aims to educate on the diverse clinical presentations of GSF and offer a single-patient surgical management and a complete 1-year follow-up experience. Through this case presentation, we want to provide our experience to help guide decision-making and the lessons we have learnt.

CASE PRESENTATION

A 60-year-old Caucasian man with no significant medical history presented to his primary care doctor complaining of diffuse muscle pain and weight loss. A thorough physical examination and laboratory studies revealed a left upper quadrant tenderness to deep palpation, microcytic anaemia and elevated C-reactive protein. These unspecific findings and the rapid progression of symptoms of the patient warranted the primary care physician to order a CT scan of the thorax, abdomen and pelvis. It revealed an enlarged spleen and no significantly enlarged lymph nodes. Nonetheless, the diagnosis of cancer was evoked by the radiologist and the patient was quickly referred to internal medicine for a thorough investigation.

On referral to the internal medicine team in our facility, a [18F]fluoro-D-glucose (FDG) positron emission tomography and CT (PET-CT) scan were ordered. The imagery showed a prominent spleen (22 cm long axis) and an infracentimetric retroperitoneal lymph node with high FDG uptake located behind the left psoas. A fine-needle biopsy of the lymph node was performed under CT guidance by an interventional radiologist, which confirmed the pathological diagnosis of DLBCL. The patient was referred to the haemato-oncology team for treatment and was almost immediately started on a regiment of rituximab+dose-adjusted etoposide, prednisone, vincristine, cyclophosphamide and doxorubicin regimen[4] and Methotrexate.

INVESTIGATIONS

After completing two cycles of treatment, which the patient tolerated well and was completely asymptomatic from, a follow-up FDG-PET scan was ordered by the haemato-oncology team as it is the standard modality for the evaluation of response to therapy in our centre. It revealed an 18.8×14.8 cm spleen with a central necrotic hypometabolic and a hypodense mass of 13.2×10.7×11.3 cm (Standard unit value (SUV) 13.3) with a new subcapsular collection located at the inferior pole of the spleen (see figure 1). The cavitary mass seemed to contain debris and displayed air bubbles in its midst suggesting an abscess. The diagnosis of GSF was suggested on the radiological evidence of a communication between the necrotic mass and the gastric body through a 12 mm fistula (see figure 2).

General surgery was consulted for this patient. On initial evaluation, the patient was completely asymptomatic. He had no abdominal pain, no signs of infection and no history of upper or lower gastrointestinal bleeding. He was afebrile and presented no haemodynamic compromise. Laboratory studies identified a normocytic anaemia

To cite: Mansouri S, Assane C, Malo F-C. *BMJ Case Rep* 2022;**15**:e244259. doi:10.1136/bcr-2021-244259

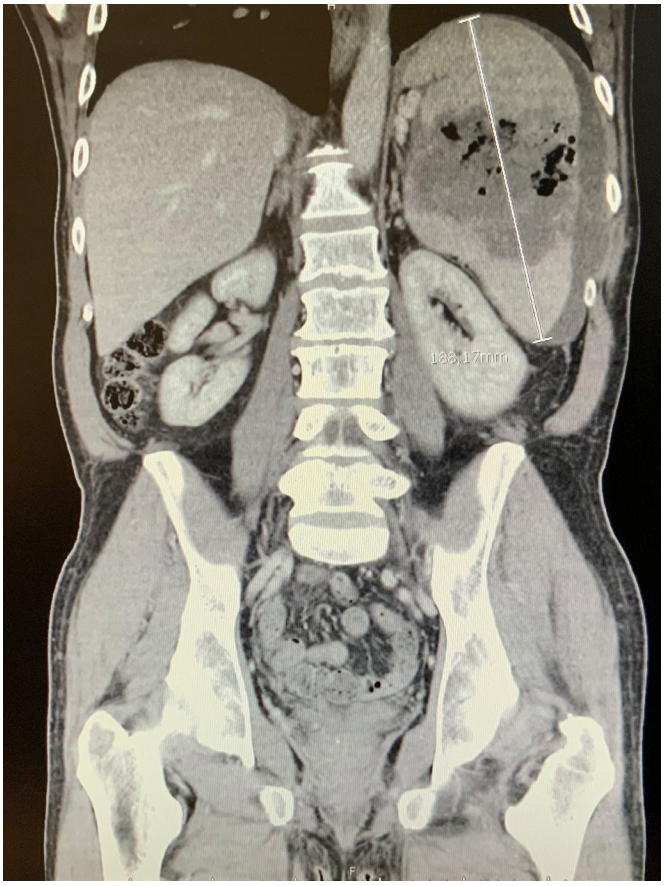

Figure 1 Coronal image of the abdomen on a CT scan with intravenous contrast—enlarged spleen with the gastrosplenic fistula outlined by the presence of air bubbles and hypodense material in the centre.

(haemoglobin 77 mg/dL), a normal white blood cell (5.4×10^9/L) and normal platelet count (149×10^9/L). Liver and renal function tests were normal. The C reactive protein was marginally raised (49,.3 mg/L). After a lengthy discussion with the surgery, radiology, gastroenterology and the haemato-oncology teams,

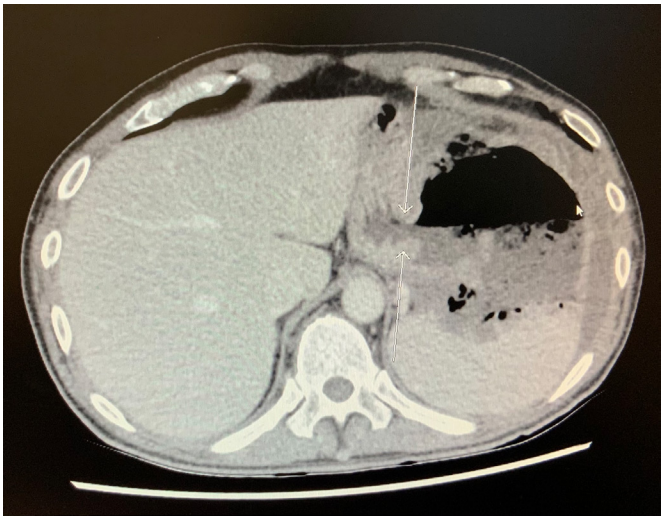

Figure 2 Axial image of the abdomen on a CT scan with intravenous contrast—fistulous tract between the stomach and the spleen (white arrows).

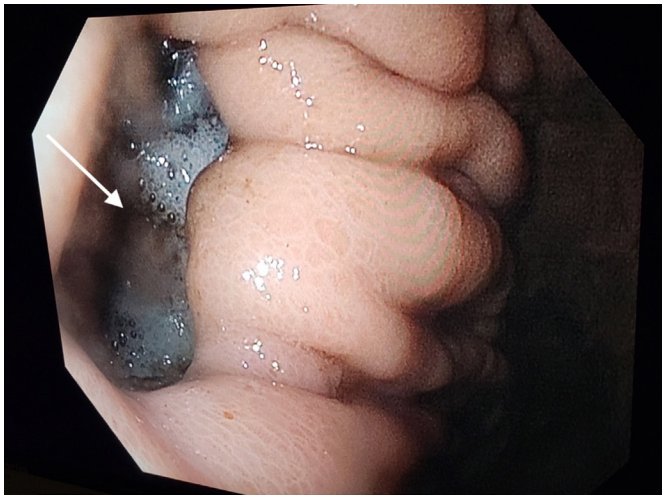

Figure 3 Endoscopic findings—gastroscopy done prior to surgery showed that the GSF (white arrow) was on the posterior wall of the fundus and at a safe distance from the gastro-oesophageal junction. Endoscopic imaging was also important to assess that there was no neoplastic involvement of the stomach itself and therefore plan accordingly for the proper type of resection. GSF, gastrosplenic fistula.

it was established that the best course of action was a surgical treatment. The different therapeutic options and risks, including conservative management, were presented to the patient. An informed consent was obtained for an open en-bloc splenectomy and partial gastrectomy. The surgery was scheduled for the next day. Presplenectomy vaccines were given prior to surgery despite the patient being under rituximab. An esophagogastroduodenoscopy was performed to inform the surgical plan. It showed that the GSF was at a safe distance from the gastro-oesophageal junction and there was no neoplastic involvement in the stomach itself (see figure 3).

DIFFERENTIAL DIAGNOSIS

The differential diagnosis included a splenic abscess given the radiological findings on the CT, a GSF or a necrotic cancerous mass.

TREATMENT

The patient underwent open splenectomy and a partial gastrectomy (en-bloc resection) under general anaesthesia. The procedure was well tolerated by the patient, and total blood loss evaluated at 1500 cc. One unit of packed red blood cells was administered. The primary closure of the fistula was not an option. The junction between the stomach and spleen where the fistula was lying was in a big inflammatory stump, rendering a safe dissection impossible, thus the partial gastrectomy (see figure 4).

OUTCOME AND FOLLOW-UP

After the surgery, the patient was transferred to the intensive care unit for surveillance. Overall, he presented a favourable evolution and was discharged from the hospital at postoperative day 3. He was vaccinated again 2 weeks after surgery at the outpatient clinic. At 1 month postoperatively, the patient was doing well and had no complication of the surgery. Chemotherapy (RDEAPOCH) was continued and completed after surgery for a total of six cycles. Complete metabolic response was confirmed by radiological imaging achieved 2 months following the surgery.

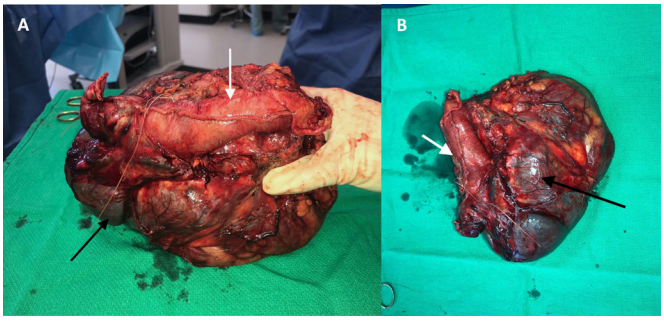

Figure 4 Voluminous surgical specimen containing the postero-lateral aspect of the stomach (white arrow) and the enlarged spleen (black arrow) that are fused together rendering an *en-bloc* resection necessary. (A) Medio-lateral view. (B) anterior view.

Twenty-eight months after the surgery, a follow-up observation is ongoing on an outpatient basis. The patient is asymptomatic and there is no sign of disease recurrence on follow-up FDG-PET scan imaging.

DISCUSSION

GSF is a rare entity. It can arise from many pathologies including Crohn's disease, gastric adenocarcinomas, benign peptic ulcers, after a laparoscopic sleeve gastrectomy or a trauma. Most cases reported in the literature arise from splenic or gastric lymphomas, with DLBCL histopathological type being the most prevalent.[5] Most patients presented with symptoms. The most common reports being left upper quadrant abdominal pain, weakness, GI bleeding and fever.[6] Asymptomatic cases are rare and the only other case of asymptomatic GSF is a 57-year-old Indian man in whom a GSF was incidentally revealed on PET/CT following three cycles of chemotherapy for a DLBCL. The patient refused any surgical treatment and was later lost to follow-up.[3]

Lymphoma-induced GSF occurs either spontaneously by direct invasion of adjacent structures or after chemotherapy.[6] GSF following chemotherapy treatments, as is in our case, results from the rapid lysis of tumour cells that infiltrated the splenic and gastric walls.[7] As previously stated, there have been only 30 cases of GSF reported in the literature. Here, we report the 31st case and its particularity is that it is in an asymptomatic patient, an entity even more rare.

Due to the rarity of the condition, thus the scarcity of data, there are no guidelines on the best treatment approach for these patients. It is theorised by some that uncomplicated GSFs (non-perforated, no GI bleeding) could be safely treated solely with chemotherapy.[6] Chemotherapy alone has been tried in a total of four patients as a definitive management strategy.[6–9] Two out of four patients were either lost to follow-up or died after 2 months of treatment.

On the contrary, our patient did benefit from early surgical intervention as do most patients with GSF. Splenectomy and partial or radical gastrectomy are the most common interventions performed, and laparotomy is the most common approach used. However, there has been a reported case of successful laparoscopic surgery.[5] Laparotomy was chosen in our case as the surgical specimen was deemed to voluminous to be safely extracted laparoscopically and risk of rupture was too high to attempt a laparoscopic approach.

The decision to operate was taken after a lengthy discussion with the patient and the consultants. The risk of spontaneous rupture and fatal bleeding outweighed the operative risks (considered low in this patient) even if he was completely asymptomatic from his GSF.

One crucial aspect that needs to be addressed when consenting a patient for surgery is postoperative complications. It is even more important in the context of DLBCL as chemotherapy is the only curative treatment. Complications can cause delays in the initiation of systemic treatments or render completion of chemotherapy impossible leading to disease progression or recurrence.

Current guidelines recommend vaccination of patients against encapsulated bacteria 2 weeks prior to elective splenectomy and 2 weeks after surgery for urgent splenectomy. Most centres vaccinate patients undergoing urgent splenectomy prior to discharge as there is important loss to follow-up in this patient population.[10] Our institution usually follows these recommendations. However, since the authors had a 24-hour window before surgery, it was agreed with haemato-oncology and infectious disease teams our patient could benefit from receiving his vaccines a first time prior to surgery and again at discharge. The postoperative period was short and uneventful, and the patient was discharged on postoperative day 3. It seemed premature to immunise the patient at that time as evidence suggests vaccination is not effective before the 2-week mark.[10] Therefore, the patient was vaccinated again 2 weeks after surgery at the outpatient clinic.

Because of the potentially life-threatening complications such as upper GI bleeding following splenic rupture, sepsis, shock, etc, most authors recommend surgery as a definitive management modality. The authors of this case report strongly advocate for a personalised approach. Conservative management should be considered very carefully and reserved to patients who are not candidates to surgery, for either medical reasons or personal preferences. The decision is not one to be taken lightly and warrants a comprehensive and multidisciplinary approach tailored to the patient's comorbidities and the aetiology of their disease.

In conclusion, the rarity of this pathology combined with the lack of data highlights the challenges that clinicians and radiologists must face in diagnosing and managing GSFs. A high degree of suspicion, a multidisciplinary approach with oncologists, gastroenterologists, surgeons, radiologists, etc are essential to a successful management of this potentially fatal entity.

Learning points

- Gastrosplenic fistula is a rare entity. Most cases arise from gastric or splenic lymphoma.
- It can present with various symptoms including weight loss, abdominal or flank pain, gastrointestinal bleeding and infection. It can very rarely present without symptoms.
- A high degree of suspicion should be held for gastrosplenic fistula (GSF) with patients known for a gastric or splenic lymphoma.
- Early diagnosis and a multidisciplinary approach are key in the successful managing GSF before potentially fatal complications such as haemorrhage, sepsis, peritonitis, shock arise.
- The most common management of gastrosplenic fistula is surgery. Chemotherapy alone could be an acceptable alternative for certain patients but data to support this claim are lacking to this day.

Acknowledgements The authors would like to thank Mr Aulin Monette for his enthusiasm in collaborating with our team.

Contributors SM: conceptualisation, acquisition of data, writing—original draft, writing—review and editing. CA: conceptualisation, writing—original draft, writing—review and editing. F-CM: conceptualisation, writing—original draft, writing—review and editing. All authors have read and agreed to the published version of the manuscript.

Funding This study was funded by Université de Sherbrooke (Département de chirurgie).

Competing interests None declared.

Patient consent for publication Consent obtained directly from patient(s)

Provenance and peer review Not commissioned; externally peer reviewed.

Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

ORCID iDs
Sarah Mansouri http://orcid.org/0000-0002-0034-8088
Chadey Assane http://orcid.org/0000-0002-4180-1926
Francois-Charles Malo http://orcid.org/0000-0002-4876-7185

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