Intrapancreatic Accessory Spleen: A Rare Cause of Recurrence of Immune Thrombocytopenic Purpura

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Abstract

A 44 years-old-woman was admitted to our clinic because of development of thrombocytopenia and skin purpura. Abdominal ultrasonography and Computed Tomography (CT) did not show spleen or liver disorder. First line treatment with orally steroids showed a transitory response, with thrombocytopenia relapse in few months. Then laparoscopic splenectomy was accomplished successfully. The patient developed relapse in platelet count after 45 days of procedure, requiring treatment with thrombopoietic drugs and new diagnosis procedures. Nuclear medicine scintigraphy using heat-damaged Tc99m-labeled red blood cells revealed two images of uptake in left subdiaphragmatic region. Then the patient underwent intraoperative gamma-probe guidance after injection of Tc99m-labeled denatured erythrocytes, through left subcostal laparotomy with resection of two accessory spleens, one of them inside the pancreatic tail. After this procedure, the platelet count remains stable with lower dose of steroid treatment.

Keywords: Laparoscopic splenectomy; Immune thrombocytopenic purpura; Accessory spleen; Intrapancreatic spleen

Introduction

Excluding trauma, Immune Thrombocytopenic Purpura (ITP) is the most common indication for splenectomy, being an attractive therapeutic option for those who fail to respond to 4 weeks to 6 weeks of medical therapy with steroids or other agents [1]. The prevalence of accessory spleen tissue is about 10-40% of autopsies [2], and does not require treatment unless it is associated with hematological diseases. In the context of immune thrombocytopenic purpura, accessory spleens can cause recurrent or persistent disease after splenectomy.

Materials and Methods

A 44 years-old-woman, with a medical history of primary biliary cirrhosis and asymptomatic HBV infection, was admitted to our clinic because of development of thrombocytopenia (platelet count of 19,000/microL) and skin purpura since 2013. The patient seemed to be well on physical examination, except for the isolated skin purpura in upper extremities abdomen without masses. Low platelet count with no other disorder in blood sample. Abdominal ultrasonography and Computed Tomography (CT) did not show spleen or liver disorder. First line treatment with orally steroids showed a transitory response in her platelet count, with thrombocytopenia relapse in few months. Then laparoscopic splenectomy (with four trocars placed in left subcostal region) was accomplished successfully without intraoperative evidence of accessory spleen (abdominal cavity was explored in order to search for the presence of accessory spleens in their most common locations) and the specimen was introduced into a retrieval bag (800 ml) for removal by morcellation.

After surgery, the patient showed a platelet increase in the first few weeks with subsequent relapse after 45 days of procedure, requiring treatment with thrombopoietic drugs (eltrombopag) in order to maintain platelet count. Nuclear medicine scintigraphy using heat-damaged Tc99m-labeled red blood cells revealed two images of uptake in left subdiaphragmatic region (Figure 1).

The patient underwent intraoperative gamma probe guidance surgery after injection of Tc99m-labeled denatured erythrocytes in early 2015, through left subcostal mini-laparotomy. Systematic scan of the left upper quadrant was performed, with care taken to angle the probe away from the liver so as to avoid interference. Higher activity was noted next to the tail of the pancreas where the exploration revealed a soft red mass embedded in the pancreatic tail (Figure 2) and another one in the retrocolic fatty tissue. Resection of two accessory spleens was completed. The histological study confirmed the presence of splenic tissue in both pieces. The patient did not present an immediate increase in platelet count, so thrombopoietic treatment was needed again for 3 months. Nowadays, with lower doses of steroids treatment, platelet count remains stable.

Discussion

The response rate of splenectomy in patients affected with ITP is around 70%. An estimated 15-30% of patient’s shows relapse after surgery, and at least one third of these relapse is due to accessory

Keywords: Accessory spleen; Intrapancreatic spleen; Laparoscopic splenectomy; Immune thrombocytopenic purpura; Accessory spleen; Intrapancreatic spleen

Figure 1: Gamma graphic study with red blood cells and Tc99m with image of abdominal computed tomography overlapped.

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Accessory spleen, although relatively rare, can cause recurrence of idiopathic thrombocytopenic purpura (ITP) if studies are suggestive of residual functional splenic tissue. In such cases, accessory spleen may be responsible for thrombocytopenia, and surgical removal can be beneficial.

In our case, mini-laparotomy was the selected approach due to the rare location of accessory spleen that we suspected in preoperative imaging studies. A mini-laparotomy in the left subcostal region allowed us to use the gamma probe to detect the splenic tissue and to remove it through the same incision. Based on literature and on our case, we recommend the excision of accessory spleen in case of recurrence in hematological disease, because a minimally invasive approach removal (laparoscopic or even mini-laparotomy with gamma probe guidance) is safe and can be beneficial to patients with recurrent ITP and documented accessory splenic tissue, although response usually is not durable and satisfactory.

References