Abdominal splenosis

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ABSTRACT

Splenosis is a benign condition caused by an ectopic autotransplantation of splenic tissues after splenic trauma or surgery. It usually occurs within the abdominal and pelvic cavity. Patients are generally asymptomatic and this entity is diagnosed accidentally. However, occasionally extensive abdominal splenosis poses a significant diagnostic dilemma for gastroenterologists, especially when this condition manifests as a disseminated metastatic malignant disease on abdominal imaging.

This paper presents a concise review of the literature on this often misleading disorder. The crucial role of taking a thorough patient’s medical history concerning splenic trauma in the past, the need for differential diagnosis of tumor-like lesions disclosed on abdominal imaging and novel diagnostics modalities that allow avoiding unnecessary laparotomy in case of abdominal splenosis are stressed.

The increased prevalence of abdominal trauma due to road accidents and the growing armamentarium of available imaging modalities suggest that abdominal splenosis may be expected more often than ever.

In order to prevent any possible diagnostic doubts and unnecessary future invasive examinations, confirmed splenosis should be recorded in the medical documentation of the patient.

Key words: Abdominal splenosis. Metastatic disease. Diagnostics.

INTRODUCTION

Splenosis is a benign condition caused by an ectopic autotransplantation of splenic tissues onto exposed vascularised intra- and extraperitoneal surfaces following splenic injury (road accident, stab wound, gunshot, etc) or elective splenectomy. It usually occurs within the abdominal and pelvic cavity, involving visceral and parietal peritoneum. Patients are generally asymptomatic and in the majority of cases this entity is diagnosed accidentally. However, occasionally extensive abdominal splenosis poses a significant diagnostic dilemma for gastroenterologists, especially when this entity manifests as a disseminated metastatic malignant disease on abdominal imaging. Moreover, this rarely diagnosed disorder may be responsible for unnecessary surgical interventions in a significant number of affected patients.

ABDOMINAL SPLENOSIS

Splenic nodules following iatrogenic or accidental splenic trauma have been described in all intraperitoneal and some extraperitoneal sites. The majority of implants are found in the left upper quadrant of the abdomen (1). According to Brewster D.C. the most common sites, in order of frequency, are the serosal surface of the small bowel, the greater omentum, the parietal peritoneum, the serosal surface of the large intestine, the mesentery and the diaphragm (2). However, unusual location of splenic tissue like extensive involvement around the celiac axis and in the right paracolic gutter (3) as well as the liver (4-8), the pancreas (9), stomach, gall bladder, appendix (10), kidneys (11), ureters, lesser omentum, uterus, urinary bladder or fallopian tubes have been also described (12). Splenic autotransplants may be found retroperitoneally (13). Moreover, in cases of thoracoabdominal trauma with diaphragm rupture, thoracic splenosis, even involving the pericardium, may accompany abdominal splenosis (14-17). The rarest form of this entity is subcutaneous manifestation (18-21), including an unusual presentation of splenosis in a port site...
after laparoscopic splenectomy in an 8-year-old boy with congenital spherocytosis (22) as well as intracerebral location in the occipital pole of the brain (23).

Albrecht’s report from as early as 1896 is regarded as the first recorded case of splenosis in the human (24). The description of post-traumatic splenic autotransplantation in the human peritoneal cavity following splenectomy dates back to the beginning of the 20th century (Kuttner, 1910) (25), but the medical term “sLENOSIS” was first used by Buchbinder and Lipkopf in 1939 (26).

Due to the lack of relevant epidemiological data the true incidence of abdominal splenosis in the general population remains unknown, but given its requirement for splenic injury before development, the prevalence seems to be low. However, according to Muller and Ruthlin who performed ultrasonographic follow-up studies in patients after post-traumatic splenectomy, presumed abdominal splenosis (without histological confirmation) occurred in one-third of these patients (27). Losanoff and Jones claim that the incidence of abdominal splenosis in patients who underwent splenectomy for trauma is as high as 65-67% of splenic rupture cases (28) and possibly 18% with regard to thoracic splenosis (29).

A review of splenosis found that almost all cases (93%) were the result of trauma with subsequent splenectomy, and that 70% of these patients suffer the trauma during their teenage years (2) with thoracic splenosis diagnosed more frequently in males than females, possibly due to the higher incidence of trauma in young men. The average interval reported between trauma and abdominal splenosis is 10 years with a range of 5 months to 32 years and is shorter than in thoracic splenosis (30). One of the authors’ own experience shows that it may be even as many as 42 years (31).

PATHOGENESIS

Rupture of a pathologic spleen is more likely to occur than that of a normal one, and may be either spontaneous or traumatic. In newborn infants splenic rupture may occur in severe hemolytic disease whereas in the older children and adolescents such rupture most commonly occurs in infectious mononucleosis. Traumatic rupture of the spleen may occur from a sharp blow to the left flank, as, for example, in the automobile accidents, a direct blow or striking a projecting object while running, sledding, bicycling or the like. Removal of the spleen offers the greatest possibility of recovery and usually no deleterious effect upon the subse

CLINICAL PRESENTATION

Patients are usually asymptomatic and the splenic implants are found accidentally during unrelated diagnostic imaging or surgery (34). However, occasionally splenosis can cause serious problems including hemoptysis, pleurisy (35), symptoms mimicking myocardial infarction (36), pyrexia of unknown origin (37) or spontaneous rupture with massive bleeding into body cavities (38). Moreover, abdominal splenosis may be responsible for severe gastrointestinal hemorrhage when tissue implantation occurs in the stomach or small bowel (1,39), abdominal pain due to bowel obstruction, intraperitoneal nodule infarction, hematoma from trauma to a preexisting splenic implant (40), abdominal or pelvic mass (31,41,42), flank pain from ureteral compression and hydronephrosis (43). Recurrence of hematologic diseases, previously treated with splenectomy, may also be symptomatic (44).

A definite preoperative diagnosis of abdominal splenosis requires a high index of suspicion and should be established cautiously. A detailed medical history concerning previous abdominal trauma as well as thorough physical examination is essential for making a preliminary diagnosis of abdominal splenosis. Lack of typical chronic changes in the blood count often present after splenectomy like Howell-Jolly bodies, increase in the number of reticulocytes, sometimes also lymphocytes, monocytes, eosinophils, and trombocytosis and protective levels of antipneumococcal antibodies in a non-vaccinated patient should make a gastroenterologist consider this rare condition.

DIAGNOSIS

Widely available imaging modalities like abdominal ultrasound examination, radiological studies and standard magnetic resonance imaging (MRI) show only limited value in the diagnostic management of abdominal splenosis.

Sonographic findings are not specific in this entity and reveal round and oval soft-tissue masses in the various abdominal locations. The low density of splenic tissue makes it also difficult to visualize on standard x-rays.

Computed tomography (CT) reveals the number, shape, size, location but not identity of the nodules (Fig 1). Thus, CT scans usually show unspcific findings: oval, rounded, sessile to pedunculated (as they grow on serosal/peritoneal surfaces) multiple nodules that, initially small (from a few
millimeters), may grow over time to become quite sizeable (up to 12 cm), but usually their diameter limited by the blood supply is restricted to less than 3 cm, like in the case described by the author (31). They have density and enhancing characteristics similar to the rest of the spleen or expected density of the spleen in a splenectomized patient. One hundred or more individual splenic nodules are commonly found in splenosis and greater than 400 have been also reported (10). It should be noted that intraoperative extension of the disease is larger than previously pictured on CT scanning (45). Therefore, non-characteristic sonographic and radiological picture of abdominal splenosis may be confused with numerous conditions such as metastatic disease (31), abdominal lymphoma, carcinomatosis, hemangiomatosis, peritoneal mesothelioma, multifocal endometriosis, adenomas, primary renal or hepatic malignancy, gliomatosis peritonei, granulomatous peritonitis as the consequence of disseminated infection such as tuberculosis or histioplasmosis, foreign materials, rupture of the tumor or a hollow viscus, or, simply, reactive adenopathy.

Standard MRI with splenic implants looking like normal spleen (if present), hypointense or hyperintense depending on presentation (T1 and T2, respectively) with heterogenous enhancement C+ (GAD) is not very useful in differential diagnosis (46). Recently, several papers have proposed as a novel technique for diagnosing splenosis ferumoxides-enhanced MRI after IV administration of supramagnetic iron oxide particles that are removed from the circulation by the phagocytic reticuloendothelial system of the liver and spleen (47,48). This type of MRI combines a physiologic test of reticuloendothelial system uptake with the anatomic detail of MRI and, in the opinion of some researchers, allows the diagnosis of splenosis to be made, especially if scintigraphy is not available, so that biopsy and surgery can be avoided (3).

However, at present, there is a general consensus among experts that noninvasive Technetium (Tc) 99m radionuclide scanning is the mainstay in the diagnosis of splenosis regardless its location (Fig. 1). Tc-99m sulphur colloid is sequestered in the reticuloendothelial system and detects heterotropic splenic tissue as long as the splenunculus is at least 2 cm in diameter (49). Intraoperative extension of the disease correlates with postoperative assessment with Tc-99m sulphur colloid (45). This confirms the usefulness of scintigraphic assessment in preoperative diagnosis in order to avoid laparotomy. However, if the diagnosis is confirmed preoperatively by appropriate radionuclide modalities in a patient with history of abdominal trauma, laparotomy can be avoided.

If differentiation from hepatic tissue is necessary, for example in rare cases of suspected hepatic splenosis, a 5 mCi (185 MBq) Tc 99m-tagged heat-damaged autologous read blood cells (RBCs) or Indium 111-labeled platelets scintigraphy, more sensitive and specific for the diagnosis of splenic sequestration and phagocytosis than Tc-99m sulphur colloid scanning, can be performed (50). RBCs scintigraphy, although not free from the risk of adverse effects (fever, inflammatory response after i.v. administration of radiolabeled cells) has also been shown to be more sensitive in early splenosis, functional hyposplenism or poor splenic uptake as well as when the spleen and the liver overlap, causing poor visualization of splenic tissue by the sulfur colloid test (33,51,52).

DIFFERENTIAL DIAGNOSIS

Abdominal splenosis should be distinguished from an accessory spleen present in up to 40% of autopsies (53). Accessory spleens are congenital and arise from the left side of the dorsal mesogastrium during the embryological period of development. Supranumerary spleens may undergo hyperplasia after removal of the principal organ. These two conditions can be sometimes distinguished from one another on the basis of the patient’s medical history, the number, distribution and additional features of nodules (Table I).

Accessory spleen resembles in miniature the structure of the principal spleen, is usually single (rarely more than three) and supplied by a branch of the splenic artery. Over 75% of accessory spleens are found in the splenic bed: immediate vicinity of the splenic hilum and pedicle (if the principal spleen is present), but may be widely scattered and occasionally found in other locations: gastrosplenic ligament, retroperitoneal region near the tail of the pancreas, greater omentum, splenocolic ligament, mesentery of the large and small bowel, left adnexa in the female and left scrotum in the male.

It is usually found by accident during ultrasound examination of the abdomen in the left upper quadrant as small, solid focal changes that sometimes, just like abdominal splenosis, may be taken for enlarged lymph nodes, metastases, gastric, renal, suprarenal or pancreatic tumor.
POSSIBLE CONSEQUENCES

There has been a debate concerning the functional significance of the splenic autotransplants in a generally asplenic patient. A recurrence of Felty syndrome 2 years after splenectomy (54) and idiopathic thrombocytopenic purpura due to splenosis 12 years after splenectomy (30) as well as amyloid deposits and miliary tuberculosis obtained from splenosis implants (7) suggest a functional reticuloendothelial system within the implants.

However, the most controversial issue was the question about the immune status and host defense in the splenectomized patient with abdominal splenosis since it was not clear whether it played a protective role against postsplenectomy sepsis. It has been reported that the overall incidence of sepsis and mortality is significantly higher in cases of incidental splenectomy than in post-traumatic splenectomy (55). Nevertheless, the significant increase in the serum levels of antipneumococcal antibodies in patients with splenic autotransplantation, proved their adequate humoral response to pneumococcal infections and the additional protective immunologic effect of splenosis indicate that this rare condition actually may provide protection against serious postsplenectomy infection or sepsis (1,16). In fact, implanted splenic tissue may be beneficial and protect against systemic encapsulated bacterial infection, which can be a major problem in asplenic patients (56).

MANAGEMENT

For many decades splenosis used to be diagnosed at the time of surgery. Also nowadays, due to sometimes alarming radiologist’s misinterpretation of the CT findings, splenic implants may be mistaken for neoplastic masses/adenopathy and the patient is subjected to the prompt exploratory laparotomy.

At present, laparoscopy provides a port for minimally invasive entry for the visualization of suspected masses, and allows access for potential subsequent biopsy or resection (57). At laparoscopy/laparotomy splenosis differ in colour and consistency from malignant tumors and in consistency from fibroids. These implants are often bluish, but may vary in color from pink to dark red/greenish black, have no hilum and are supplied by local arteries that penetrate their fibrotic capsule. Lack of adhesions within the abdominal cavity is quite a characteristic feature of splenosis.

If the biopsy is performed without a preoperative suspicion of splenosis, the frozen section should be carefully examined for evidence of splenosis, but its quality may not be sufficient enough for detailed assessment of the splenic tissue. Macroscopically and histologically two adjacent implants may differ markedly. The tissue in splenosis often reveals distorted architecture with no hilum and a poorly formed capsule. Most reports describe the tissue as lacking trabecular structures, having less elastic tissue than a normal spleen and poorly

### Table I. The differential diagnosis between splenic autotransplants and accessory spleens

<table>
<thead>
<tr>
<th>Splenic autotransplants</th>
<th>Accessory spleens</th>
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</thead>
<tbody>
<tr>
<td><strong>Medical history</strong></td>
<td>Splenic trauma or splenectomy</td>
</tr>
<tr>
<td>Location</td>
<td>Any widespread intraperitoneal and extraperitoneal, including intrathoracic, subcutaneous and intracranial</td>
</tr>
<tr>
<td>Number</td>
<td>Numerous, sometimes even 400</td>
</tr>
<tr>
<td>Size</td>
<td>Usually less than 3 cm in diameter</td>
</tr>
<tr>
<td>Shape</td>
<td>Oval, rounded, sessile or pedunculated, without hilum</td>
</tr>
<tr>
<td>Blood supply</td>
<td>Small vessels entering capsule at periphery at any site of nodule</td>
</tr>
<tr>
<td>Histology</td>
<td>Microscopic structure may vary from identical to the normal spleen to various stages of degeneration with distorted architecture and poorly formed capsule</td>
</tr>
<tr>
<td>Function</td>
<td>Same as normal spleen in well-developed autotransplants</td>
</tr>
<tr>
<td>Clinical significance</td>
<td>Important in differential diagnosis, e.g of malignant diseases</td>
</tr>
<tr>
<td></td>
<td>Congenital normal variant</td>
</tr>
</tbody>
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Based on references 10, 18, 32, 58, 59.
formed or deficient white pulp with normal appearing red pulp, but splenosis with histology and immunohistochemistry indistinguishable from the normal spleen have been also described (58,59). Unless complicated, there is no inflammatory reaction in the adjacent tissue (32). The pathologic differential diagnosis may include lymph nodes in any reactive condition and low grade lymphoma.

The current opinion is that when the splenosis is incidentally diagnosed in an asymptomatic patient, complete surgical removal is not indicated (41). The surgical approach is recommended in cases of symptomatic or complicated splenosis and in some patients with hematological disease for whom splenectomy is beneficial and splenic autotransplantation must be avoided (like congenital splenomegaly). In this subset of patients preoperative use of imaging methods might improve diagnostic certainty and contribute to a well-planned surgical intervention.

The surgical approach should be also recommended in case of an uncertain diagnosis, especially when scintigraphic methods are not readily available and there is a suspicion of a malignant disease, because distinguishing the nature of the abdominal mass considerably modifies the management.

Interestingly, most patients who have an exploratory laparotomy for abdominal pain have cessation of the pain after the procedure, regardless of whether the splenic nodules have been removed or not (31,60).

CONCLUSIONS

To conclude, with the increased prevalence of abdominal trauma due to road accidents and the growing armamentarium of available imaging modalities abdominal splenosis may be expected more often than ever. Occasionally, it leads to a great diagnostic dilemma because splenic implants may be misinterpreted as neoplastic lesions or adenopathy. Presumed diagnosis can be made in a patient with absence of siderocytosis and Howell-Jolly bodies in the blood smear and a history of splenectomy or severe abdominal trauma in the past. Scintigraphy using heat damaged Tc-99m-labelled autologous RBCs is a reliable non-invasive diagnostic method of choice in this rare condition and may allow to avoid unnecessary abdominal surgery. In order to prevent any possible diagnostic doubts and unnecessary future invasive examinations, confirmed splenosis should be recorded in the medical documentation of the patient.

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