



Thoracic splenosis

Report of a case and review of the diagnostic workup

Ann. Ital. Chir.

Published online (EP) 29 July 2016

pii: S2239253X16025834

www.annitalchir.com

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Thoracic splenosis. Report of a case and review of the diagnostic workup.

Splenosis is the autoimplantation of splenic tissue to ectopic sites after spleen injury. Although splenosis most commonly occurs in the abdomen, it may occur in the thorax in case of diaphragm rupture. Thoracic splenosis (TS) is often asymptomatic and is diagnosed incidentally in the course of chest examination.

We describe the case of a man, with a history of thoracoabdominal trauma, undergoing routine chest radiography with the evidence of radio-opaque images close to the inferior left curvature of the cardiac shadow, which resulted to be nodules of TS at the contrast enhanced CT scan.

CT scan or MRI and an accurate anamnesis are usually sufficient to diagnose TS, otherwise scintigraphy with ^{99m}Tc is needed. Imaging-guided biopsy and thoracoscopy should be performed if scintigraphy is unavailable or results are inconclusive. It is not usually necessary to remove the TS because the splenic tissue is slow growing, non-invasive and benign. Rarely surgery may be required for symptomatic TS (hemoptysis, cough or pleuritic chest pain).

TS may be difficult to diagnose, especially if features suggesting TS are not recognized and the anamnesis is not known. This may lead to an extensive work-up and unnecessary invasive diagnostic procedures (including biopsy, thoracoscopy, up to thoracotomy). In the workup of thoracic nodules TS should be considered in patients with a history of trauma and spleen injury.

KEY WORDS: Thoracic splenosis, Spleen, Mediastinal nodules, Thoracoabdominal trauma

Introduction

Splenosis is the condition of heterotopic autotransplantation and implantation of splenic tissue to ectopic sites. It is also known as ectopic spleen and it is a very rare

occurrence due to the spillage of splenic tissue and cells usually after an abdominal trauma. Splenic implants may be located anywhere in the peritoneal cavity, more often on the serosal surface of the small bowel and colon, the parietal peritoneum and the subdiaphragmatic areas ¹. Thoracic splenosis (TS) is a rare condition resulting from concomitant rupture of the spleen and the left hemidiaphragm. Most cases of TS are asymptomatic and are incidentally diagnosed during the investigation of other clinical presentations.

The first description of TS is by Shaw in 1937: he observed the post-traumatic spleen autotransplantation into the thorax ². Buschbinder in 1939 first used the term splenosis to describe the intra-abdominal splenic implants after a trauma ³. Since these reports, many cases of abdominal splenosis have been described, but only

Pervenuto in Redazione Maggio 2016. Accettato per la pubblicazione Giugno 2016.

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66 cases of TS have been reported in the English-language literature up to 2010, as written by Khan, and no more than 10 case-reports have been published since 2010 to date ⁴.

The diagnosis of TS may be challenging if the anamnesis is not known, thus leading to an extensive workup and unnecessary invasive procedures (PET-CT, biopsy, thoracoscopy, thoracotomy). Therefore, an understanding of this occurrence is important both to avoid invasive time-consuming workups and in the further management of this entity.

Case Report

A 59-year-old man underwent chest radiography as routine checkup because of his previous professional exposure to asbestos. The chest radiography showed an irregular profile of the inferior left curvature of the cardiac shadow, with radio-opaque images (Fig. 1). Therefore, he underwent contrast enhanced chest computed tomography (CT) scan which showed multiple nodules ⁸ in the inferior anterior left mediastinum between the parietal pleura and the left ventricle. They were ovoidal and their dimensions varied from 1 to 5 cm. The vascularization pattern of the nodules was the same of that of spleen parenchymal (Fig. 2). He had undergone a thoracoabdominal trauma with hemothorax and hemoperitoneum due to splenic and left hemidiaphragm rupture 33 years before. He was completely asymptomatic and he is still so after 8 years from the diagnosis of TS.

Discussion

Splenosis is due to autotransplantation after spleen removal secondary to traumatic rupture or surgery. The prevalence of splenosis after spleen injury ranges from 16% to 67%, but the true prevalence is unknown because splenosis is usually an incidental finding at imag-

ing, surgery or autopsy ^{5,6}. Only 18% of the cases of splenosis are TS ⁷. The essential condition for the passage of the splenic tissue to the thorax is the rupture or a defect of the diaphragm. Splenosis may occur very rarely as a result of congenital malformations due to an incomplete fusion of the dorsal mesogastrium ⁸.

The review by Khan reported that the mean age of presentation of TS is 44 years (range 15-79), with a male predominance (76%) ⁴. However, these numbers are poorly significant because TS is caused by thoracoabdominal traumas (more frequent in young people) and its diagnosis is very often incidental. Freise described the only one case to date of TS resulting from a trauma without post-traumatic splenectomy ⁹. It is an exceptional entity but it is reasonable to think that abdominal splenosis will likely be found more and more in the near future in patients undergoing spleen injury treated conservatively without surgery.

The splenic pulp has an high ability to implant itself onto the serosal surfaces of the thorax, abdomen and pelvis and to draw its own blood supply from the submesothelial vasculature or from the surrounding circulation (e.g. the pulmonary circulation in the case of TS) ⁴. Splenic implants subsequently grow into histologically mature splenic tissue with further neovascularization ⁴. Microscopic examination of nodules shows that blood vessels do not enter through a designated hilus (contrary to the normal spleen) but branch into smaller vessels, which penetrate the collagenous capsule around its circumference. This capsule is poorly formed and merges with surrounding tissues. The splenic pulp is the same as the spleen with both well-formed or loose lymphoid follicles and trabeculations ⁴. The differential diagnosis of TS includes pleural mesothelioma, primary lung malignancy, metastatic lesions, mediastinal tumor, lymphoma, neurogenic tumor, infectious lesion and thorax endometriosis. Therefore, accurate anamnesis, clinical evaluation and blood examination are mandatory. The diagnostic workup depends on whether the radiologist considers TS in the differential diagnosis of suggestive findings.



Fig. 1: Chest radiography shows an irregular profile of the inferior left curvature of the cardiac shadow, with multiple radio-opaque nodular images.

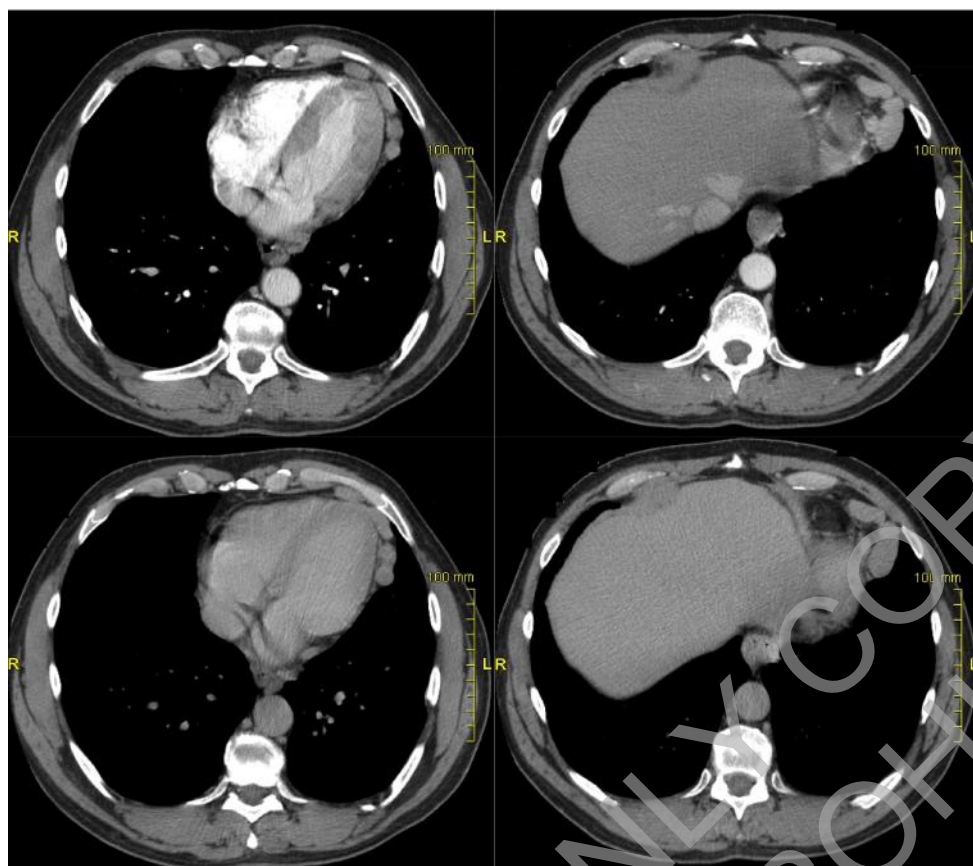


Fig. 2: Axial CT images in arterial and venous phases show well defined pericardial-based multiple ovoidal nodules with homogenous enhancement resembling that of the normal spleen.

The chest radiography is usually the first examination which shows mediastinal nodules or pulmonary lesions. They are further investigated with CT scan or magnetic resonance imaging (MRI). Contrast enhanced CT or MRI examinations are essential to study the vascularization of the thoracic lesions. Splenic nodules in the thorax are always seen on the left side, most of them being pleura-based. Their size varies from 0.5 to 7 cm in the cases reported, and only 12% of them had a single lesion⁴. On contrast enhanced CT scan splenic nodules have smooth ovoidal or round shape with the same enhancement pattern of the normal spleen. In the case of serosal implants, the nodules show homogenous and uniform enhancement.

On conventional MRI splenic nodules are hypointense on T1-weighted images and hyperintense on T2-weighted images, similar to the normal spleen. Contrast-enhanced MRI with superparamagnetic iron oxide (SPIO) is useful because the SPIO, which is taken up by reticuloendothelial cells in the liver and spleen, produces local inhomogeneities in the magnetic field that cause rapid dephasing of transverse magnetization, resulting in a decrease of signal intensity of those organs on T2-weighted images.

Scintigraphy with ^{99m}Technetium is performed when the diagnosis of TS is equivocal. Two techniques may be used: ^{99m}Tc-labeled sulfur colloid and ^{99m}Tc-labeled heat-

damaged red blood cell. ^{99m}Tc-labeled sulfur colloid is sequestered in reticuloendothelial cells and thus it can be useful for detecting all ectopic splenic tissue. ^{99m}Tc-labeled heat-damaged red blood cell scintigraphy is preferred because of reduced uptake by normal liver tissue, resulting in an improved target-to-background ratio¹⁰. This technique is considered more sensitive and specific for the diagnosis of splenic tissue.

Image-guided biopsy (with fine-needle aspiration) is also a diagnostic tool for thoracic nodules of unknown origin: it can be performed if scintigraphy is unavailable or results are inconclusive. Often the location of a lesion precludes the fine-needle aspiration.

Video-assisted thoracoscopic surgery has been described as a tool of diagnosis, but it could be performed only when the imaging diagnosis is equivocal or the TS is symptomatic. Moreover, thoracoscopy may be limited by thick pleural adhesions and inaccessible nodules, which result in the need for thoracotomy.

If the diagnosis is confirmed by imaging, surgery is not indicated unless the patient is symptomatic. It is not usually necessary to remove the thoracic nodules because the splenic tissue is slow growing, non-invasive and benign. Rarely the TS is symptomatic, with hemoptysis, cough or pleuritic chest pain.

TS is thought to have all the functions of normal spleen. It has been reported that the incidence of sepsis is sig-

nificantly lower in splenectomized patients who have splenosis than in those without it ¹¹. Moreover, it has also been shown that among splenectomized patients, the number of pitted red blood cells in peripheral blood smears is significantly lower in patients with splenosis following trauma than in those who have undergone elective splenectomy, thus indicating the ability of splenic implants to remove defective and old red blood cells ^{4,11}. Finally, ectopic splenic tissue is protective against systemic infections by encapsulated bacteria (*Neisseria meningitidis*, *Streptococcus pneumoniae*, and *Haemophilus influenzae*), which might be a serious problem in asplenic patients ^{11,12}.

Conclusion

TS is a rare occurrence that must be considered in patients with asymptomatic left pleural-based thoracic nodules and a history of thoracoabdominal trauma with splenic injury and left diaphragmatic lesion.

The differential diagnosis of TS includes pleural mesothelioma, primary lung malignancy, metastatic lesions, mediastinal tumor, lymphoma, neurogenic tumor, infectious lesion and thorax endometriosis.

In the workup contrast, enhanced CT and/or MRI may be adequate to diagnose TS. Scintigraphy is a very useful tool, which can preclude more invasive diagnostic procedures such as fine-needle biopsy and thoracoscopy. Surgery should be recommended only in case of uncertain diagnosis or symptomatic TS.

Riassunto

L'impianto di tessuto splenico in siti ectopici dopo un trauma della milza viene definito splenosi. Sebbene si verifichi più comunemente in addome, essa può localizzarsi anche nel torace in caso di rottura del diaframma. La splenosi toracica (TS) è spesso asintomatica ed è diagnosticata nel corso di indagini radiologiche del torace. Descriviamo il caso di un uomo, con storia di pregresso trauma toraco-addominale, che ha eseguito una radiografia del torace per routine con evidenza di immagini radioopache in adiacenza dell'arco inferiore sinistro dell'ombra cardiaca, che sono risultate essere noduli di TS confermate dalla CT con mezzo di contrasto.

La CT o la MRI unite ad una anamnesi accurata sono di solito sufficienti per diagnosticare la TS, altrimenti è necessaria la scintigrafia con ^{99m}Tc. La biopsia guidata dall'imaging e la toracosopia dovrebbero essere eseguite solo quando la scintigrafia non è disponibile o i risultati delle altre metodiche non sono risolutivi. Normalmente non è necessario rimuovere la TS perché

il tessuto splenico ha una crescita lenta, non invasiva e benigna. Raramente la chirurgia può essere necessaria quando la TS è sintomatica (emottisi, tosse, dolore di tipo pleuritico).

La TS può essere difficile da diagnosticare, specialmente se l'aspetto dei noduli non è univoco e l'anamnesi non è nota. Ciò può portare a un iter diagnostico eccessivo e a procedure non necessarie (biopsia, toracosopia fino alla toracotomia). Nel work-up dei noduli toracici la TS dovrebbe essere considerata nei pazienti con storia di trauma e rottura della milza.

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