Ectopic splenic tissues mimicking gastro-intestinal stromal tumour in a patient after splenectomy for a giant epithelial cyst of spleen: A case report

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ARTICLE INFO

Article history:
Received 16 January 2015
Accepted 27 June 2015
Available online 10 July 2015

Keywords:
Splenic giant epithelial cyst
Splenectomy
GIST

ABSTRACT

INTRODUCTION: Ectopic splenic tissues left after a previous splenectomy can masquerade as a gastro-intestinal stromal tumour (GIST).

PRESENTATION OF CASE: Splenectomy was carried out for a 17-year-old girl with a giant epithelial cyst of spleen. Four years later, an upper endoscopy carried out for dyspepsia revealed two sub-mucosal lesions at the posterior wall of the gastric fundus. Computed tomography diagnosed a GIST. At operation, a dump-bell shaped extragastric mass was excised. Histology showed normal splenic tissues.

DISCUSSION: Giant epithelial cyst of spleen is rare. It is even rarer for ectopic splenic tissues left after splenectomy to masquerade as a GIST.

CONCLUSION: Ectopic splenic tissues should be included as a differential diagnosis in a patient who has a history of splenectomy presenting with a sub-mucosal gastric tumour.

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1. Introduction

Giant epithelial cyst of spleen is a rare condition and splenectomy is the treatment of choice [1]. The removal of spleen will stimulate compensatory hypertrophy of any accessory spleen, if present, to give rise to a suspicious mass on routine follow-up surveillance. Alternatively, part of the splenic pulp may detach from the specimen and subsequently implanted on an ectopic site, which then subsequently enlarges, creating diagnostic confusion. We report a young lady with ectopic splenic tissues at the posterior wall of the gastric fundus mimicking a gastro-intestinal stromal tumour (GIST) after total splenectomy for a giant epithelial splenic cyst.

2. Presentation of case

A 17-year-old girl presented with abdominal pain and physical examination revealed a large left upper quadrant mass. After laboratory and radiological investigations, the diagnosis was a giant epithelial splenic cyst. The serum carbohydrate antigen CA19.9 was 906 U/ml. The patient was vaccinated with meningococcal, pneumococcal and haemophilus influenza vaccines. Laparotomy was performed in 2007 and a giant cyst arising from the spleen was found (Fig. 1). Splenectomy was performed and histologic examination confirmed giant epithelial splenic cyst. CA 19.9 returned to normal after surgery.

Fig. 1. A giant epithelial cyst of spleen was removed by laparotomy in 2007.
She recovered uneventfully from the operation. In 2011, she presented with dyspepsia and upper endoscopy revealed two sub-mucosal lesions at the posterior wall of the gastric fundus. The appearance was compatible with gastro-intestinal stromal tumours (GIST). Computed tomography (CT) scan and positron emission tomography (PET) scan were performed and a dump-bell mass was shown on the posteromedial aspect of the gastric fundus, measuring $1.1 \times 0.9 \times 1.1$ cm and $1.9 \times 1.6 \times 1.7$ cm, respectively, in the 2 dump-bells. The lesion was closely abutted to the wall of the gastric fundus and corresponded with the endoscopic finding of a submucosal lesion (Fig. 2 and Fig. 3). PET-CT of the dump-bell gastric lesion showed normal metabolic activities (SUVmax 1.4 and 1.0, respectively). She was subsequently seen by a gastro-enterologist and endoscopic ultrasound showed the lesion to be submucosal, favouring the diagnosis of a GIST. However, biopsy of the lesion only revealed reactive gastritis.

She eventually came back to us and laparotomy was offered with the presumptive diagnosis of a GIST. The mass at the gastric fundus was found at operation to be extra-gastric in origin and it did not invade into the gastric wall. The mass was excised (Fig. 4) and histologic examination confirmed normal splenic tissues. She recovered uneventfully from the operation and was discharged home on day 10.

3. Discussion

Giant epithelial cyst of the spleen is rare [1–3]. Percutaneous drainage usually results in re-accumulation [4] and marsupIALIZATION is often ineffective [4]. Surgical removal, and in most cases, splenectomy, either open or laparoscopic [1–6], is the preferred treatment. Unlike operations for immune thrombocytopenic purpura (ITP), no attempt should be made to search for and remove any accessory spleen. However, compensatory hypertrophy of accessory spleen(s) after splenectomy can create diagnostic confusion, especially in this era when CT scan is performed commonly. Extensive studies have been conducted to identify the common sites of accessory spleens to facilitate complete removal of all splenic tissues for ITP [7,8]. The splenic hilum situated in the inferior third of the posteromedial compartment and anterolateral to the upper pole of left kidney is the commonest location of an accessory spleen [8]. On retrospective review, $^{99m}$Tc Technetium sulfur colloid scintigraphy can be performed to confirm the splenic nature of the lesion [9].

Splenosis is an acquired condition as a result of auto-transplantation of splenic cells after splenectomy [8]. The location may be highly variable but enlarged splenosis located at the gastric fundus mimicking a GIST or a submucosal tumour has been reported [10,11], and in one case even ended up in a partial gastrectomy, which is obviously futile and unnecessary [10]. In this case, we have rightly performed a laparotomy to ascertain the lesion being extra-gastric. Since preceding investigations favoured the diagnosis of a GIST, we had no choice but to remove the lesion for histologic confirmation.

The ligation of short gastric vessels while performing splenectomy may include some splenic pulps which may subsequently enlarge after splenectomy. This case explains the location of ectopic splenic tissue in this case. The incidence may be under-reported but with liberal indications for upper gastrointestinal (GI) endoscopy and CT scan, this lesion can create diagnostic confusion and it should be included as one of the differential diagnosis when a submucosal tumour or a GIST is diagnosed, especially in patients having previous splenectomy. $^{99m}$Tc Technetium sulfur colloid scintigraphy is a useful adjunct and should be considered in doubtful cases.
In conclusion, ectopic splenic tissues should be included as a differential diagnosis in a patient who has a history of splenectomy presenting with a sub-mucosal gastric tumour.

**Conflict of interest**

All the authors had no conflict of interest. This study was not supported by any grant.

**Sources of funding**

There is no financial support for this study.

**Consent**

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

**Author’s contribution**

Chung K.M.: Patient management, surgery, acquisition of data and drafting of the article. Lau S.H.Y.: Analysis and interpretation of data, editing the article. Lau W.Y.: Surgery, literature review, decision making and final revision of the article.

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