INTRODUCTION

Accessory spleens (AS) are frequent entities but, abdominal pain caused by their infarction is a very rare diagnosis; furthermore infarction caused by venous congestion and not arterial torsion is even rarer. We present the case of a 38-year-old female patient with a three-week history of abdominal pain and an abdominal CT scan revealing a 7 cm mass near the spleen and tail of pancreas with segmental portal hypertension. The patient had no hematologic disease. This mass was surgically resected, with a pathological diagnosis of an infarcted accessory spleen.

CASE

A 38-year-old female patient was admitted with a three-week history of left upper quadrant abdominal pain and anorexia. She had left subcostal tenderness on physical exam. Laboratory studies revealed normal WBCs, platelet count, creatinine, electrolytes, liver function tests, amylase, and lipase. Hemoglobin was 9.6 g/dl, and CRP 24 (normal < 3,19). Abdominal CT scan showed a 7 cm mass close to the infero-medial aspect of the spleen and to the pancreatic tail, with thrombosis of the distal splenic, superior mesenteric, and the portal veins; the proximal vein is patent and drained by multiple collaterals (Fig. 1).

A full coagulation profile and liver function tests were normal. The patient had no bleeding history. Because of persistent abdominal pain and the indeterminate nature of the lesion, the patient underwent a laparotomy which revealed a segmental portal hypertension, and a mass in surface contact with the splenic hilum, the tail of the pancreas, and the splenic flexure of the colon. An en bloc resection of the mass, tail of pancreas, left colonic flexure and spleen was performed with primary colonic anastomosis.

The patient had an uneventful postoperative course. Gross pathologic examination revealed a well circumscribed 7 cm rubbery mass, with a regular 5 mm-thick capsule, abutting the adjacent resected organs; cut surface showed homogeneous rubbery mahogany colored material (Fig. 2). Microscopic examination and special stains confirmed the diagnosis of infarcted accessory spleen. The patient was discharged home on the fifth postoperative day, totally asymptomatic.

DISCUSSION

Accessory spleens (AS), the most common congenital splenic anomaly, are present in 10% to 30% of individuals [1]. They represent failure of the splenic buds to fuse, with subsequent ectopic location during migration of the embryologic spleen. The most common location of AS is the splenic hilum. The pancreatic tail is the next favorable location. Other common locations include the gastrosplenic, gastrocolic and splenorenal ligaments, the omentum, and the small bowel mesentery. Rare locations include the kidneys and the testicles [2]. There have been reports localizing the accessory spleen intrathoracically [3], intracranially [4], and even in a transplanted pancreas [5]. AS are not pathologically important, unless when splenectomy is done for haematological disorders because, if not removed, they will grow and cause recurrent disease as they will function as the native spleen. Abdominal CT can establish the diagnosis by showing a mass with the same density and enhancement as the normal spleen [7], the MRI also shows a hypointense mass which has the same signal intensity as the normal spleen [7]. Angiography can show the blood supply from the main splenic artery or its branches and a homogeneous accumu-
Scintigraphy with different radionuclide agents (technetium 99m-sulfur colloid or technetium 99m-labeled heat-damaged red blood cells) is very specific for the detection and localization of accessory splenic tissue [8]. Recently, superparamagnetic iron oxide has been applied to confirm the ectopic splenic tissue [6]. AS seldom cause symptoms and usually are incidental findings. Very rarely, as in our case, they can undergo infarction and cause severe abdominal pain. Infarction in the majority of the reported cases is due to torsion of the AS; however, in our case there is no torsion, and the infarction is due to arterial stasis from the venous congestion. Venous congestion can be due to hematologic [10] or liver [11] disease. However, our patient had no bleeding history, and her coagulation profile and LFTs were totally normal; in addition, systemic diseases cause total portal hypertension. The partial venous hypertension, in our case, can be explained by the mass effect of the AS on the mesenteric and portal vessels, resulting in a portal hypertension where the compressed vessels by the AS are the only ones affected, and the hypertension is termed “segmental”.

CONCLUSION

Even though very rare, an infarcted AS is a differential to consider in the diagnosis of abdominal pain. It is managed via surgical resection and the outcome is excellent. Infarction can be due to arterial torsion or stasis caused by venous congestion. Venous congestion can be due to several causes, and to the best of our knowledge this is the first reported case to be due to segmental portal hypertension caused by the mass effect of the AS.

REFERENCES