Splenosis of the peritoneal cavity resembling an adnexal tumor: case report

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Summary

Introduction: Splenosis is the autoimplantation of ectopic spleen tissue in various anatomic cavities of the body resulting after trauma or rupture of the splenic parenchyma. The major localization sites of this phenomenon are mainly intraperitoneal, the gastrenteric tract, genitalia, intrahepatically and the kidneys. Extraperitoneal locations occur less frequently and include the thorax and brain. Also locallization in the subcutaneous fat has been described. *Case report:* We present the case of a 32-year-old woman with symptomatic peritoneal cavity splenosis occurring ten years after traumatic splenectomy. The patient was admitted to our department with the clinical presentation of an adnexal tumor. US and CT confirmed an adnexal mass. Exploratory laparotomy was performed and multiple focal lesions were noticed on the uterus, ovaries and intestinal tract. Biopsies were taken and sent for histological analysis. The pathology specimen revealed ectopic splenic tissue. After surgical intervention the patient remained asymptomatic. *Conclusion:* Splenosis is a rare phenomenon which clinicians should be aware of in order to spare patients from pointless surgical interventions. Patients with abdominal masses and post-traumatic splenectomy should be checked for splenosis.

Key words: Splenosis; Autoimplantation; Splenectomy; Adnexal tumor.

Introduction

The first description of the splenosis phenomenon in humans was made by Albrecht [1] and later by Schilling [2] in 1907. The mechanism of autoimplantation was first analyzed by von Kuttner [3] in 1910. The term splenosis was first coined in 1939 by Buchbinder and Lipkoff during exploratory laparotomy for an abdominal tumor [5].

Splenosis may present in 16-67% of patients with traumatic rupture of the spleen [6]. Approximately 100 cases have been reported universally [7]. Spenosis is a rare condition which can progress asymptomatically. Its clinical performance is not characteristic and varies depending on whether the site is intraperitoneal or extraperitoneal. It may be characterized by mild pelvic pain or may demonstrate acute abdominal manifestations in patients with abdominal masses of ectopic splenic tissue [8]. Cases involving the ileum have been described [9], as well as gastrointestinal hemorrhage following rupture of the capsule of the ectopic splenic tissue, [10] and hydronephrosis because of amenable compaction of the ureter in the small pelvis [11].

The diagnosis of splenosis is difficult to make with the usual imaging media, such as ultrasound (US) and computed tomography (CT), giving false imaging findings which refer to neoplastic procedures [5]. Sometimes an erroneous clinical estimate leads to pointless surgical management [5]. The use of radioactive technetium-99 intravenously provides important information on the existence of ectopic splenic tissue [12].

Peripheral blood examination may show the existence of Howell-Jolly bodies which are frequently recognized in splenosis cases. These bodies are intracellular deposits in the red blood cells, nigrescent after Wright staining, and are constituted by condensate DNA of fragmented splenic cell nuclei.

Case Report

A 32-year-old woman came to the Outpatient Gynecology Department with atypical abdominal pain and accompanying symptoms of dysmenorrhea, dyspareunia and a feeling of meteorism. The most remarkable fact in the patient's history was a splenectomy, ten years before, because of traumatic rupture of the organ in a car accident. She had no gynecological or pathological history. Her menses were normal with a 28-day-cycle and duration of five days.

The patient brought the results of the US of her genitalia and CT of the upper and lower abdominal area.

US revealed that the uterus was of normal size, with 9 mm thick endometrium. In the left adnexa a firm tumor measuring 35 x 40 mm was found but there was no fluid in the Douglas space. Upper and lower abdominal CT proved the absence of the spleen and confirmed the existence of a firm left adnexal tumor measuring 35 x 50 mm, with no other complementary pathological findings. The patient was hospitalized in the clinic for further investigation and management.

No pathological findings were discovered during physical examination and inspection of the external genitalia. The vagina and cervix were normal. The uterus was mobile, normal in size but mild sensitivity was found in the anatomic region of the left adnexa during the gynecological examination with both hands. The abdomen was smooth, easily compressed, with moderate pain.

The laboratory blood tests of the patient showed hematocrit 37%, Hgb 12.2 g/dl, platelets 200,000, WBC 12.5, slightly

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increased (INR 1.07). The remaining laboratory biochemical tests were absolutely normal. Tumor markers CA 125 and CA 19.9 were within normal limits. The presurgical diagnosis was left adnexal tumor. The patient was subjected to exploratory laparotomy. Exposure of the abdominal canity revealed a normal uterus with multiple angiomatous, blue-red lesions, measuring up to 1 cm in the anterior surface. The adnexa presented multiple varices on both sides and were normal in texture. On the surface of the large intestine serosa, as well as in the greater omentum, the same lesions as those in the anterior surface of the uterus were observed. Biopsy samples of various abdominal sites were taken and sent for rapid biopsy. It should be noted that the biopsy sampling was an extremely hemorrhagic procedure and that hemostasis was difficult. The frozen section results were negative for malignancy and indicated the existence of ectopic splenic tissue in the sampled lesions. Cytology lavage of the peritoneal fluid was also carried out and the cytological examination was negative. The patient was not subjected to total surgical desection of the foci and only selective biopsy sampling was performed.

Results

The final histological diagnosis verified the results of the rapid biopsy and determined the existence of ectopic splenic tissue with a surrounding capsule. The postsurgical course was normal with no symptomatology. The patient remained in the clinic for three days and was in good condition when released. Six months after the exploratory laparotomy the patient remains asymptomatic.

Discussion

Post-traumatic splenosis of the genitalia and peritoneal cavity is a rare clinical entity which presents characteristics common to different diseases such as endometriosis, benign and malignant adnexal neoplasms, hemangiomas of the gastrointestinal tract, lymphomas etc. [8]. The patient's history should be taken under serious consideration when traumatic spleen rupture or splenectomy is noted [6]. Apart from the history, the diagnosis of splenosis can be determined with the help of laboratory findings, such as the presence of Howell-Jolly bodies in peripheral blood, absence of spherocytosis and with imaging techniques – by means of scintigraphy and using Tc-99m tagged red blood cells [13]. Usually splenosis is found in multiple sites, is asymptomatic and most times is discovered accidentally [14]. Experimental studies have shown that subcutaneous fat, the greater omentum and peritoneum are the ideal tissues for autoimplantation of ectopic spleens [15].

As has already been mentioned, splenosis should be distinguished from the congenital abnormality of secondary spleens that originate from the posterior mesogastrium and are found the entire length of the gastrosplenic ligament [16], and rarely in the pancreas or in the pelvis [17]. Splenosis of the female genital tract should be taken under consideration in cases of non-diagnosed chronic pelvic pain, as well as in those with the presence of a pelvic mass of unknown origin in patients with splenectomy history [6]. The main difference between splenosis and endometriosis is that splenosis does not create adhesions [18].

If we consult the international literature we can see that intrapelvic splenosis may give symptomatology in the majority of cases compared to intraperitoneal splenosis where patients have a higher probability of not developing clinical symptoms [19].

It is impossible to predict which patients will develop splenosis post-traumatically [8].

The immunological significance of post-traumatic splenosis is still under dispute and study. It has been shown that patients who have been subjected to post-traumatic splenectomy have less risk of developing infection compared to patients undergoing selective splenectomy due to myelodysplastic syndrome [20]. A dangerous complication which is observed is the so-called OPSI (overwhelming postsplenectomy infection) is rapid post-splenectomy sepsis, with 60-80% morbidity in the first 48 hours [21]. This is due to post-traumatic autoimplantation of splenic tissue resulting in the maintenance of basic immunological functions such as phagocytosis, antibody production against pneumococcus and opsonophagocytosis. Studies report that roughly 20-30% of splenic tissue is required to have an immune response against bacteremia [20].

Complete surgical excision in patients with diagnosed disease is contraindicated laparoscopically. In recent years the destruction of splenosis foci has been performed with an argon laser [22]. When splenosis is diagnosed casually in symptomatic patients complete surgical excision should not be performed [8].

Splendosis represents a problem of differential diagnosis that may often lead to confusion and cause the patient to be subjected to pointless surgical operations [5].

Conclusions

Splenosis is a rare pathology which, in most cases, progresses asymptomatically. Its clinical performance has not been clarified and depends on the location. The diagnosis is difficult and misleading. Imaging findings many times lead the physician to an erroneous estimation and treatment. The history of post-traumatic splenectomy and accompanying pelvic pain should always be taken into consideration. In case of pelvic pain with vague etiology, surgical investigation should always be taken into account. In asymptomatic patients with a diagnosis of splenosis made casually surgical excision is contraindicated [23].

In general, this is a benign disease of immunological importance concerning the body's defenses.

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