Case Report

Splenorenal fusion mimicking renal cancer: One case report and literature review

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Abstract

Splenorenal fusion is an extremely rare benign entity. This abnormality is presented in a case of a 29-year-old-male patient. We discuss the distinction between this condition and renal splenosis and their embryology. The course of this condition and modalities of investigation including radiological imaging, management, and pitfalls are reviewed.

Keywords: Accessory spleen, ectopic spleen, splenic heterotopia, splenorenal fusion, splenosis

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INTRODUCTION

Renal neoplasms are common and are easily diagnosed with the widespread use of the various imaging techniques. However, literature is scarce regarding the lesions mimicking renal neoplasms as subsequent histopathology and/or surgery prove a wrong radiologic diagnosis. These masses may be composed of normal or benign renal tissue (renal pseudotumors), but atypical renal masses may be resected unnecessarily for the concern of renal malignancy. Of these tumors, an accessory spleen in the form of a splenorenal fusion. Gonzalez-Crussi *et al.* first reported splenorenal fusion in 1977. It presents in patients with no history of splenic trauma or splenectomy and can mimic primary renal neoplasms or metastatic disease.[1] It usually involves the left kidney, although the right side location has also been documented.[2]

CASE REPORT

We report a 29-year-old male patient with no medical, surgical, or traumatic history who was referred by his family physician to the urology outpatient clinic for an unexplained increased urinary frequency explored subsequently by an ultrasound showing a left upper pole kidney tumor. His blood work was normal. A computerized tomography (CT) scan revealed a 4-cm homogenous but hyperdense lesion located at the left kidney's upper pole [Figure 1] and a normo-anatomic spleen. Furthermore, a renal biopsy was noncontributive displaying richly vascularized granulation tissue and inflammatory cell infiltrates. After discussing the case in a multidisciplinary team meeting, the patient underwent a laparoscopic partial nephrectomy to manage a hypothetical malignancy suspected because of the large size of the tumor. The pathology report revealed a capsulated accessory spleen inside the kidney measuring 25 mm independent of the renal parenchyma [Figure 2]. The patient's renal function remained normal at a 1-year follow-up.

DISCUSSION

During the fifth embryonic week, the spleen arises as the result of a proliferation of peritoneal cells of the dorsal

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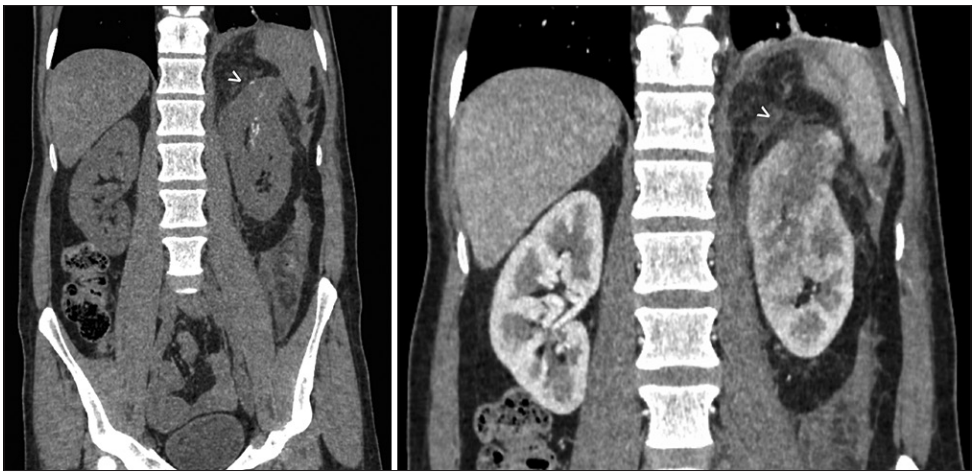

Figure 1: Coronal reconstructed computed tomography scan images. Left side: Showing the tumor (arrow head) with no contrast. Right side: Enhancement of the renal mass in the arterial phase (arrow head)

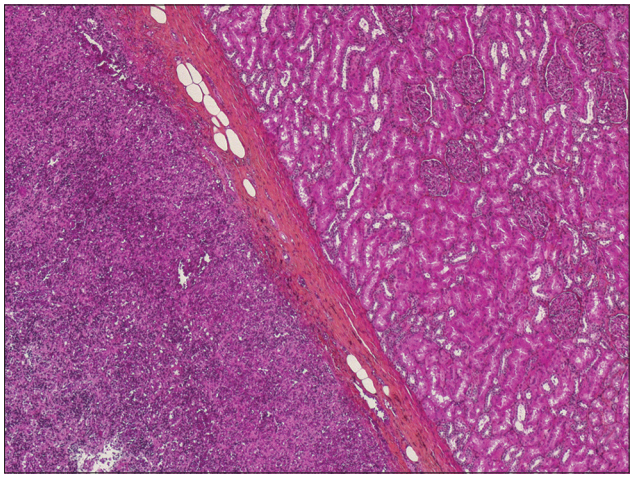

Figure 2: Histology of renal tissue (right side), which can be easily distinguished from splenic tissue (left side) separated by the capsule of the accessory spleen (H and E, ×40)

mesogastrium from mesenchymal cells that migrate between the leaves of the mesentery.[3] A failure of fusion of the mesenchymal cells results in accessory spleens.[4] In addition, as an outcome of the rotation of the embryonic gut, the splenic primordium lies close to the left mesonephric ridge.[5] There are two conflicting theories of spleno-visceral fusion.[6] The first one is the continuous theory that describes a band of tissue that adheres the spleen with the organ between the 7th and 8th week of gestation. The second theory is the discontinuous type that postulates migration of the splenic cells caudally to reach the developing mesonephric ridge and retroperitoneum, where fusion takes place. The only objection to this theory is that crossing of splenic tissue to the right side would be expected since there is no obstacle in the retroperitoneum.[2] The forms of ectopic splenic tissue (splenules) can be found as two types: accessory spleens and splenosis. Accessory spleens are congenital and arise from the left side of the dorsal mesogastrium during the embryological period of development.[5] Splenosis refers to splenic tissue that has been autotransplanted in a heterotopic location after splenic traumatism or splenectomy as defined by Beneke in 1910. We consider spleno-organ fusion a subtype of accessory spleens. Accessory spleens always conform to their embryological topographical limits even when reported within the pancreas, kidney, and scrotum and as an adnexal mass. Splenosis can be found in any location. Most usual locations (most frequent to less) are the peritoneum, omentum, and mesentery.[4] Splenosis has been reported in the pericardium, subcutaneous tissue, and even the brain. Epidemiologically, accessory spleens are found in up to 30% of the population.[2] The incidence of splenosis remains unknown, but its occurrence seems to be highest in males of young age that may reflect the increased capability of splenic cells to implant or it may simply represent the increased incidence splenic injury in young males. Most patients with splenic heterotopia are usually asymptomatic and found incidentally. Symptomatic patients may present with pain caused by infarction, intestinal obstruction due to the adhesive bands of the splenic implants, gastrointestinal hemorrhage, hydronephrosis caused by a mass compressing the ureter, as well as an enlarging abdominal mass. Patients can also present with symptoms of hypersplenism.[1] Radiological studies by ultrasound or CT scan and invasive laparotomies are all well-documented ways of incidental uncovering of ectopic spleens. Ultrasound examination will show round hypoechoic masses with homogenous echogenicity, and the nodules show a rim of a thin hyperechoic wall. CT scans show a uniform soft tissue density with homogenous, hypodense, contrast enhancement that differs clearly from the normal heterogeneity of the spleen.[7] None of these techniques is specific or sensitive. Recent reports have shown that ferumoxide-enhanced magnetic resonance imaging is a novel technique for diagnosing splenosis.[8] Ferumoxides

are superparamagnetic iron oxides that are removed from the circulation by the reticuloendothelial system making them tissue specific. The gold standard diagnostic modality of choice is nuclear scintigraphy (splenic scan). The ^{99m}technetium (^{99m}Tc) sulfur colloid test of the liver and spleen was first used to diagnose accessory spleens and/or splenosis due to its ability to localize the reticuloendothelial system.[3,5] Moreover, scintigraphy using ^{99m}Tc heat-damaged red blood cells (RBCs) or indium^{111}-labeled platelets is more sensitive and specific for splenic uptake, making these tests the current diagnostic tools of choice.[9] RBC scintigraphy was shown to be more sensitive than the sulfur colloid test in early splenosis, situations where minimal splenic tissue is present, functional hyposplenism, or poor splenic uptake or overlapping of the liver and spleen, causing poor visualization of splenic tissue.[4] Radiological-guided biopsy has been reported but was not useful for the diagnosis so far. Cytological smears alone may be insufficient since RBCs and white blood cells will appear on the slides and distinguishing the lesion from an organizing hematoma or a chronic inflammation can be cumbersome for pathologists, if not impossible.[5] The histology of an accessory spleen is identical to that of the spleen. Splenosis' tissue usually reveals bizarre architecture without a hilum and scantily formed capsules and tissue of any shape or size. Reports have described the tissue as lacking trabecular structures or having less elastic tissue than a normal one and presenting ill-formed or scarce white pulp with usual appearing red pulp. Two cases of splenosis described by Carr and Turk showed that the histology and immunohistochemistry were indistinguishable from that of a normal spleen.[10] Most reported cases of splenorenal fusion underwent total nephrectomy. To our knowledge, we report the first case treated by tumorectomy, thus preserving the kidney. In conclusion, splenorenal fusion is a congenital anomaly in which splenic tissue is present within the renal capsule. This condition mimics primary or secondary tumors on imaging. Although found incidentally by imaging techniques, it can present with symptoms. The diagnosis can be established with ^{99m}Tc-sulfur-colloid scan or ^{99m}Tc-labeled, denatured RBC scan. Recognition of this entity is important to avoid unnecessary surgery.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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