Case Report

SPLENOSIS: AN UNCOMMON CAUSE OF RIGHT "ADRENAL INCIDENTALOMA." CASE REPORT AND REVIEW OF THE LITERATURE

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ABSTRACT

Objective: To present an interesting case of splenosis mimicking a right adrenal incidentaloma.

Methods: We discuss the clinical presentation, diagnostic evaluation, and management of this case and review the literature for reports of right-sided retroperitoneal splenosis mimicking an adrenal mass.

Results: A 4-cm right suprarenal mass was incidentally discovered during the work-up of abdominal pain in a 38-year-old female smoker with a history of hypertension and prior splenectomy following abdominal trauma 32 years earlier. Adrenal function tests were compatible with a nonsecreting adrenal incidentaloma. Imaging characteristics of the mass were not diagnostic. Diagnostic laparoscopy revealed ectopic splenic tissue, independent of the normal-appearing adrenal gland. Three cases of right-sided retroperitoneal splenosis mimicking an adrenal mass were identified in the literature and summarized.

Conclusion: Splenosis should be included in the differential diagnosis of any patient with a history of posttraumatic splenectomy presenting with an adrenal incidentaloma. **(AACE Clinical Case Rep. 2015;1:e100-e104)**

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Abbreviations:
CT = computed tomography; **HU** = Hounsfield units; **MRI** = magnetic resonance imaging; **NRR** = normal reference range; **PET** = positron emission tomography

CASE REPORT

A 38-year-old woman with a past medical history of hypertension, postviral cardiomyopathy, smoking, and posttraumatic splenectomy at 6 years of age, complicated by recurrent streptococcus infection thereafter, presented with flank and abdominal pain and was hospitalized. *Clostridium difficile* colitis was diagnosed from analysis of stool samples. However, contrast-enhanced computed tomography (CT) performed during initial evaluation demonstrated a 4.2-cm right suprarenal soft tissue mass with 2 separate nodules in the left upper quadrant, suggestive of splenules, measuring 2.5 cm below the diaphragm near the splenic fossa and 1.4 cm anterior to the greater curvature of the stomach; additional nodules were identified anterior to the pancreas and in the left lower quadrant. She denied any symptoms suggestive of adrenal hormone hypersecretion, and the physical examination was negative for any signs of hyperfunctioning adrenal tumor. Adrenal function tests were performed: aldosterone, 41 ng/dL (normal reference range [NRR], 4 to 31 ng/dL); plasma renin activity (PRA), 14.6 ng/mL/h (NRR, 0.65 to 5 ng/mL/h); aldosterone to PRA ratio, 2.8; serum cortisol at 8 am after overnight dexamethasone suppression test, 0.8 µg/dL; plasma norepinephrine, 502 pg/mL (NRR, 120 to 680 pg/mL); plasma epinephrine, 18 pg/mL (NRR, <60 pg/mL); plasma dopamine, <15 pg/mL (NRR, <87 pg/mL); plasma metanephrine, <25 pg/mL (NRR, <58 pg/mL); plasma normetanephrine, 108 pg/mL (NRR, <149 pg/mL). Additional diagnostic imaging examinations including magnetic resonance imaging (MRI) and positron emission tomography (PET)/CT were subsequently performed. MRI demonstrated an ovoid, smoothly marginated, well-

demarcated 4.2 (anterior-posterior) × 2.8 (transverse) × 3.6 (cranio-caudal) cm homogeneous soft tissue mass involving the right adrenal gland demonstrating intermediate T1-weighted signal intensity without evidence of signal suppression on opposed phase gradient echo sequences, moderately increased T2-weighted signal intensity, and mild diffuse homogeneous intravenous contrast enhancement without evidence of internal necrosis or hemorrhage. These findings were not consistent with a lipid-rich adrenal adenoma, and the size of the lesion (>3 cm) was felt to be larger than typically seen with incidental adrenal adenomas. Pheochromocytoma was considered but thought unlikely based on the imaging characteristics, including T2-weighted signal intensity and biochemical results. The lesion did not demonstrate bulk or macroscopic fat to suggest myelolipoma. Lesion homogeneity and well-demarcated margins argued against primary adrenal cortical carcinoma, but metastatic disease could not be excluded. Unenhanced CT performed with PET imaging demonstrated an internal attenuation value of 50 Hounsfield units (HU), whereas PET showed only minimal fluorodeoxyglucose (FDG) avidity, with standardized uptake value of 2.2; there was no evidence of primary neoplasm identified elsewhere. As such, the likelihood of malignancy was thought to be low but not entirely excluded. The absolute washout of the lesion was 50% (estimated retrospectively on hard copy films of the initial multiphasic CT scan using the formula: absolute washout = [enhanced HU – delayed HU]/[enhanced HU – unenhanced HU] × 100), indicating retention of contrast and therefore suggesting the possibility of malignancy such as metastasis. Representative diagnostic images of the findings in this case are presented in Figures 1 and 2.

Although past medical history and imaging findings did raise the possibility of splenosis in this case, persistent diagnostic uncertainty and lesion size greater than 3 cm precluded exclusion of a potential neoplastic process. Therefore, the patient agreed to proceed to the operating room for further evaluation. Laparoscopy revealed a purple encapsulated mass, independent of the normal appearing adrenal gland, which was dissected free from the retroperitoneum and resected. Frozen section pathologic evaluation was consistent with an accessory spleen. Microscopic evaluation showed unremarkable splenic tissue with normal white pulp and congested red pulp without any evidence of malignancy (Fig. 3).

DISCUSSION

Adrenal incidentalomas are found in approximately 4% of CT examinations performed for a variety of clinical indications (1). The differential diagnosis of adrenal masses is broad, as shown in Table 1. Most incidental adrenal nodules prove to be nonfunctional (~80%) or hypersecreting (6%) benign adrenocortical adenomas. Most functional adenomas show autonomous cortisol secretion (often subclinical), whereas others secrete aldosterone. The remaining cases are largely comprised of small percentages of silent pheochromocytoma, myelolipoma, primary adrenocortical carcinoma, and metastatic disease (2). Surgery is currently recommended for malignant lesions, functioning or symptomatic adrenal adenomas, and tumors over 3 to 4 cm in size with indeterminate or suspicious imaging characteristics (3).

In the present case, there was no evidence of autonomous hormone secretion. Testosterone and dehydroepiandrosterone sulfate were not measured, despite considering malignancy in the differential diagnosis, because the patient had no signs or symptoms suggestive of hyperandrogenism. In fact, the patient became pregnant during the study of the mass; however, the pregnancy was voluntarily terminated during the first trimester because it was

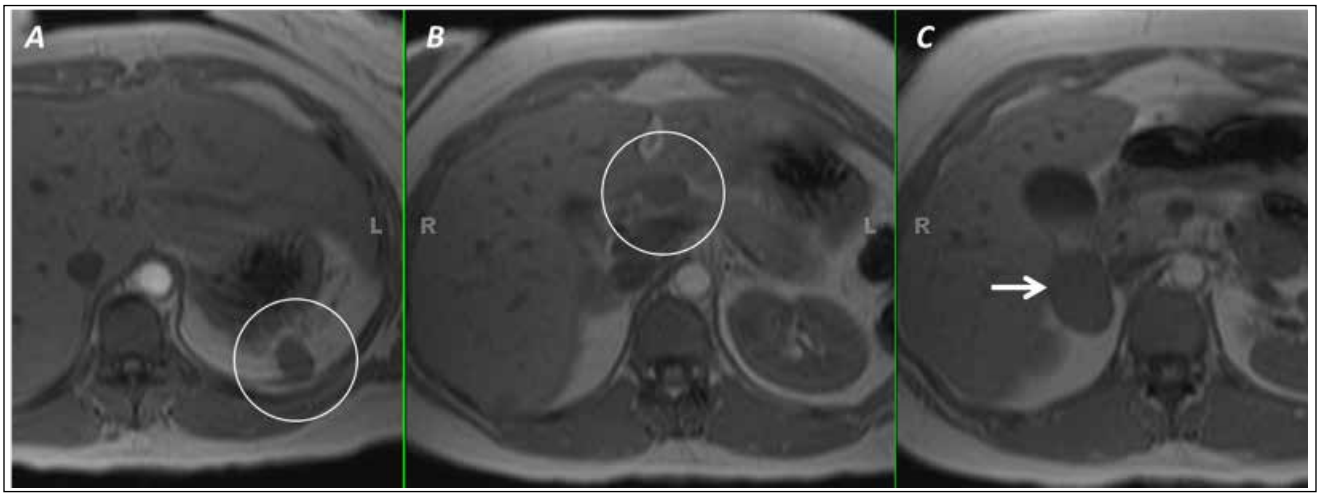

Fig. 1. Axial T1-weighted gradient echo magnetic resonance imaging (cranial to caudal) demonstrates (*A*) left subphrenic nodule, (*B*) peripancreatic nodule, and (*C*) right suprarenal mass.

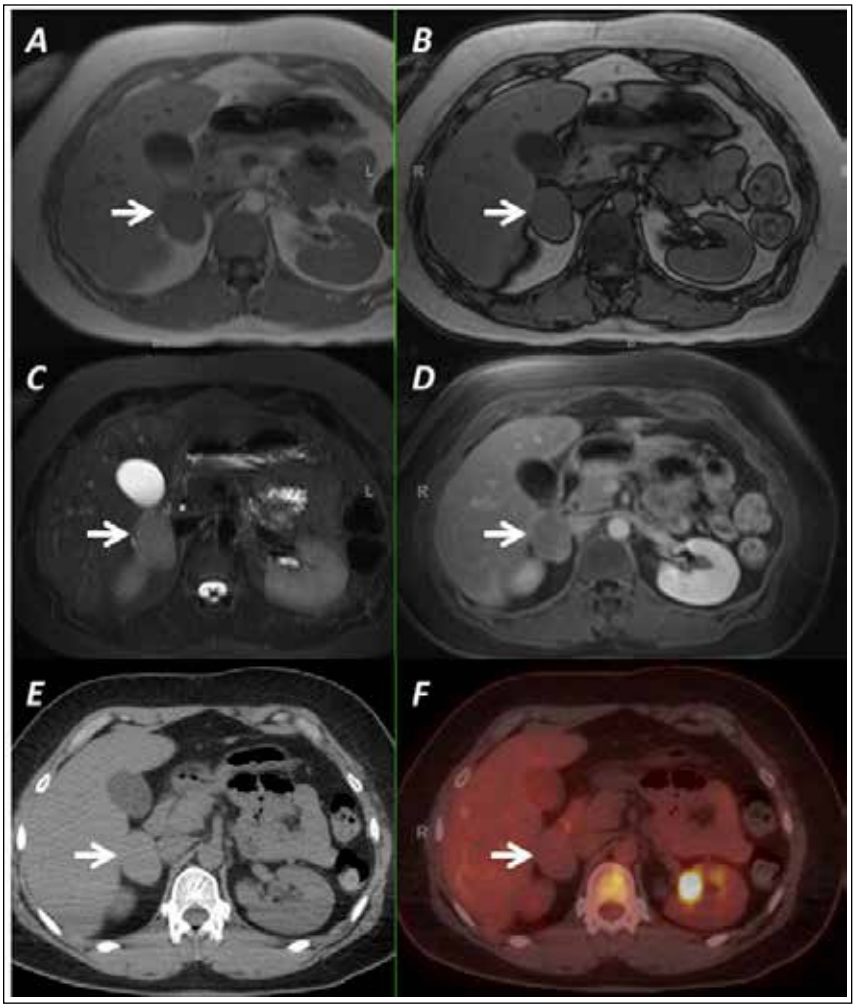

Fig. 2. (*A*, *B*) Axial in and opposed phase T1-weighted gradient recalled echo imaging demonstrates lack of signal suppression, which excludes lipid-rich adenoma. (*C*, *D*) Axial fat-suppressed T2-weighted and contrast-enhanced magnetic resonance imaging demonstrate homogeneous, moderately T2-hyperintense, mildly enhancing soft tissue mass. (*E*) Axial unenhanced computed tomography imaging demonstrates soft tissue mass with internal attenuation value of 50 HU. (*F*) Positron emission tomography/computed tomography fusion image demonstrates low-level fluorodeoxyglucose uptake.

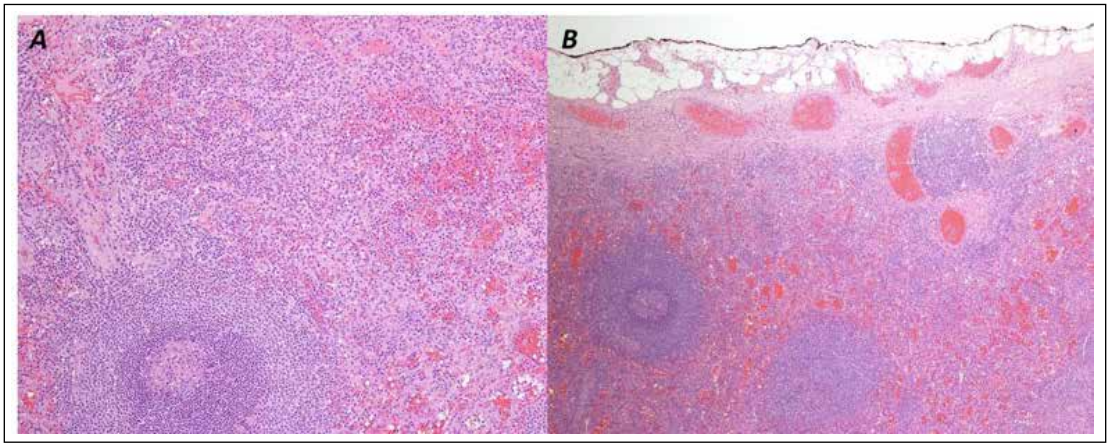

Fig. 3. (*A*) Sections show normal splenic tissue with red and white pulp (hematoxylin and eosin stain; 10× magnification). (*B*) Sections show normal splenic tissue with red and white pulp. The adjacent soft tissue margin of resection is inked black. No adrenal gland tissue is present (hematoxylin and eosin stain; 4× magnification).

Table 1
Differential Diagnosis of Adrenal Incidentaloma[a]

Benign nonfunctioning mass	**Hyperfunctioning mass**
- Adenoma	- Congenital adrenal hyperplasia
- Adrenolipoma	- Masculinizing or feminizing tumor
- Amyloidosis	- Nodular hyperplasia
- Cyst	- Pheochromocytoma
- Ganglioneuroma	- Preclinical Cushing syndrome
- Granuloma	- Primary aldosteronism
- Hamartoma	- Primary malignancy
- Hematoma	**Pseudoadrenal mass**
- Hemangioma	- Mistaken vasculature
- Infection (fungal, tuberculosis, echinococcosis, cryptococcosis, nocardiosis, paragonimiasis)	- Liver
- Leiomyoma	- Lymph nodes
- Lipoma	- Pancreatic mass
- Myelolipoma	- Renal mass
- Neurofibroma	- Spleen
- Pseudocyst	- Stomach mass
- Teratoma	- Technical artifact
Malignant nonfunctioning mass	
- Angiosarcoma	
- Ganglioneuroblastoma	
- Leiomyosarcoma	
- Malignant schwannoma	
- Metastatic carcinoma	
- Primary malignancy (adrenocortical carcinoma)	
- Primary malignant melanoma	

[a]Reprinted from (2), with permission from Elsevier.

unplanned and due to the high risks associated with pregnancy in the cardiomyopathy setting. The tumor exceeded 4 cm, and although imaging studies were not clearly suspicious for malignancy, findings did not allow for a diagnosis of lipid-rich adrenal adenoma or other benign entity. As such, surgical resection was offered to the patient. At laparoscopy, the lesion was clearly identified as a structure independent from the adrenal gland; splenic tissue was confirmed at pathologic examination. Of note, the washout rate of the lesion was not available at the time of the patient's evaluation but was calculated retrospectively on scanned hard copy films of a CT image for informative purposes only and therefore may not have been absolutely accurate. Having said that, the absolute washout rate, if available during the study period, would have lent support to surgical resection, as was performed in this case.

Splenosis, or ectopic autotransplantation of viable splenic tissue, has been reported in as many as 76% of patients after splenectomy for traumatic splenic rupture (4). It is usually asymptomatic and incidentally detected during unrelated diagnostic imaging. When this is the case, treatment is generally not indicated. Most implants are found in the abdomen or pelvis, most commonly in the left upper quadrant, but splenosis can occur anywhere within the peritoneal cavity. Retroperitoneal implants have been described, as have intrathoracic, intracranial, and subcutaneous deposits (5). Splenosis occurring in close proximity to the adrenal glands is rare but may mimic an adrenal tumor, especially when inseparable from the adrenal by imaging (as in the present case) or coincident with symptoms, such as uncontrolled hypertension, which might resemble hormone hypersecretion (6-10). Most previously reported retroperito-

neal splenules resembling an adrenal mass have been found on the left side ipsilateral to the spleen.

When splenosis is suspected, the absence of Howell-Jolly bodies in peripheral blood erythrocytes indicates the presence of functioning splenic tissue, and scintigraphy using Tc-99m heat-damaged erythrocytes or Tc-99m sulfur colloid is usually diagnostic. Although nuclear medicine imaging was suggested by the radiologist in this case, it was not performed, as the mass was indistinguishable from the right adrenal gland, mimicking a right adrenal neoplasm, an unusual location and presentation of splenosis.

To our knowledge, only 3 cases of right retroperitoneal splenosis have been reported (8-10). The patients in all 3 cases had experienced posttraumatic splenectomy many years before discovery of splenules (31, 24, and 36 years earlier, respectively) and were found with ultrasound during studies performed for abdominal pain, hematuria, or suspected pheochromocytoma due to refractory hypertension. In the prior case reports, the suprarenal masses were similar in size to the case presented here, ranging from 3.4 to 6 cm and were described as homogeneous and nonspecific on ultrasound or MRI (low signal on T1 and intermediate signal on T2). However, no other radiologic features were described, preventing further comparisons. Final diagnosis was achieved after surgery in 2 of the 3 cases. The other patient had end-stage renal failure treated with periodic hemodialysis and was not operated on due to the lack of radiologic features suggestive of malignancy, absence of hormonal activity, and inadequate control of the hypertension. This patient was followed with ultrasound 6 months later (which confirmed size stability) and was ultimately diagnosed using Tc-99m heat-damaged erythrocyte scintigraphy.

CONCLUSION

The finding of an adrenal incidentaloma in a patient with a history of posttraumatic splenectomy should raise the possibility of splenosis. Scintigraphy using Tc-99m heat-damaged erythrocytes or Tc-99m sulfur colloid might be indicated in such cases to rule out this rare condition. Consequently, surgery may be avoided for this entirely benign and asymptomatic condition.

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DISCLOSURE

The authors have no multiplicity of interest to disclose.

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