The surgical paliative treatment of myelofibrosis
Personal experience

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We report a case of significant splenomegaly by myelofibrosis, treated in our institution, that we believe relevant to the completeness parade of symptoms and the exceptional size.

KEY WORD: Myelofibrosis, Splenomegaly.

Introduction

The myelofibrosis is a chronic myeloproliferative disease, as the essential trombocitemia and polycythemia vera, described in literature for the first time by Heuck in 1879 [1], characterized by the release of circulating immature myeloid precursors that tend to accumulate in the reticuloendotheliale system [2,3]. This process, called extramedullary haemopoiesis, leads to secondary complications, such as hepatosplenomegaly. In particular, it becomes a cause of symptoms characterized by mechanical obstruction, such as early satiety, difficulty breathing and pain, and from haematological disorders such as anaemia, thrombocytopenia and, more rarely, leukopenia. In the case of massive splenomegaly, a sector portal hypertension, caused by compression and subsequent obstruction of the flow in the portosplenomesentheric system, is associated. We report a case of significant splenomegaly by myelofibrosis, treated in our institution, that we believe relevant to the completeness parade of symptoms and the exceptional size.

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Case report

A male 64 years old came to our observation for a consultation requested by internists for the presence of a massive splenomegaly. He had been hospitalized in the department of internal medicine sent from the emergency ward, where the patient had gone for a few episodes of melena during the week and the outbreak of asthenia, weight loss and dyspnea. Medical history showed the presence of a myelofibrosis since 1992, treated with cycles oncosorbide, diabetes mellitus type 2 under insulin therapy, and the gallbladder lithiasis. The patient was forced to bed in clynorthostatic position in order to compensate dyspnea, because the pathological spleen occupied the entire left emiabdomen (Fig. 1). This anatomical condition caused gastrointestinal-related synthomatology, characterized by nausea, early satiety, canalization alterations and dispeptic syndrome. The patient was bedridden also to prevent any likely, and deadly complications from rupture of the spleen, in the case of effort and even minor entity trauma.
The clinical examination showed pale and dried skin and mucousas, he had crepitating wheezings at the bases and the breath was harsh and widespread. The abdomen was globular and painfull, palpating we could appreciate a significant splenomegaly, that occuped entirely the left emiabdomen and spill over the midline. Blood tests showed a moderate anaemization (hb: 7.5 mg\dl) of normocytic normocromic type, increased indices of flogosys

and LDH. Emogas test showed hypoxaemia with hypocapnia. In the emergency room had performed an ultrasound which showed moderate hepatomegaly with an angioma of 10 mm of 6th segment, lithiasis of gallbladder and significant splenomegaly with multiple hyperechogenic parenchimal lesions. During the hospitalization in medicine he underwent to blood transfusion and enteral nutritional support with high protein rate. A gastroscopy showed the presence of esophageal varices in the distal thirdo of esophagus and a "ab extrinsic" compression of the duodenal bulb. The patient has undergone surgery through a laparotomy by a double undercostal access extended in median line. The vascular time was praticated by the ligation first of splenic artery and after of the splenic vein (diameter of about 30 mm), after isolation from the tail of the pancreas and exposure for approximately 8 cm. As we completed deafferentation of vascular hilum, we proceeded to splenectomy. The inspection of the gallbladder showed a chronic hepatic empiema abscessed in the bed, with an intense pericolecistitys that creates adhesions with gastric antro and cholic angle right. Then we proceeded to difficult cholecystectomy, after emptying the gallbladder of purulent content (about 300 cc) and suture of artery and cystic duct in a tough fibrous sclerotic gangue. Two drains in hepatic and splenic loggia. The weighting of fresh spleen was 7 kg. The histological examination on the operative piece documented widespread myeloid metaplasia of the red pulp with extensive areas of haemorrhagic necrosis.

The patient, after surgery, was taken into intensive care, given the high operative risk (ASA III). The postoperative course was without complications. in the day III the patient was channeled, has resumed normal diet and was mobilized first with aid and physiotherapy, and thereafter independently. in the day VI drains were removed and in the XVI patient was discharged. At diacharge, the patient was no longer dispnoyc, moved independently and adequately nutrished. Postoperative control after one month, the patient appeared in good general conditions, he had returned to drive the car and to have a normal life. Subjectively he felt satisfied with his current quality of life. After two months in good health condition, an intense headache appeared in the night with cold sweat, lipotimia and aphasia. He was transported to the Emergency department and underwent a skull TC, that showed a major cerebral hemorrhage from rupture of aneurysm. A craniotomy was performed to drain blood and was transferred to intensive care where he died in secomùnd postoperative day.

Discussion

The myelofibrosis is a haematological idiopathic disease whose treatment appears represented by allogeneic myeloablative transplantation [4], which, in about half of patients treated, enables the cell's establishment with durable remission of the disease. However, in view of high mortality for transplantation in elder patients and in advanced stage of disease, this method is a therapeutic choice only for patients diagnosed before 45 years and that is not feasible in this case we reported [5]. Other medical therapies as already seen, are always to be considered only supportive or palliative.

The role of surgery is now considered only a second-line procedure, implemented in the presence of a massive splenomegaly [6]. Splenectomy for myelofiborsis appeared for the first time in literature in 1937, published by Hickling [7] reported that the treatment of 25 patients with high perioperative mortality (56%). The indications for splenectomy are the presence of local symptoms (dyspnea, changes in digestion, pain), frequent transfusions [8], portal hypertension and risk of rupture, and then the correction of secondary symptoms, leading to an improvement of quality of life, but not preventing the progression of disease. Recently was published a retrospective study of the Mayo clinic reported 314 cases of splenectomy for myelofibrosis in the last 30 years; in this retrospective study particularly evident is the reduction of perioperative mortality by up to 10%, with

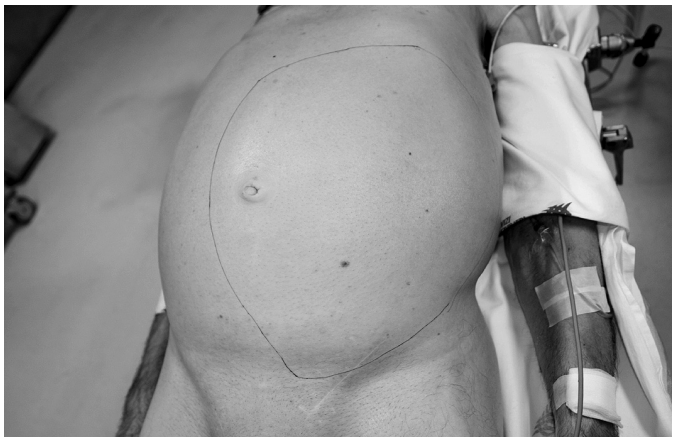

Fig. 1: The pathological spleen occupied the entire left emiabdomen.

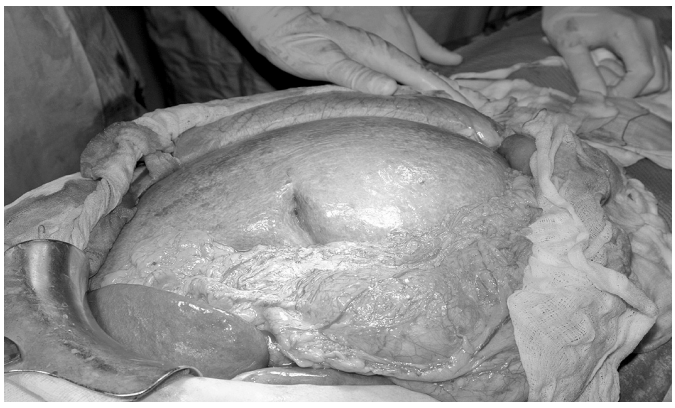

Fig. 2: Intraoperative view:the size of the liver were already increased.

significant improvement of symptoms of obstruction, of anemia and the thrombocytopenia of portal sector [9].
The surgical options in the treatment of myelofibrosis applied are exeresis of total and partial splenic resection. Currently both techniques can be performed by laparoscopy. In our case, the laparoscopic approach was impractical because of the extent of splenomegaly, which unables also the placement of only optical trocar. The emisplenectomy is used by some authors for the advantages it offers in terms of prevention of post splenectomy epathomegaly, at the cost of increased complexity of intervention and an increased risk of postsurgical haemorrhagia [10,11]. In our case we have not considered the emisplenectomia a valid therapeutic option because the size of the liver were already increased (Fig. 2) and also part of the exeresi would not have resolved the symptoms of encumbrance.
In dealing with the case came to our observation, we paid particular attention to the real need for a splenectomy. The presence of significant local symptoms that forced the patient to his confinement to bed and prevented them from proper nutrition, the detection of esophageal varices on gastroscopy posed a strong indication for surgery, on the other hand, the poor nutritional conditions and the precarious hematologic state were important risk factors for prognosis [12]. The preoperative approach has resulted in an improvement of clinical conditions, using the integration of nutrition and blood, to control the anemia. In accordance with an infettivologic counseling, antipneumococcic vaccines against Haemophilus influenzae type B and against meningococcal C were also charged in order to prevent OPSI [13].
Some authors stressed the importance of a few notes of surgical technique when splenectomy is performed in myelofibrosis. Access to abdomen must ensure the best exposure for a safe vascular isolation of the splenic ilum, reached by opening the gastrocolic ligament. In our case, more than bilateral subcostal incision to perform splenectomy and cholecystectomy, it was necessary to extend the incision in the xifopubic median line to control the entire spleen, whose inferior pole was located in correspondence of ala ossis ilii. The correct exposure of the operative field has allowed to execute the maneuver of isolation of the splenic vein from the lower margin of the pancreas. This procedure allows to prepare a log for a surgical clamping in emergency and will also enable us to identify and bind, as soon as possible, the same vein, in order to eliminate the risk of forming thrombus in the remaining stump, and to perfect binder of all vessels within the ilum [14,15].

Conclusions

The therapeutic process we follow confirms the palliative role of surgery in the treatment of myelofibrosis. Splenectomy does not alter the progression of disease, but significantly change the quality of life in patients, bringing them to resume their habits and autonomy, to the point of driving the car again. The lack of perioperative and postoperative morbidity is certainly due, in addition to the exemplary implementation of the operative technique, to the preparation of the patient includes the multivalent vaccination and restoration of a clinical sufficiently valid condition. For this reason, we recommend performing splenectomy, if possible, always in election, before increase of spleen size, thinning of capsule and parenchimatous infarcts lead organ to failure. It's obvious that a fracture of the spleen, in our case, was not devoid of complications, probably fatal, due to the difficulty of mastering the huge haemorrhage. Some authors recommend to bind splenic vein as distal as possible to prevent theombosys. This complication, reported in literature as 3.3%, does not justify in our opinion, the isolation of the vase from the bottom edge of the pancreas because of the risks of this maneuver, related to hypertension and the portal sector size of the vein, but has importance to us in ensuring a safety margin in the control of any bleeding and for this reason we always recommend the practice.
The patient's death, which occurred in the well-being three months later after surgery for massive hemorrhage from rupture of aneurysm vertebrobasialre, was totally unpredictable and not attributable either to disease or surgery and does not affect the validity of therapy followed by us.

Riassunto

Riportiamo il caso clinico di un'importante splenomegalia trattata nel nostro Istituto, che descriviamo sia per il quadro sintomatologico che determinava, che per l'eccezionale dimensione della milza stessa.

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