

Thoracic splenosis mimicking a pleuropneumonia

A case report

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Abstract

Rationale: Splenosis is the development of one or more heterotopic splenic tissue autoimplants following rupture of the spleen and remains mostly asymptomatic.

Patient concerns: We report a case of a 50-year old post-traumatic splenectomized man admitted for a left side community acquired pneumonia resistant to antibiotics.

Diagnoses: The diagnosis of intrathoracic ectopic spleen was suspected because of the history of spleen trauma with diaphragm rupture and the absence of Howell-Jolly bodies.

Interventions: Technetium (Tc)-^{99m} colloid scintigraphy SPECT, fused with CT scan showed an intense radionuclide uptake on hyper vascularized masses without any additional pathologic uptake and confirmed the diagnosis of thoracic splenosis.

Outcomes: Despite any lifelong penicillin prophylaxis, he had no history of infections eight years after the diagnosis.

Lessons: Physician must be aware of this differential diagnosis and of its consequences. Depending on its size and location, it may lead to incorrect diagnosis (tumor, empyema, abscess ...), treatment and invasive procedures while the diagnosis of splenosis only relies upon imaging studies associated with functional study of the uptake of particles or cells.

Abbreviations: CMV = cytomegalovirus, CT = computed tomography, MRI = magnetic resonance imaging, OPSI = overwhelming post-splenectomy infection, Tc = Technetium, TS = thoracic splenosis, VATS = video-assisted thoracoscopic surgery.

Keywords: ^{99m}Tc-sulfur colloid scintigraphy, computed tomography, thoracic splenosis

1. Introduction

Splenosis is defined as autotransplantation of splenic tissue into another anatomic compartment, after rupture or trauma of the spleen. Most cases occur in the peritoneal cavity and the thoracic location is a comparatively rare finding.^[1-3] Incidence of splenosis after splenic trauma ranges from 58% to 65%.^[1,2] In case of diaphragmatic rupture, the prevalence of thoracic splenosis (TS) has been described as high as 18%.^[1] Patients with

TS are mostly asymptomatic and TS is usually diagnosed on routine chest radiography. However, chest imaging typically shows well defined mass which can mimic malignant tumor, abscess, or empyema.^[3] The diagnosis of TS may be challenging for physicians, leading to invasive procedures. We report a case of intra-thoracic splenosis that mimicked left side community acquired pleuropneumonia and led to delay in correct diagnosis. Patient gave informed consent and ethical committee review was not applicable.

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2. Case report

A 50-year-old posttraumatic splenectomized man was admitted to our Infectious Diseases Department because of fever and chills. His medical history was marked by a car crash occurring 34 years before. At that time he had suffered from a hemoperitoneum and a hemothorax with left diaphragm rupture leading to a resection of the lacerated spleen. The patient recalled having been told that a splenectomy had been performed. A review of his medical records showed that he had been taking penicillin V as prophylaxis for 2 years. He received only 1 shot of pneumococcal polysaccharide vaccine, 25 years later. He had no history of infection (pneumonia, meningitis, sinus infection...) since this surgery. Two weeks before his admission, he developed fever, night sweats, and cough. He denied other constitutional symptoms including weight loss, chest pain, or hemoptysis. Physical examination only revealed fever (temperature 38.4°C) and was consistent with a left pleural effusion. No lymphadenopathy or tumoral syndrome was noted. Since the chest x-ray showed left lower lung zone opacity (Panel A), he was considered to have community-acquired pleuropneumonia and received

antibiotic with no efficiency. Blood tests showed a lymphocytosis (12×10^3 cells/mm³), mild increased protein C reactive (44 mg/L), and hepatic cytolysis with an increased level serum alanine transferase and aspartate aminotransferase upper to 6 times the limit of normal. No Howell–Jolly bodies were described. Cytology, immunophenotyping, and bacterial analyses of pleural effusion were normal or negative. The patient was eventually found to have primary cytomegalovirus (CMV) infection and fever resolved within 15 days. One month later the computed tomography (CT) scan demonstrated the persistence of contrast-enhanced masses on both side of left diaphragm, abdomen, and peritoneum without pleural effusion. Upper and lower thoracic nodules measured $3 \times 3 \times 2$ cm and $1.5 \times 1.5 \times 1$ cm, respectively. No suspect lymph nodes were described. At this period, the patient was asymptomatic with no fever, weight loss, or even night sweats. C-reactive protein, liver blood tests, blood cell count, and plasma lymphocytes immunophenotyping results were within normal limits. However, because of the absence of any symptoms, the history of spleen trauma with diaphragm rupture and the absence of Howell–Jolly bodies in this splenectomized patient, the diagnosis of intrathoracic ectopic spleen was suspected. As shown in Fig. 1, Technetium (Tc)-99m colloid scintigraphy single photon emission computed tomography (Panel C, E, G) was fused with CT scan (Panel B, D, F) and showed an intense radionuclide uptake on hyper vascularized masses without any additional pathologic

uptake. These findings confirmed the diagnosis of left thoracic and hypocondrium splenosis and no invasive procedures to explore these lesions were done. The patient received immunizations against *Haemophilus influenzae* and *Streptococcus pneumoniae*, and was informed of an increased risk of bacterial infection. Despite any lifelong penicillin prophylaxis, he had no history of infections 8 years after the diagnosis.

3. Discussion

TS is defined as an autoimplantation of viable splenic tissue in the thoracic cavity. While early reports of TS in middle of 20th century suggested that TS was a consequence of the development of embryonic rests of splenic tissue, it is now clear that traumatic disruption of spleen is a crucial step. Diaphragm rupture is essential for dissemination and proliferation of splenic tissue to the left pleural cavity. After autoimplantation, splenic pulp derive its own blood supply from the adjacent tissue and the surrounding circulation and grows up slowly into mature splenic tissue which appears to preserve some degree of splenic function such as the elimination of aged blood cells.^[3–5]

Since the first case of TS reported by Shaw et al^[6] in 1937, almost 100 cases have been referred to in the available English language literature.^[1–16] Khan et al^[3] reported a total of 66 cases in 2010. During the last 6 years most of cases were case reports.^[8,11–13,15]

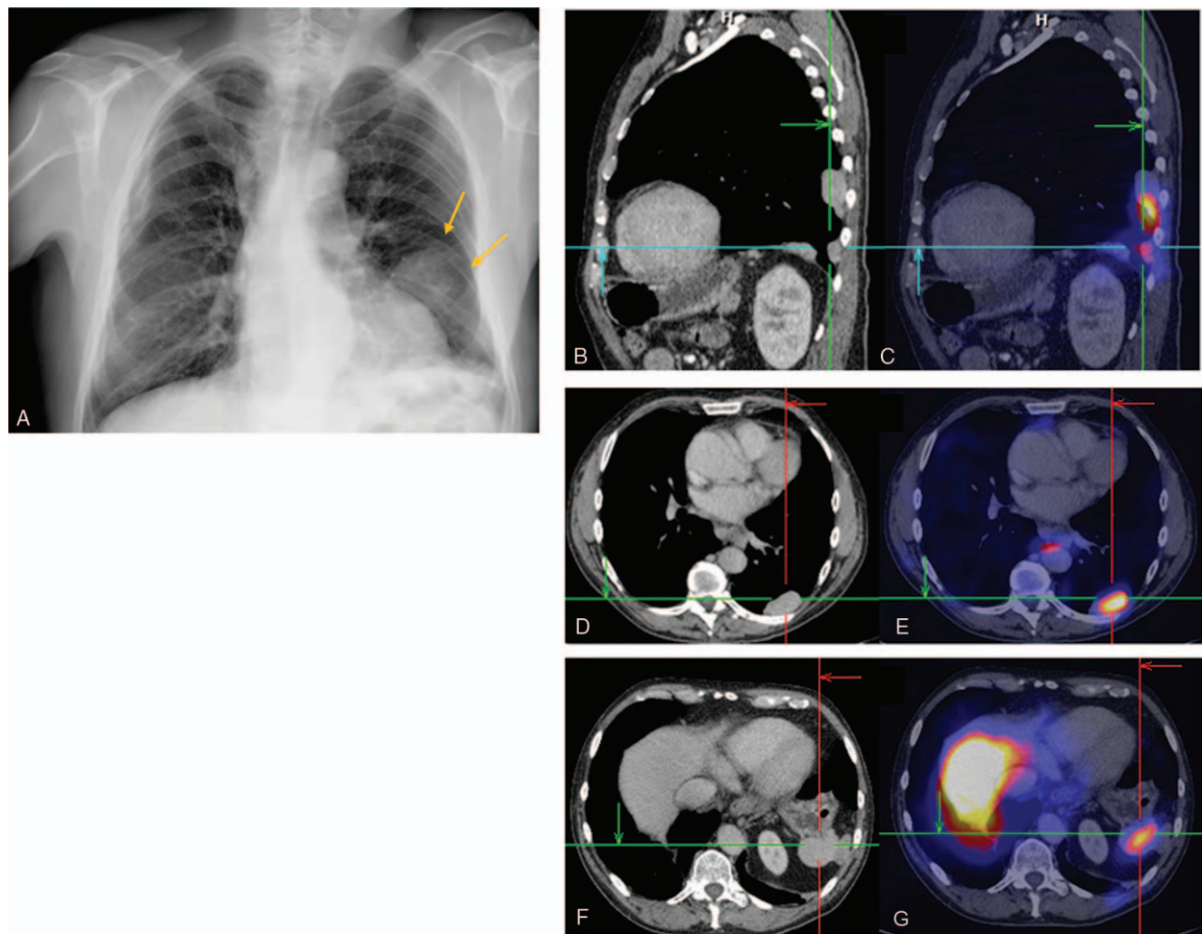


Figure 1. A 50-year-old male patient with thoracic splenosis mimicking a pleuropneumonia. Chest x-ray (A) showed a left side opacity. A CT scan (B, D, F) was fused with Tc-99m colloid scintigraphy SPECT (C, E, G) and showed an intense radionuclide uptake on thoracic (B, C, D, E) and abdominal (F, G) hyper vascularized masses corresponding to splenic tissue. CT=computed tomography; SPECT=single photon emission computed tomography.

Because splenosis is mostly asymptomatic, the time between the traumatism and the development of TS is not precisely known, ranging from 1 to 45 years with a mean delay of up to 21 years, suggesting that the incidence of this sequela might be underestimated.^[7,16] In our case TS was diagnosed 34 years after splenectomy, which presents one of the longest time period described in the literature. TS mainly occurs after gunshot wounds or motor vehicle injuries which can explain that most of cases of TS are described in men.^[3,4,7,16] Abdominal splenosis is found to be associated in 24% of TS cases.^[3] While peritoneal splenosis is frequently symptomatic with abdominal pain, gastrointestinal obstruction, or hydronephrosis, TS has been exceptionally described as the cause of hemoptysis, thoracic pain, or cough.^[3,7,9] TS are often discovered on routine thoracic imaging and only concerns left hemithorax.^[2,3,7,9,13,16] Chest CT features consisted of homogeneous, non-calcified pleural nodules or masses; lung parenchyma splenosis is described only after a history of lung laceration. These lesions are mainly multiple with an attenuation similar to normal spleen.^[2,3,7,9,13,16] Single lesion is described in less than 15% of TS.^[3] Their size varies from few millimetres to 8.5 cm in diameter.^[3,7,10,14] Magnetic resonance imaging (MRI) appearance of TS is also similar to normal spleen with isointensity nodules on T1 and T2 weighted images.^[2,7]

However, because of the rarity of TS, this diagnosis may be challenging for physicians and differential diagnosis should be considered first.^[3] The differential diagnosis for unilateral pleural nodules includes infectious lesions, pleural metastases, primary lung carcinoma, asbestos-related pleural disease, lymphoma, thymoma, localized fibrous tumor of the pleura, neurogenic tumor, or auto-immune diseases (rheumatoid arthritis). In the case of our patient, no asbestos or tobacco exposure was reported. One month after the primary CMV infection he did not have any general symptoms such as recurrent fever, weight loss, asthenia or night sweats or cough, without any treatment. All these findings and the lack of pleural plaques, mediastinal lymph nodes or mass or pleural effusion on the CT scan contributed to exclude the diagnosis of hematological malignancy, thymoma, lung carcinoma, and mesothelioma. In pleural lymphoma (Hodgkin and non-Hodgkin diseases) recurrent pleural effusion is usually reported and the size and number of nodules increase within a brief time period.^[5,17] Pleural parenchymatous of our patient did not show any abnormal cells in immunophenotyping and cytology analyses. An infection presenting as pleural or pulmonary nodules was also excluded because of the immunological status of the patient, the CT scan features, and the absence of clinical manifestations. Fungal infection such as aspergillosis, mucormycosis, cryptococcosis mainly occurs in immunocompromised patients. Bacterial abscess related to *Nocardia spp*, tuberculosis infection or pyogenic bacteria are always associated with clinical manifestations (cough, night sweats, cutaneous nodules, fever, ...).^[18] Pulmonary nodules may occur in less than 1% of rheumatoid arthritis. These intraparenchymatous lesions are usually multiple, located in the middle and superior peripheral lobes and may cavitate in up to 50% of cases.^[19] If malignancy remains part of the differential diagnosis, fine-needle aspiration biopsy and video-assisted thoracoscopic surgery (VATS) or thoracotomy should be performed.

The history of thoraco-abdominal injury leading to splenectomy, the presence of left sided pleural-based pulmonary nodes and the absence of Howell-Jolly bodies or siderocytes should suggest a potential diagnosis of TS. The absence of Howell-Jolly bodies or siderocytes in peripheral blood indicates the presence of

functional splenic tissue (in patients with splenectomy).^[4] Thirty years ago, diagnosis of TS was usually confirmed by studying tissue samples obtained by such invasive measures (thoracotomy, VATS, needle biopsy).^[2,3,9] However it is now well-established, that a definitive diagnosis can be reached using non-invasive diagnostic tools such as imaging studies associated with functional study of the uptake of particles or cells: Tc-99 sulfur colloid scintigraphy, Tc-99 sulfur heat-damage erythrocytes scintigraphy, Tc-99 white blood cell scan or indium 111-labeled platelet, or super-paramagnetic iron oxide particles MRI.^[1-3,5,16,20] Among these investigations, scintigraphy using heat-damaged erythrocytes tagged with Tc-99 is considered as the most specific nuclear imaging. Heat damage erythrocytes deposit in splenic tissue throughout the body. Radionuclide tagging of these erythrocytes can lead to identify ectopic splenic tissue with a higher specificity.^[1,2,5,16,20] Super-paramagnetic iron oxide particles MRI, which localize to sites of phagocytic reticuloendothelial cells, is an alternative technique. This technique seems to have a better resolution than other nuclear imaging modalities and may be more commonly used in the future despite the absence of comparative studies especially for the diagnosis of TS.^[1,2,5,16]

Once the diagnosis of splenosis has been established in a patient, it is of importance to emphasize that little is known about the degree of recovery of splenic function in such cases.

Overwhelming post-splenectomy infection (OPSI) is a fulminant sepsis due to encapsulated bacteria with a high rate of mortality. The overall incidence of infection in splenectomized patients varies from 2.3 to 7.7 per 100 persons-years, depending on underlying conditions.^[21,22] The incidence of infection among children and adults seems to be similar while the incidence of death is higher among children.^[21] Although the risk of infection is increased within the 2 years following splenectomy, 30% to 40% of OPSI have been documented after 5 years and case reports have been reported 40 years post-splenectomy, suggesting a long-lasting infection risk in these patients.^[22] However, the degree of immunoprotection offered by ectopic splenic tissue remains unclear. Several experimental studies in partially splenectomized animals or using spleen autotransplants in splenectomized animals have demonstrated that the degree of recovery of a normal spleen immune function was highly variable and relies especially on the mass, the location of residual spleen tissue, and the quality of spleen vascularization. It seems that when splenosis represents less than one-third of weight of the original spleen, host defenses are not restored, with poor phagocytic capacity, antibody response, or serum lysozyme levels.^[4,23,24] Incidence of OPSI in patients with splenosis is unknown, especially because most of human data come from case reports. Even though the absence of Howell-Jolly bodies (or of pitted erythrocytes as well) could suggest some recovery of macrophagic spleen function, the physician must keep in mind that the risk of OPSI, especially by encapsulated bacteria remains higher. Indeed Connell et al^[4] reported 16 cases of serious infections due to encapsulated bacteria, leading to death, despite a large amount of splenosis. These OPSI can occur up to 17 years after splenectomy. Therefore, even in patients with TS and non Howell-Jolly bodies, the management of splenosis consists on preventive measures to reduce the risk of OPSI. Penicillin prophylaxis and vaccinations against encapsulated bacteria should be given to these patients in the same way it is done in any asplenic or hyposplenic patient.^[3,4,9,25] Surgical excision of splenic tissue should only be considered for symptomatic patient (chest pain, hemoptysis).^[3-5,9,11,15]

4. Conclusion

TS remains a rare entity that is probably underdiagnosed considering that splenic implants are mostly asymptomatic and may be of a small size. However, physicians must be aware of this differential diagnosis and of its consequences. Depending on its size and location, it may lead to incorrect diagnosis (tumor, empyema, abscess, . . .), treatment and invasive procedures while a simple question regarding abdominal trauma in a patient's history can lead the clinician to consider this diagnosis. Final diagnosis of splenosis only relies upon imaging studies associated with functional study of the uptake of particles or cells. Since the recovery of splenic function cannot be ascertained on an individual basis, even in those patients without Howell-Jolly bodies, it is careful to prevent the occurrence of OPSI in patients with splenosis.

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