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original article

Postsplenectomy recurrence of idiopathic thrombocitopenic purpura: role of laparoscopic splenectomy in the treatment of accessory spleen

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SUMMARY: **Postsplenectomy recurrence of idiopathic thrombocitopenic purpura: role of laparoscopic splenectomy in the treatment of accessory spleen.**

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Aim. *Idiopatic thrombocytopenic purpura (ITP) is the most common indication for splenectomy. The failure rate of surgery is about 8% and the failure rate after splenectomy is approximately 28% for all patients. When the presence of an accessory spleen is diagnosed, splenectomy is recommended. Laparoscopic approach is considered the first choice.*

Patients and methods. *At our Department, between July and November 2011 two patients underwent laparoscopic accessory splenectomy for recurrence of ITP. Both patients had a previously laparoscopic splenectomy. Preoperative Magnetic Resonance (MR) was performed in both the cases revealing the presence of an accessory spleen.*

Results. *The operative time was 105 and 100 minutes respectively. No perioperative complications occured. Hospital stay was four days in both cases. The first patient had a disease free period of two months; the second one of one month. Both patients restarted immunosuppressive therapy.*

Conclusions. *The relapse of thrombocytopenia post-splenectomy can be associated with the presence of an accessory spleen. The laparoscopic accessory splenectomy should be considered the first choice approach. Surgical accessory splenectomy allows a transitory remission of the disease.*

KEY WORDS: Laparoscopy - Splenectomy - Accessory spleen - Idiopatic thrombocytopenic purpura.

Introduction

Excluding trauma, ITP is the most common indication for splenectomy in Europe (1). Splenectomy is a therapeutic option for adults who failed the 4-6 weeks medical therapy with steroids or other agents, who require toxic medication dose to maintain the remission, or who relapse after an initial response to medical therapy (2). Splenectomy is an effective treatment for ITP. The failure rate of surgery is about 8% and the failure rate at 5 years after splenectomy is approximately 28% for all the patients (3). In these cases, the presence of an accessory spleen (AcS) should be demonstrated (4).

Case reports

Case 1

A 16 years-old Indian female patient affected by refractory ITP since she was 10 years old. The diagnosis of ITP was made in 2005 by the onset of purpura, headache and vomit associated with a platelet count of 3000/mmc and subdural bleeding demonstrated with Magnetic Resonance (MR).

She was treated with: intravenous immunoglobulin (IgIV 20g/IV/die for five days); steroids (Prednisone 25 mg/die for 4 days, decreased to the interruption after 2 months) from November 2005 to 2011; monoclonal autoantibodies anti-cd20 (May 2006); laparoscopic splenectomy (March 2007); Dapsone from May 2007 to April 2011; Romiplostin and Estrombopag (tTPO-mimetics molecules) from September 2009 to June 2010; Azathioprine (100mg/die) from 2010. The patient came again to our attention in June 2011 for a new episode of cerebral bleeding with illness, headache and vomit. At physical examination diffused petecchiae and ecchymosis and oral bleeding syndrome were present. The vital parameters were normal with blood pression (BP)

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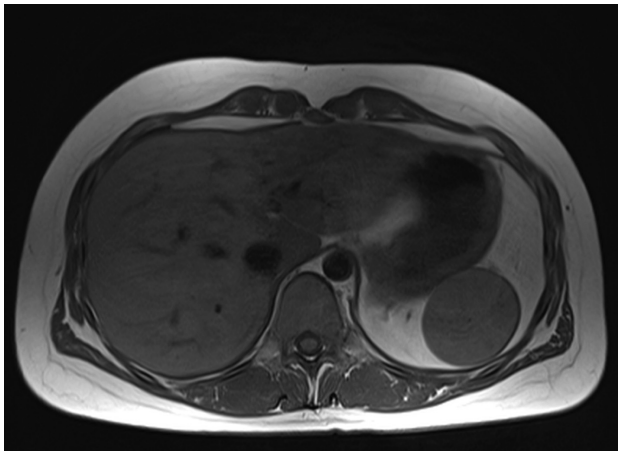

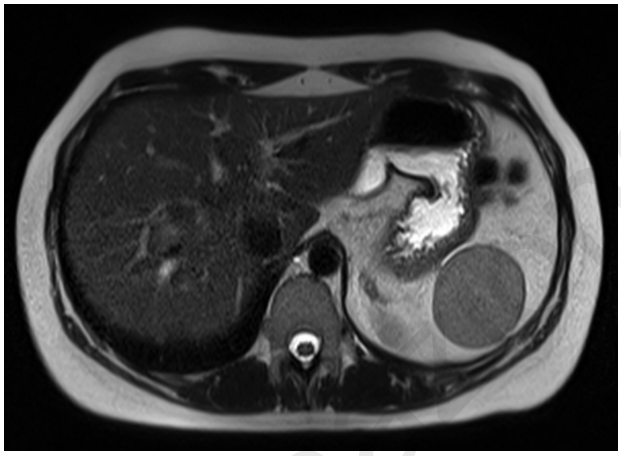

Figures 1 and 2 - A 5.5 cm accessory spleen showed with abdominal MRI. In these sections we can see the lesion near the gastric curve and the pancreatic tail.

of 135/80 mmHg, cardiac frequency (CF) of 80 bpm. The platelet count was 1500/mmc, the WBC was 13800/mmc, hemoglobin was 9.2 g/dL. The platelet count was in progressive decreased to 1000/mmc. Therapy with IgIV, Tranexamic Acid and Factor VII was started and the steroid was increased. Abdominal MR shows a 3.2 cm lesion in continuity with the distal extremity of pancreatic tail and near the cranial pole of left kidney suggestive for accessory spleen.

The laparoscopic accessory splenectomy (LAcS) was performed in July 2011. The perioperative platelet count was 61.000/mmc. The intraoperative setting confirmed the radiological data. The operation time was of 105 minutes without intraoperative complications. The postoperative course was regular. The blood tests demonstrated a progressive increase of the platelet count. The platelet count at the discharge was 74000/mmc. There was not markers of purpura at the physical examination or manifestations of bleeding. The platelet count made one week later was 127.000/mmc. Azathioprine was stopped and the steroid was progressive decreased. The hematological response was only transitory: in September 2011 therapy with Azathioprine was started again and a partial response was obtained so the Cyclosporine was started.

Case 2

A 27-years-old female affected by ITP since 20 years old. The diagnosis of ITP was made in 2003 with platelet count inferior to 100.000/mmc. She underwent the following therapies: steroids (Prednisone 25 mg/die for 4 days, with interruption for 2 months in 2003, 2006, 2008, and 2009); IgIV; monoclonal autoantibodies anti-cd20 from April 2004; laparoscopic splenectomy in July 2005; Dapsone from 2009 to 2011. The patient came again to our attention in 2011 when the piastrinopenia relapsed. Abdominal MRI shows a 5.5 cm lesion near the gastric curve and the pancreatic tail suggestive for accessory spleen (Figs. 1-3). Another similar 1.5 cm lesion sized was found in the mesenterial fat tissue. Splenic scintigraphy with ^{99m}Tc nanocolloid was performed and only the lesion in the left hypocondrium confirmed the suspicion of accessory spleen.

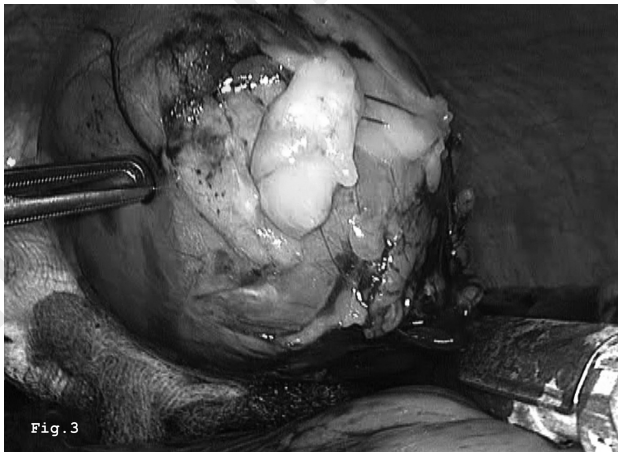

Fig. 3 - Laparoscopic approach confirm the lesion previously visualised with the abdominal MRI.

A LAcS was performed .The intraoperative setting confirmed the radiological data. The surgical time was 100 minutes without complications. The postoperative course was regular. The platelet count on first post-operative day was 154.000/mmc. She continues the therapy with Prednisone 25 mg until 2 weeks after the intervention. The blood tests made one month after the LAcS demonstrated a new relapse of piastrinopenia with a platelet count of 5000/mmc. The patient started the therapy with Prednisone 25 mg.

Surgical technique

The primary splenectomy was performed using the lateral position on right side and three trocars in both

cases. For laparoscopic accessory splenectomy the patients was placed in lateral position on right side. The trocar of Hasson was introduced according to the open technique. After camera introduction and pneumoperitoneum induction, two further trocars were inserted under vision: a 10-mm trocar in the left side and a 5 mm trocar in the left hypochondrium. Ligation of the vascular pedicle was obteined by Ultracision® and Endo-loop®.

Discussion

The AcS is the result of three possible mechanisms: (a) accessory spleen missed at the initial operation, (b) splenic tissue spilled at the initial operation with subsequent hypertrophy, and (c) compensatory hypertrophy of small splenic rest (5, 6). Lack of immediate and satisfactory post-splenectomy result or recurrence of thrombocytopenia are almost entirely connected with the remaining splenic tissue (7-13). Most authors, despite the use of different imaging techniques (ultrasonography, US; classic and spiral Computed Tomography, CT; and scintigraphy) underline the lack of reliable ways of preoperative assessment of ectopic splenic tissue (9, 10, 14, 15). Gigot et al. (9) studied CT scan results, demonstrating preoperative diagnosis of AcS, verified during laparoscopic splenectomy, only in 25% of patients. Morris, Szold and Targarona concluded that the key point should be the identification and the removal of the accessory splenic tissue during the primary surgery (7, 8, 15).

The recurrence of thrombocytopenia after splenectomy is an indication for scintigraphy (8, 9). Gigot et al. (9), in the postoperative study, revealed with scintigraphy the presence of remaining splenic tissue in nine patients (50%) previously operated for ITP. Interestingly, CT scan revealed the presence of the splenic tissue only in 5%. Despite that, only three patients (16%) manifested clinical thrombocytopenia (9).

Stanek et al. (16) report the recurrence of ITP in 9% of patients; in all cases the presence of splenic tissue was proved by scinthigraphy. Three additional accessory spleens were also identified and excised in three cases in which no preoperative data of their presence were present. Their conclusions were that preoperative imaging lacks in sensitivity and does not provide reliable evidence for the presence of accessory spleens.

In our experience 8 of 59 patients (13.6%) with ITP had an intraoperative diagnosis of accessory spleen. In these patients the accessory spleen was excised during the laparoscopic surgery. No one of these patients had a preoperative diagnosis of accessory spleen. Relapse of ITP was found in 2 of 59 patient (3.39%). In both these patient there was not a diagnosis of accessory spleen before and during the intervention and the accessory spleen was found with postoperative abdominal MRI.

The European Association of Endoscopic Surgery (EAES) has recommended a routine search for accessory spleens intraoperatively along with preoperative CT scan to achieve the highest detection rates and prevent disease recurrence for autoimmune haematological disorders (1). However the value of preoperative imaging to detect AcS remains unclear. Stanek et al. (16) found a level of sensitivity of the preoperative CT scan in the 43% of the cases. This lack of sensitivity can be caused by insufficient resolution of the imaging or by the missing of the small accessory organ most frequently located adjacent to the main spleen. In 58 patients operated for ITP there were three accessory spleens diagnosed preoperatively and confirmed intraoperatively. Three others were noted only intraoperatively, and one was diagnosed postoperatively. Therefore the incidence of accessory spleens reached 12.1%. In their study the recurrence of ITP was noted in three patients (5.17%). In all those cases the presence of splenic tissue was proved with scintigraphy. The features of the splenic tissue can be correlated to an accessory spleen only in one of these cases. In this case a laparoscopic splenectomy was performed and the presence of an accessory spleen was confirmed. They concluded that the preoperative imaging lacks sensitivity and does not provide reliable evidence for the presence of accessory spleens. The essential part of the laparoscopic splenectomy must be a thorough visual examination of the abdomen cavity with respect to most frequent sites of accessory spleens.

Napoli et al. and Gigot et al. (24,25) demonstrated 100% sensitivity in detecting accessory spleens with CT scan. Conal et al. (26) found an accessory spleen in 11 of the 58 patients (19%). However accessory spleens were confirmed in 9 patients intraoperatively. At laparoscopic splenectomy a total of 14 accessory spleen were found in 13 of 58 patients (22%). Four patients of this group had a negative preoperative CT scan. One accessory spleen was removed in 12 patients and two in 1 patient. All removed accessory spleen were confirmed at the histologic examination. Conal et al. (26) revealed a sensitivity of 60% for the detection of accessory spleens of the preoperative CT scan, whereas laparoscopy had a sensitivity of 93%. They found that laparoscopy is more effective in detecting or excluding the presence of accessory spleen than preoperative CT scan. In four patients an accessory spleen that the CT scan failed to reveal was found. It seems that the role of preoperative CT scan in detecting accessory spleens is of doubtful value if a meticulous laparoscopic approach is adopted at the time of splenectomy and for the following arguments. First, the majority of accessory spleens lie in the vicinity of the spleen and are readily detectable during laparoscopy. Sec-

ond the role of accessory spleens in haematological diseases remains controversial and the cause of relapse are multifactorial (9). Other sites for platelet destruction, such as the liver or residual splenic tissue (RST), may result in the recurrence of ITP (25). Third, the identification of accessory spleens postoperatively with imaging can be due to hypertrophy of the RST that would not have been detected on preoperative imaging (27). Their experience suggests that accessory spleens can be readily detected at laparoscopy near to the spleen and that preoperative CT scan specifically for their detection and localization may be not necessary. A search for accessory spleens during laparoscopy is mandatory and remains the most successful method for their detection.

Abdulmalik et al. affirmed that perioperative localization studies helps in the intraoperative identification of an accessory spleen (28).

In our experience 59 patients with ITP had a preoperative US. No one of those patient had a preoperative diagnosis of accessory spleen. We found accessory spleen in 8 patients (13.6%) during the surgical intervention. The accessory spleen was excided.

During the follow-up ITP relapsed in two patients respectively after 4 and 6 years from the laparoscopic splenectomy. No one of those patients had a preoperative or intraoperative diagnosis of accessory spleen. A magnetic resonance was performed in both these patients and an accessory spleen was detected near the pancreatic tail. In one of those patients the MR detected another lesion that was similar to an accessory spleen and located in the mesenterial fat tissue. A scintigraphy with ^{99}mTc nanocolloid was performed and it confirmed the presence of only one accessory spleen (near the pancreatic tail).

A review of the literature confirms our experience that LAcS is technically feasible. Szold et al. (7) reported successful laparoscopic accessory splenectomy in 8 of 8 patients. All patients have been discharged on the first postoperative day without complications. Other studies support these results. Morris et al. (8) reported no conversion to open surgery and no complications with all the discharged patients on postoperative day one. Yong et al. (23) reported a laparoscopic accessory splenectomy performed without complications; the patient was discharged on the second postoperative. Therefore seems that the laparoscopic resection of residual accessory splenic tissue is favored in patients with ITP. In our experience the laparoscopic accessory splenectomy was performed without complications. No technical difficulties wer met. The postoperative course was regular in both of these cases. The feeding was resumed on the third day post-operative and the patients were discharged on fourth postoperative day.

At last we discuss about the recurrence of ITP after the LAcS and the real efficacy of the surgery in the treatment of this disease. Therefore ITP is the most common indication for elective splenectomy, unfortunately, even when surgery successfully achieves platelets count remission, there is not guarantee that the disease will not recur. A review of the literature shows ITP recurrence following splenectomy ranging between 18% to 38% (17-9). In a significant number of these patients, residual accessory splenic tissue can be found. Targarona et al. (20) reported the findings of an accessory spleen in 33% of the patient who had not clinical improvement after laparoscopic splenectomy for treatment of ITP. An association between the presence of missed accessory splenic tissue and recurrent or unresolved ITP after initial splenectomy has been reported (9-20).

When the accessory spleen is diagnosed, the AcS is considered a potential ethiology and is usually excised as its removal may result in a second remission of the disease (9). A small number of case series and reports exist in the literature on LAcS. The literature provides a wide disparity in the efficacy of repeat surgery. After splenectomy the succesful response has been reported in 26.7% to 75% (7, 8, 21). Although a complete response following accessory splenectomy may not be possible, most patients can realize some benefits from the intervention because of the decreased requirement for systemic immunosuppressive therapies. As for our experience, the patients with relapsed thrombocytopenia obtained a rapid increased of the platelet count after some weeks and started again medical therapy. LAcS has been shown to be effective and safe (1, 22).

Conclusions

The imaging before the laparoscopic splenectomy is not sensitive for the detection of accessory spleens. During the laparoscopic surgery is recommended the research of accessory spleens. The relapse of thrombocytopenia can be associated to the presence of an accessory spleen and its excision allows a transitory remission of the disease. The LAcS is safe and feasible. Surgery obtaines only a transitory response of thrombocytopenia.

The preoperative US has no efficacy for the detection of AcS. The laparoscopy had a higher sensitivity for the individuation of AcS. At the relapse of ITP the MRI is a sensitive techniques to detect AcS, but the scintigraphy with nanocolloid is more specific.

The role of accessory spleen in the recurrence of ITP is not clear and their excision does not produce a durable response. Therefore more study with larger numbers of patients are needed to establish more about the effectiveness of laparoscopic accessory splenectomy for recurrent or unresolved ITP and to underscore the importance of perioperative localization studies or early diagnosis at the recurrences.

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