

Ectopic Spleen with Segmental Portal Hypertension, About a Case

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Abstract

Ectopic spleen is a condition in which the spleen does not sit in the left hypochondrium but has an atypical, often pelvic, location. We report the case of a 35-year-old patient suffering from chronic abdominal pain for 11 years associated with an episode of hematemesis, pollakiuria and dysuria, with anemia and notion of multiple transfusions. Abdominal ultrasound found an ectopic spleen in the pelvic position, enlarged in size, with multiple collateral venous circulations and extensive partial thrombosis of the splenic vein. A total splenectomy by midline laparotomy was performed. Perioperative exploration found a bulky pelvic spleen with a long and tortuous splenic pedicle, involuted in 3 turns without splenic infarction. When the splenic artery is clamped, the colonic and gastric varicose veins lose half their diameter, so the decision is made to perform total splenectomy. Post-splenectomy antibiotic prophylaxis and vaccination was administered. The follow-up was favorable, with a follow-up of 4 years. The patient was then lost sight of.

Keywords: Ectopic spleen, Portal Hypertension, Splenectomy.

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INTRODUCTION

Ectopic spleen is a condition in which the spleen is in a location other than the usual upper left quadrant, often lower in the abdomen or pelvis. This condition should be differentiated from accessory spleen or supernumerary spleen, where, in addition to the spleen present in its normal anatomical location, splenic tissue is found elsewhere [3]. Chronic volvulus of the ectopic spleen and its pedicle can lead, in rare cases, to the development of collateral venous circulation in the left gastroepiploic and short gastric veins and segmental portal hypertension. The objective of this work is to report a rare case of segmental portal hypertension in a pelvic ectopic spleen and to discuss it in the light of the literature.

CASE REPORT

We report the case of a pelvic ectopic spleen revealed by segmental portal hypertension, which was diagnosed and treated in the medicine gastroenterology department C and in the surgery department B of the Ibn Sina hospital in Rabat.

Our observation relates the case of Mrs. N. A aged 35, G3, P3, having as a history an iron deficiency anemia under replacement therapy for 11 years, a

uterine fibroid with menometrorrhagia and notion of multiple transfusions. She was admitted for chronic abdominal pain evolving for 11 years by paroxysmal painful crises of the left flank with torsion type, without irradiation and spontaneously resolving, associated with pollakiuria and dysuria, with notion of a single episode of hematemesis. for a year (not explored). On examination, the patient was in good general condition, hemodynamically stable with slightly discolored conjunctivae and a body mass index of 18 kg/m².

Abdominal ultrasound showed an ectopic pelvic spleen increased in size (15 cm) with collateral venous circulation, dilation of the splenic vein at 11.3 mm, tortuous, site of a thrombus and a small accessory spleen. The liver is of normal size, homogeneous, with regular contours. The trunk is 11.5 mm permeable, the supra-hepatic veins clearly visible. Abdominal CT angiography also showed ectopia of the spleen, at the pelvic level with rolled appearance of its pedicle and of the tail of the pancreas, partial and extensive thrombosis of the splenic vein, portal hypertension with portocaval diversion: recanalization of the umbilical vein, pericardial and perigastric varices as well as trans parietal gastric varices.



Figure 1: Axial computed tomography section with injection of contrast product, at the arterial phase highlighting the absence of the spleen in the HCG

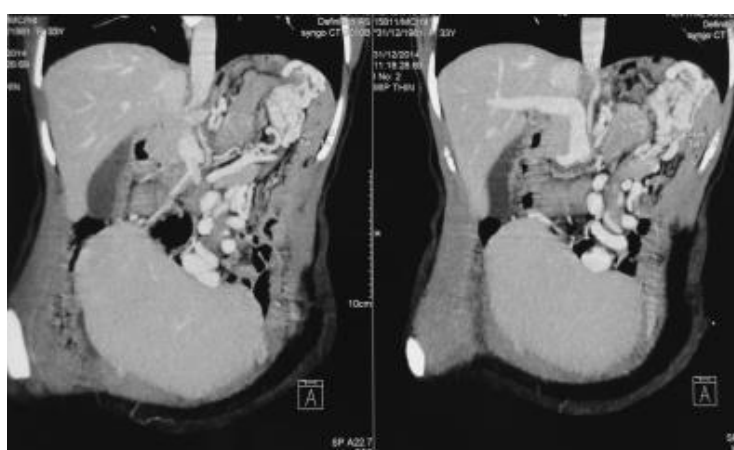


Figure 2: Abdomino-pelvic CT scan with injection of contrast product, in the portal phase: ectopic pelvic spleen; volvulus of the splenic pedicle in several turns of whorls; splenic vein thrombosis; trans gastric and perigastric varices

Esogastroduodenal fibroscopy showed gastric varices without red signs.

The biological assessment showed hypochromic microcytic anemia with a hemoglobin level of 6 g/dl, a ferritinemia of 5 ng/ml. The patient received a transfusion of 4 red blood cells. Other laboratory tests including liver tests and ionogram are normal.

The etiological assessment of portal hypertension finds at the liver biopsy puncture: hepatic suffering, no dysplasia, no fibrosis. The hepatic veins are thin, permeable to Doppler, partial thrombosis of the splenic vein, thrombophilic assessment: decrease in protein S activity to 27%, and on bone marrow biopsy: hyperplastic marrow with slight exaggeration of the

reticulin framework suggesting idiopathic myelofibrosis in its initial phase.

After a multidisciplinary consultation meeting, the patient was transferred to surgery department B on 02/20/2015 for a splenectomy.

Intraoperative exploration finds a large spleen in the pelvic position with a long and tortuous splenic pedicle passing in front of the mesocolon and the transverse colon, a large tortuous splenic vein, the splenic compartment is empty, occupied by small loops, multiple varicose veins pancreatic, transverse mesocolon, transverse colon and stomach. The splenic pedicle is volvulated 3 turns clockwise but without splenic infarction. When the splenic artery is clamped, the colonic, transverse and gastric veins and varices quickly collapse to more than 50% of their initial diameter. We decide to do a splenectomy.

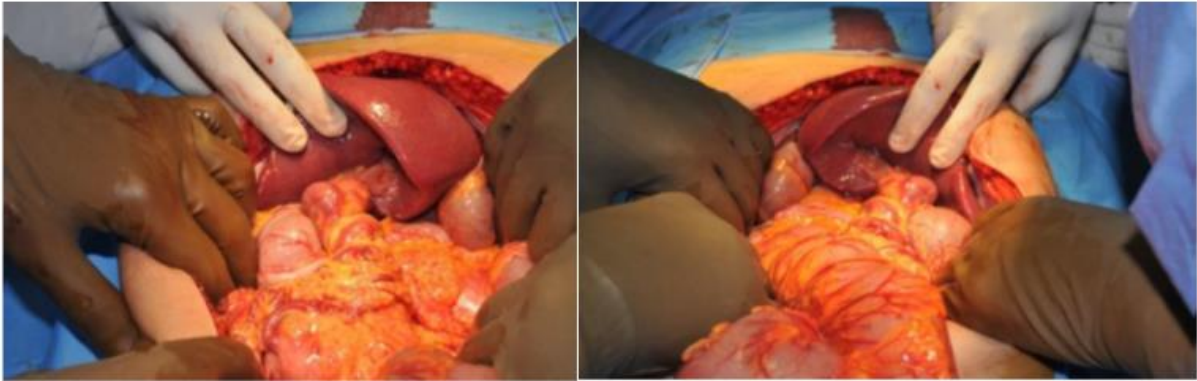


Figure 3: Intraoperative views showing a large spleen in the pelvic position with a long and tortuous splenic pedicle passing in front of the mesocolon and transverse colon

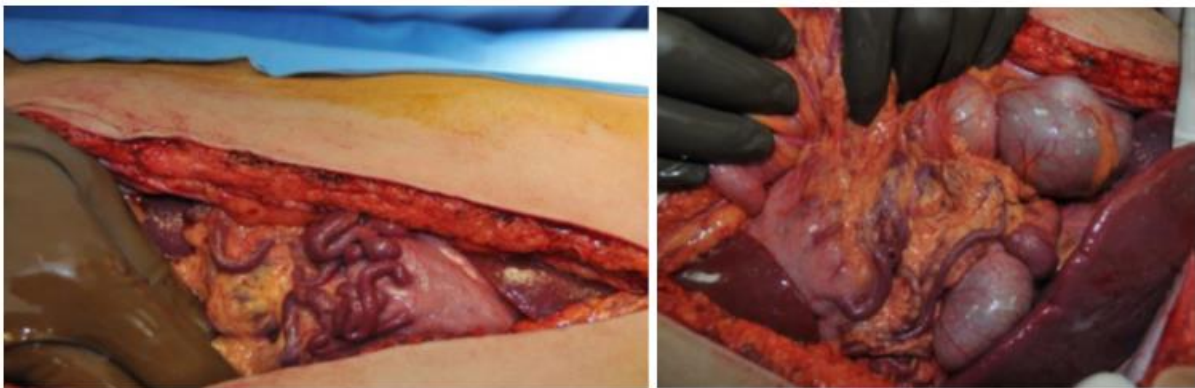


Figure 4: Intraoperative images showing the pelvic ectopic spleen with large varices of the transverse mesocolon

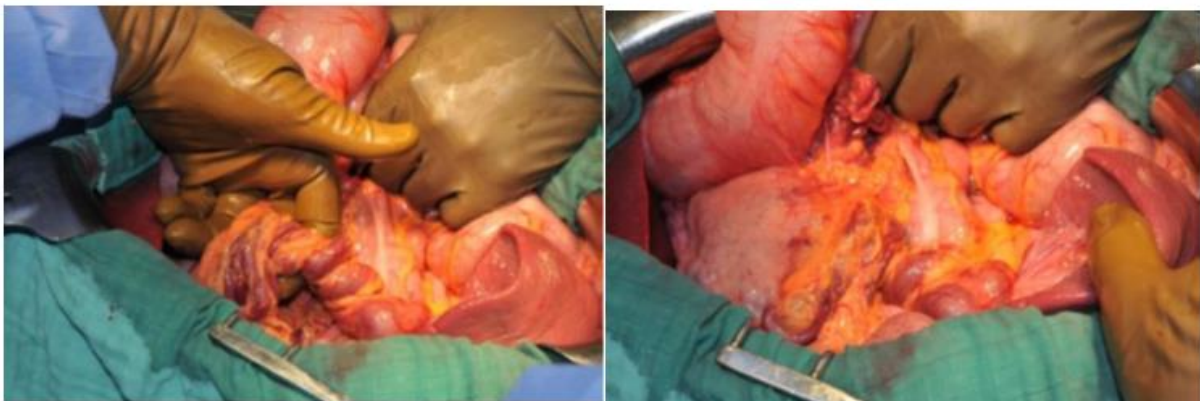


Figure 5: Intraoperative views showing the splenic pedicle volvulated 3 turns clockwise without splenic infarction

The postoperative course was simple. The patient received vaccination against pneumococcus and Haemophilus influenza and antibiotic prophylaxis based on Oracillin. The patient was discharged on D6 postoperatively.

The patient was seen in a gastroenterology consultation on 06/08/2015: in good general condition, without abdominal pain, exteriorized digestive bleeding or anemic syndrome, all suggesting a favorable evolution.

Histological examination revealed a congestive spleen and splenic vein thrombosis, with liver biopsy showing preserved morphology.

It is therefore a discovery of an ectopic spleen in the pelvic position in a 35-year-old multiparous patient suffering from iron deficiency anemia. She complained of an obstructive urinary syndrome, related to compression of the urinary tract by the ectopic spleen, associated with chronic and intermittent abdominal pain related to chronic torsion of the splenic pedicle responsible for portal hypertension with upper

gastrointestinal bleeding. remained hidden for a long time.

DISCUSSION

The ectopic spleen is an extremely rare pathology, its frequency varies from 0.2 to 0.5% [4].

The mode of revelation by an array of hemorrhage by HTP is exceptional. In the series of cases in the literature, we found only 2 cases whose mode of revelation is similar to our observation.

This pathology is observed at any age and in both sexes but is more frequent in women during the period of genital activity and in children.

The ectopic spleen is characterized by the disappearance of its means of fixation in its compartment, with an elongated pedicle and excessive mobility. The factors favoring its constitution are congenital or acquired.

The congenital anomaly is due to a defect in the development of the dorsal midgut responsible for hyper laxity of the suspensory ligament of the spleen or its total absence, which would deprive the spleen of any connection with the diaphragm in addition to an elongation of the pedicle. splenic (containing the splenic vessels and often the pancreatic tail [3]); associated with an almost constant absence of the gastro-splenic, spleno-colic and spleno-pancreatic ligaments [5, 6].

The pelvic position of the spleen would be explained by the fact that it adheres to the left gonad and that it would follow this one in its migration.

Acquired abnormalities favoring displacement of the spleen are [1, 5-12]:

- Hormonal changes during pregnancy in multiparous women;
- Splenomegaly and its causes such as leukemia, lymphomas, Hodgkin's disease and Gauche's disease;
- Laxity of the abdominal wall, gastric distension, the existence of a diaphragmatic hernia, operative ATCD and abdominal trauma;
- Infectious mononucleosis, malaria, hypertrophy or absence of the kidney and connectivities.

Our patient presents with a long splenic pedicle, an increase in the volume of the spleen with early idiopathic myelofibrosis and the notion of multiparity; From our case and from the literature, we find that often the two acquired and congenital anomalies are associated.

The clinical presentation of an ectopic spleen is variable and polymorphic:

- The fortuitous discovery of an abdominal mass by the patient himself, during a systematic abdominal examination or during an imaging assessment carried out for another indication.
- Intraoperative discovery during an intervention for another pathology.
- Intermittent abdominal pain testifying to torsion and spontaneous detorsion crises, associated with signs of peritoneal irritation or a digestive and/or urinary obstructive syndrome, this is the case of our patient
- Acute abdominal pain related to sudden and complete torsion of the splenic pedicle, or rupture of the spleen; often associated with nausea, vomiting and fever.
- A state of hemorrhagic shock by rupture of a congestive spleen
- An upper digestive hemorrhage exteriorized in the form of hematemesis or occult in the form of an anemic syndrome, this is the case of our patient, but it remains a particular mode of revelation.
- Acute recurrent pancreatitis by torsion of the tail of the pancreas

The majority of symptomatic forms are linked to complications that are varied. Torsion of the splenic pedicle remains the main complication, it usually occurs clockwise and is precipitated by body movements, changes in intra-abdominal pressure, peristalsis or distension of adjacent organs. It can be acute, subacute or chronic. When chronic splenic volvulus sets in, the splenic pedicle is coiled, the splenic artery is narrowed, and the splenic vein is compressed or obstructed. If the left gastro-splenic and gastro-epiploic veins are present and permeable, the venous return can borrow them and supply varicose veins of the large gastric tuberosity as well as the gastrosplenic and gastrocolic omentum. Which explains the segmental portal hypertension. The arterial supply is also compromised, causing infarction (generally, persistent torsion > 180° results in splenic infarction [8]), fibrosis and necrosis.

Other complications include abscess formation, peritonitis, local obstruction and necrosis of the tail of the pancreas, upper gastrointestinal bleeding and rupture of the spleen, intestinal obstruction and necrosis of the head of the pancreas, gastric obstruction, recurrent urinary tract infections and portal hypertension.

Abdomino-pelvic ultrasound confirms the diagnosis of ectopic spleen in view of the vacuity of the splenic compartment and the demonstration of an abdominal mass with very clear contours, well limited, with a homogeneous echo structure, finer than the liver, "comma" shaped with a central vascular hilum located

in the abdomen or pelvis. Coupled with Doppler, it can highlight signs of torsion of the splenic pedicle.

CT is the imaging modality of choice to diagnose an ectopic spleen, particularly when there is suspicion of torsion, by showing an empty splenic space with the presence of an abdominal and/or pelvic mass. In case of infarction, the CT sections show a slight decrease in the spontaneous density of the spleen compared to that of a normal splenic tissue, the absence of enhancement after injection of contrast product as well as an infiltration of the hilar fat. and sometimes necrosis of the tail of the pancreas. The twisted pedicle can be seen on the scanner in the form of a swirl representing the whorls.

In our patient, CT shows, in addition to torsion of the splenic pedicle, thrombosis of the splenic vein and portocaval shunts suggesting portal hypertension.

It makes it possible to look for the signs of gravity and to eliminate the differential diagnoses of an upper digestive hemorrhage, more particularly a rupture of the VOs secondary to PH on the liver of cirrhosis. Magnetic imaging examinations are little used in the study of the spleen.

Three therapeutic approaches are possible, depending on the age of the patient, the clinical presentation and the condition of the spleen: therapeutic abstention, conservative treatment, and radical treatment, with its advantages and disadvantages.

Therapeutic abstention requires rigorous clinical and radiological monitoring, rarely chosen in current practice.

Conservative treatment consists of splenopexy, which can only be considered if the spleen is ectopic without signs of necrosis or any other complication requiring splenectomy. It is associated with a 65% complication rate.

The radical treatment is the method usually chosen in the ectopic spleen, it is splenectomy by laparotomy or laparoscopic route. It is indicated in case of functional asplenia due to torsion, splenic necrosis, subcapsular hematoma, secondary hypersplenism, any suspicion of malignancy and venous thrombosis with segmental portal hypertension [1, 7] c is the case of our patient who presents with chronic ectopic splenic volvulus responsible for splenic vein thrombosis and therefore segmental PH with multiple varices, the difficulty of predicting permeabilization of the splenic vein for the collapsed gastric varices was the reason for using splenectomy instead of splenopexy. Early postoperative complications are dominated by infectious complications (scarring, pulmonary and intraperitoneal), followed by thromboembolic and hemorrhagic complications. It is estimated that the risk

of late complications after splenectomy is preponderant in the two years after the procedure. They are first of all of the infectious type (sepsis, pneumopathies and meningitis), with a mortality of 65% during sepsis and 78% for meningitis. There are also thrombotic complications, generally unrecognized spleno-portal thrombosis in the early postoperative period, responsible for portal hypertension. The evolution was favorable in our patient. It is estimated that the risk of late complications after splenectomy is preponderant in the two years after the procedure. They are first of all of the infectious type (sepsis, pneumopathies and meningitis), with a mortality of 65% during sepsis and 78% for meningitis. There are also thrombotic complications, generally unrecognized spleno-portal thrombosis in the early postoperative period, responsible for portal hypertension. The evolution was favorable in our patient. generally unrecognized spleno-portal thrombosis in early postoperative period, responsible for portal hypertension. The evolution was favorable in our patient. generally unrecognized spleno-portal thrombosis in early postoperative period, responsible for portal hypertension. The evolution was favorable in our patient.

Anti-infectious prophylaxis consists of pneumococcal vaccination in the first place, anti-HI and anti-meningococcal in children, antibiotic prophylaxis and rigorous clinical monitoring of any signs of infection and its early management. Our patient received double vaccination and antibiotic prophylaxis.

CONCLUSION

The pelvic ectopic spleen is a rare entity, its frequency varies from 0.2 to 0.5%, which can be of congenital or acquired origin, but often the two anomalies are associated. It is seen at all ages and in both sexes; with two frequency peaks, in childhood and in adulthood in women during the period of reproductive activity.

It remains of difficult clinical diagnosis because of its many varieties of presentation. Symptomatic forms are often linked to complications. Torsion of the splenic pedicle remains its main complication; it can be acute, subacute or chronic. The mode of revelation by an array of hemorrhage by PH is particular.

The diagnosis of a chronic volvulus of an ectopic spleen must be evoked in front of an array of

chronic and intermittent abdominal pain, and clinical signs such as a mobile abdominal mass and/or signs of pelvic compression; associated with a picture of upper digestive hemorrhage revealing a picture of portal hypertension.

Ultrasound and abdominopelvic CT are sufficient to establish the diagnosis, rule out differential diagnoses and indicate surgical treatment.

Splenectomy remains the safest way to treat gastric varices in segmental PH on an ectopic spleen. Laparoscopy should be preferred whenever possible, as it allows a rapid recovery period and a shorter hospital stay, not to mention the aesthetic benefit.

Data Availability Statement

The data that support the findings of this article are available from the corresponding author upon reasonable request.

Conflicts of Interest: Drs Jihane Sabar, Moustapha Traore and Abdellatif Settaf declare no conflict of interest.

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