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Laparoscopic Gastropexy for the Treatment of Wandering Spleen With or Without Gastric Volvulus

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1. Introduction

Wandering spleen is a rare condition. This congenital or acquired pathology is found in children and adults alike. It is characterized by a hypermobile spleen causing in some cases splenic torsion with ischemia.

We will successively look at the anatomy, etiologies, epidemiology, clinical pictures, additional imaging examinations and surgical possibilities for this pathology.

2. Anatomy

Wandering spleen is caused by failed fusion of the dorsal peritoneum, or absence or abnormal development of its suspensory ligaments that hold the spleen in its normal position in the left upper quadrant of the abdomen.

The splenic ligaments are the gastrosplenic, splenorenal (splenopancreatic), splenophrenic, splenocolic ligaments. (Couinaud, 1963)

Embryologically, the splenic ligaments develop in the coeliac artery territory, from the primitive dorsal mesentery (mesogastrium), which is responsible for the formation of peritoneum, the greater omentum and the several peritoneal folds. However, developmental anomalies or variations may take place. These variations in the embryologic development of the spleen's primary supporting ligaments could explain the wandering spleen.

These ligaments may be absent, may be too long or too short, too wide or too narrow, or abnormally fused.

3. Etiology

Wandering spleen can be a congenital or acquired condition.

3.1 Congenital form

Wandering spleen is in most cases a randomly distributed birth defect but in some cases it can be part of a syndrome.

Fig. 1. Transverse section. Development of peritoneal reflexions of spleen during primitive embryonic stage. Coeliac artery territory.

Fig. 2. Transverse section. Peritoneal reflexions of spleen are developed from dorsal mesogastrium (primitive dorsal mesentery).

Fig. 3. Frontal section showing the formation of splenocolic ligament and phrenicocolic ligament

3.1.1 Association

3.1.1.1 Congenital diaphragmatic hernia

The first case of wandering spleen associated to congenital diaphragmatic hernia (CDH) was described in the literature in 1940 by Bohrer. Several cases have been reported since then (Yasuda et al, 2010; Fiquet-François et al, 2010; Yilmaz et al, 2008; De Foer et al, 1994). The diagnosis of both pathologies can occur at the same time or the diagnosis of wandering spleen can be secondary to CDH. With CDH wandering spleen can be a result of an abnormal or absence of retroperitoneal fixation. Based on these data, all patients with CDH should be considered as potential candidates for wandering spleen.

3.1.1.2 Omphalocele

Yilmaz reported the unusual case of wandering spleen associated to omphalocele. (Yilmaz et al, 2008) As a possible cause for this association they listed defects on the abdominal walls through which the organs were protruding, resulting in a restriction of the stomach and spleen normal rotation or inefficient fusion after the rotation has been completed

3.1.2 Familial wandering spleen

Ben Ely described the first case of familial wandering spleen with two sisters diagnosed at a 3-year interval. (Ben Ely et al, 2008)

3.2 Acquired form

3.2.1 Postoperative (subtotal splenectomy)

Even if these data are not found in the literature, our multicenter study (Fiquet-François et al, 2010) reported 4 cases of wandering spleen post subtotal splenectomy. They were in fact excluded from the study that only focused on congenital forms. These cases are quite

interesting and probably unveil a technical defect. When the subtotal splenectomy involves resection of the upper pole of the spleen, with the section of suspensory ligaments, promoting acquired wandering spleen. To avoid this type of complications it is preferable to preserve the upper pole of the spleen and promote resection of the lower pole. It is important to bring up the possibility of wandering spleen in case of sudden or chronic abdominal pain in a patient having a history of subtotal splenectomy.

Fig. 4. Sagittal section showing the Phrenogastric ligament. This ligament prolongs the splenophrenic ligament to the right, and this splenophrenic ligament is an extension of the splenorenal ligament.

3.2.2 Traumatic diaphragmatic hernia

As discussed above, CDH can be associated to wandering spleen; in fact traumatic diaphragmatic hernia can also generate acquired wandering spleen.

3.2.3 Malarial infection

Malarial infection has not been clearly validated as responsible for the onset of secondary wandering spleen, but it can clearly trigger the pathology, asymptomatic until then.

Cripps described the case of a patient who had a malarial infection at the age of 5 and the CT-Scan done at the time validated a normally located spleen. (Cripps et al, 2010) However at the age of 18 she developed clinical symptoms and the diagnosis concluded to wandering spleen that could have resulted from a congenital fusion anomaly or attenuation of the patient's suspensory ligaments caused by her previous malarial infection and splenomegaly. However we can wonder if the malarial splenomegaly did not simply unveil an underlying congenital abnormality.

Fig. 5. Frontal view. Peritoneal attachments of spleen. Stomach is retracted to the right

4. Epidemiology

The incidence of wandering spleen is uncertain and difficult to assess. The diagnosis is often made following complications. The incidence of this pathology is probably dramatically underestimated.

Romero and Barksdale evaluated the peak incidence for wandering spleen between the age of 20 and 40 (Romero & Barksdale, 2003; Lin et al, 2005). Generally, 70–80% of the reported cases occur in women of childbearing age. (Steinberg et al, 2002) Hormonal changes and fluctuations explain this female predominance in adults. Furthermore the literature has reported that potentially predisposing elements in this population include multiparity and abdominal laxity thought to be secondary to pregnancy-induced hormonal effects on the abdominal wall. (S. Zarrintan et al, 2007) Ghazeeri et al (Ghazeeri et al, 2010) reported the case of splenic torsion on wandering spleen in a pregnant woman in her twelfth week of twin pregnancy.

This pathology is also found in children seemingly affecting more boys than girls (Allen & Andrews, 1989; François-Fiquet et al, 2009; Fiquet-François et al, 2010). This condition can occur very early on as seen in neonatal cases (Balliu et al, 2004; Fiquet-François et al, 2010, Arleo et al, 2010). During the first years of life the sex ratio is probably reversed. (Brown et al, 2003)

5. Clinical pictures

The diagnosis of wandering spleen is extremely difficult since it is such a rare condition and is clinically non-specific. In our recent multicenter study in children (Fiquet-François et al, 2010), we reported that the abdominal pain is at the forefront of all symptoms (93 % of cases), and its severity brings 86% of all cases to Emergency Room care. Furthermore, in 57% of all cases it was their first symptomatic episode of this type. The pain location is clinically non-specific: diffuse, periumbilical, left side, pelvis, left hypochondrium... Vomiting can be associated in 57% of cases. None of the diagnoses of wandering spleen were based on clinical evidence only. Even if the diagnosis cannot solely be based on clinical observations, it is important to note that the clinical presentation for wandering spleen can be either acute or chronic pain (Fiquet-François et al, 2010). The acute clinical pictures require emergency surgery because of the high risk of ischemia.

5.1 The acute clinical picture

The acute clinical picture can show two types of presentations: splenic torsion but also gastric volvulus, associated or not to splenic torsion.

5.1.1 Splenic torsion

This is the main complication of wandering spleen, it usually reveals this abnormality. Pain is at the forefront of the symptoms. Splenic torsion is an emergency situation as it can quickly lead to irreversible splenic ischemia. In our series (François-Fiquet et al, 2010), 6 patients (43%) had splenectomy for splenic ischemia, but the torsion can complicate up to 65% of pediatrics cases (Romero & Barksdale, 2003).

5.1.2 Gastric volvulus +/- associated to splenic torsion

The clinical picture groups together painful symptoms associated to high occlusion with vomiting. In some cases patients can be in a real state of shock.

Gastric volvulus associated to wandering spleen is a rare condition, and its quick clinical improvement with a simple medical treatment often delays the diagnosis and access to proper surgical care (Fiquet-François et al, 2010; François-Fiquet 2009; Spector & Chappell, 2000; Qazi & Awadalla, 2004). The semiological difficulty is quite real when faced with complex clinical pictures associating gastric volvulus, wandering spleen and even in some cases a diaphragmatic hernia (Liu & Lau, 2007).

The combination of wandering spleen and gastric volvulus should be explored by additional imaging exams, and requires a quick and adapted therapeutic care.

5.2 Chronic clinical picture

Between 39% and 43% of children treated for wandering spleen had already presented similar symptoms. (Brown et al, 2003; Fiquet-François et al, 2010). Most often these children had been complaining about non-systematic recurrent but inconsistent abdominal pain for

the past months (even several years). Some children were even hospitalized several times before making a proper diagnosis. This is mostly due to the quick clinical improvement when the child was lying down (Fiquet-François et al, 2010; François-Fiquet et al, 2009).

The chronic clinical picture once again underlines the difficulty in making a proper diagnosis when faced with an atypical clinical picture.

6. Additional imaging examinations

Additional imaging examinations are key elements for the diagnostic evaluation of wandering spleen. The diagnosis cannot be made on non-specific clinical symptoms only.

6.1 Abdominal sonogram

An abdominal sonogram is the current diagnostic modality of choice for wandering spleen since it can validate the diagnosis without using radiation. (Fiquet-François et al, 2010; Brown et al, 2003; Di Crosta et al, 2009; Karmazyn et al, 2005).

It is essential to ask the radiologists to correctly evaluate the location and viability of the spleen when faced with gastric volvulus, but also dull abdominal pain.

6.2 CT-scan and abdominal magnetic resonance imaging

The efficacy of contrast enhanced CT-scan imaging has been validated and can be quite helpful in an emergency situation since it is not radiologist dependent and might sometimes be faster to access. Thus, it remains a perfect choice for acute pictures such as diagnostic evaluation of splenic torsion associated to a wandering spleen with a high risk of ischemia. It is the whorled appearance of the splenic vessels and surrounding fat that is considered pathognomonic of that condition. (Gomez et al, 2006). However even if this examination is well indicated in adults, CT-scan should remain a last-resort examination in children because of radiation exposure (Ben et al, 2006; Marinaccio et al, 2005). Abdominal magnetic resonance imaging (MRI) (Fig 7-8-9), since it does not require any anesthesia seems to be a good alternative to CT-scan for adults or older children with chronic pain. However, because it is not available in all clinical settings, it can limit its indications. It can also be recommended for uncomplicated chronic types.

6.3 Dynamic sonogram

Dynamic sonogram (on the side, standing up) is a simple examination that can help define the splenic ptosis and be relevant for chronic and hard-to-identify cases. It is also properly indicated for follow-up and monitoring exams.

6.4 Plain abdomen radiography

Plain abdomen radiography is still useful as first-line imaging examination. It allows for a quick diagnosis of gastric volvulus (Fig 10).

A well-designed imaging check-up can usually validate the diagnosis. But in some cases the diagnosis will only be validated during surgery.

7. Complications of wandering spleen

Splenic ischemia is the main complication of wandering spleen. It justifies in itself emergency therapeutic care. Gastric volvulus is a well-known complication of wandering

spleen; however its incidence is lower than splenic torsion. Sometimes there can be a pancreatitis and gastric outlet obstruction via direct external compression (sanchez et al, 2010) or even a pancreatic tail infarction (Dirican et al, 2009)

Fig. 7. a-b Magnetic resonance imaging abdominal frontal view. Spleen in a low position below the stomach, long pedicle, good vascularization

Fig. 8. Magnetic resonance imaging abdominal transversal view. a : not visible on a view going through both kidneys and b : well-vascularized spleen still visible in the left iliac fossa

Fig. 9. Magnetic resonance imaging abdominal frontal view. Long pedicle, good vascularization

Fig. 10. Plain Abdomen: gastric volvulus

8. Surgery

Surgery is the only option to guarantee the viability of the spleen; however it should not trigger any secondary ischemia. Its objective will be to restore the spleen in its anatomical position as close to normal as possible to avoid the dangling effect of the spleen at the end of its pedicle.

8.1 States of art

Surgery is the appropriate therapeutic choice, but many different approaches are available: laparoscopic surgery, laparotomy, splenopexy, gastropexy, and splenectomy.

8.1.1 Surgical approaches

In the literature, we found that in 49% of the cases the diagnosis was made during surgery (Brown et al, 2003). In this context, laparoscopic surgery is the procedure of choice. It allows for an etiological diagnosis, a good evaluation of the surgical situation while offering several therapeutic possibilities: splenectomy (Carmona et al, 2010), splenopexy (Hirose et al, 1998; Kleiner et al, 2006), gastropexy (François-Fiquet et al, 2009; Fiquet-François et al, 2010) or even a combination of several techniques such as gastropexy and splenopexy (Okazaki et al, 2010)

The choice for classic open surgery or laparoscopic surgery varies according to the different surgical teams. When there is no history of abdominal surgery, laparoscopic procedure seems to be the procedure of choice.

The risk of gastric perforation is an argument for laparotomy as the procedure of choice in case of gastric volvulus, but it does not seem to be a limiting factor for an experienced laparoscopic technician. (Mayo et al, 2001) The surgical treatment should only take place after medical treatment has been administered. The gastric suction avoids the risk of spontaneous or laparoscopy-induced gastric perforation.

8.1.2 Surgical procedures

Nowadays, it is commonly accepted to try and preserve the spleen, when viable, during the procedure, to avoid post-splenectomy infectious complications.

It is necessary to be aware of this rare clinical pathology in order to avoid delaying surgical care, which could lead to splenic ischemia or even gastric ischemia.

Nevertheless, splenic ischemia after torsion is quite common and the rate varies from 43% to 65% of cases according to the series (Fiquet-François 2010; Romero et al, 2003).

Splenectomy will be the gold standard for major splenic ischemia, when there is splenic necrosis after torsion repair and the spleen is no longer viable.

Faced with a viable or almost viable spleen, the surgery should aim for splenic conservation.

The surgery should focus on a fixation technique that will:

- reposition the spleen properly in order to avoid any further risks of torsion. The goal is to reconstruct the best possible physiological anatomy with surgical fixation.
- but also avoid gastric volvulus complication. This is why it is also recommended, as a preventive measure, to perform a gastropexy on patients with a wandering spleen in order to avoid any risk of developing a gastric volvulus. (Spector & Chappell, 2000; Soleimani et al, 2007)
- while limiting spleen manipulation that could be responsible for secondary splenic ischemia. (Fiquet-François et al, 2010)

Taking all these elements into account we have proposed an approach by Laparoscopic Assisted Gastropexy (LAG)

8.2 Procedure: LAG laparoscopic assisted gastropexy

This technique can be used in adults and children alike and in both cases of congenital or acquired wandering spleen. It can be done as an emergency procedure in case of splenic ischemia or scheduled for uncomplicated chronic cases.

8.2.1 Installation

After gastric tube decompression (in case of gastric volvulus), the patient is positioned supine on the surgical table.

A general anesthetic technique completed by bilateral Transversus Abdominis Plane Block (TAPB) to allow for evicition curare substances.

Tracheal tube and positive pressure ventilation with O2-air (0.5,0.5) was used. The nitrous oxide is formally cons indicated. (intestinal dilatation)

In children, the surgeon and assistant are at the right of the child. The laparoscopy column is placed at the level of the patient’s left shoulder. (Fig 11) In adults, the French lover position allows for the surgeon’s assistant to be perfectly positioned for this procedure.

Fig. 11. Diagram presenting the positions of: the patient, trocar entry sites, surgeon, surgeon’s assistant and laparoscopic column.

8.2.2 Procedure

A 10-mm camera trocar was inserted in the sub-umbilical region using open laparoscopy.

A laparoscope (0° degree) was inserted through the umbilical port.

2 additional working ports (5mm) were inserted: below and above the umbilicus. A third port can be inserted if necessary.

Laparoscopic exploration validated:

- the abnormal location of the spleen located in the lower left quadrant (in most of cases) and its lack of supportive ligaments,
- the vascularization of the spleen with or without ischemia, the aspect of the stomach. Normal or associated to gastric volvulus. In most of cases, during surgery we do not find the gastric volvulus identified by abdominal X-rays, it became devolvulated non-ischemic. However there is evidence of gastric distension with flaccid wall.

If the spleen is completely ischemic after de-torsion, we proposed a splenectomy.

Faced with splenic viability, we decided to perform a gastropexy. (Fig 12)

Fig. 12. Well-vascularized spleen in the left iliac fossa

The spleen was then moved freely from its abnormal location (left iliac fossa) to its normal one (sub-diaphragmatic). (Fig 13)

Fig. 13. Repositioning the spleen at the level of the right hypochondrium

We created an extra peritoneal pocket. We performed a parietal peritoneal posterolateral incision, opposite the large gastric curve, up to the diaphragm (7 cm). (Fig 14)

Fig. 14. a - b Parietal peritoneal posterolateral incision

Fig. 15. Gastropexy by suturing the peritoneal wall to the greater curvature of the stomach

We proceeded to the gastropexy. (Fig 18) We fixed the anterior stomach lining with sutures (Mersuture® 3/0; Johnson and Johnson, Somerville, NJ) on the free anterior peritoneum (Fig 15), in two planes. (Fig 16-17-18)

This suture can be done in separate stitches sutures or by two surgeons.

No drain was inserted. The nasogastric tube was removed at the end of the procedure.

Carbon dioxide gas was expelled, trocars removed, and incisions were are closed.

It is essential in case of splenectomy to ensure vaccination (pneumococcal, meningococcal, and haemophilus) and prescribe the usual antibiotic course post-splenectomy. In case of conservative splenic management, in spite of some signs of splenic suffering, it can be useful

in the immediate postoperative period to vaccinate as a precaution. Then, at 1-month postoperative and according to imaging controls (Doppler sonogram or contrast CT-Scan) showing the lack of spleen viability, an antibiotic course will be started.

Fig. 16. Gastropexy posterior wall suture done by one surgeon

Fig. 17. Final aspect of the posterior plane of the gastropexy

Fig. 18. Suture of the anterior plane (peritoneal-gastric) of the gastropexy

8.2.3 Postoperative care

The patient can drink on the day of the surgery after the legal delays post- anesthesia.

Eating can be started at D1. The patient will be kept laying down on this back the first 24 hours in order to limit shoulder pain.

The convalescence will last 10 days. The patient will be asked to stop all sport activities from 1 to 3 months according to patient's age, clinical picture and type of sports.

8.2.4 Follow-up and monitoring imaging examination

Children will be seen again for a surgical consultation at M1, M4, M10, M24 and postoperative follow-up then again every 3 years until adulthood. Doppler and dynamic sonograms (on the side, standing up) are the key examinations for this follow-up. They can assess the vascularization and viability of the spleen but also make sure the sutures are adequate and discard any residual ptosis.

If there is a doubt on splenic vascularization, a contrast CT-Scan will be proposed.

9. Conclusion

The diagnosis of wandering spleen is extremely difficult to establish because it is such a rare condition and is clinically nonspecific.

Early diagnosis and surgical care are the best guarantees for preserving the spleen. Additional imaging examinations, especially abdominal sonogram as the imaging examination of choice, can help establish a diagnosis when faced with an abnormal location of the spleen. Splenopexy and gastropexy are two surgical fixation approaches aiming to maintain the viable spleen in place.

The results of the gastropexy procedures seem encouraging, but faced with such a small number of cases, no conclusion can be established. Gastropexy seems to avoid the risk of

gastric volvulus by restoring the best possible physiological anatomy while preserving the spleen by lack of manipulation.

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