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The role of computed tomography in detecting splenic arteriovenous fistula and concomitant atrial myxoma

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Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Patient: Female, 45
Final Diagnosis: Arterio-venous fistula of the spleen
Symptoms: Lef-side abdominal pain
Medication: —
Clinical Procedure: —
Specialty: Surgery

Objective: Rare disease

Background: Splenic arterial-venous fistula and atrial myxoma are not rare cases but the co-existence of both lesions in the same patient is unpublished so far.

Case Report: A 45- year-old woman presented with vague left flank pain. She was initially scanned by B-dimensional echography, which revealed multiple enlarged hypo-echoic lesions in the splenic hilum. To further characterize the lesion, we performed computed tomography angiography (CTA). CTA showed dilatation of the splenic artery, and aneurismal dilatation of the splenic vein, associated with early opacification of the portal system. CTA showed also an intrasplenic venous aneurism, which was presumed to be the site of fistulous communication. Celiac arteriography confirmed the CTA findings. A left atrial mass was detected by cardiac echography, which was evaluated better by CTA, and was consistent with atrial myxoma. The patient underwent open surgery in different stage with resection of the atrial mass and spleen. The postoperative period was uneventful.

Conclusions: This is a unique case in the literature, showing the coexistence of a dual-pathology splenic arterial venous fistula and atrial myxoma.

MeSH Keywords: CT • Myxoma • Arteriovenous Fistula • Fistula

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Background

A splenic arterial-venous fistula (SAVF) is caused by an abnormal discharge of the distal splenic artery into the corresponding vein. This pathology should be especially suspected in cases when patients complain of a vague left flank pain associated with a loud murmur, or acute portal hypertension not related to chronic liver disease. They may be asymptomatic or lead rapidly to the development of portal hypertension [1,2].

Myxoma is the most common benign tumor of the heart. It is found accidentally, as in this case, or could present with various clinical signs [3,4]. To the best of our knowledge, SAVF with concomitant atrial myxoma represent a unique pathology, previously unreported in the literature.

Case Report

A 45-year-old woman presented with a vague left flank pain. The pain started 3 years ago but became more persistent and

was associated with a loud pulsatile murmur. The patient was a well nourished Caucasian female. The liver was not palpable and no signs of jaundice were observed. The rest of the physical examination was unremarkable. Heart rate was regular and rhythmic. Blood pressure was 110/80 mm Hg. The liver and kidney test results were normal, and the rest of laboratory test results were unremarkable. An ECG revealed normal sinus rhythm. The patient had no prior history of abdominal trauma, accidental injury, or abdominal surgery.

Abdominal echography showed multiple round hypo-echoic lesions in the splenic hilum, most likely representing dilated splenic vessels. CTA revealed a tortuous dilated splenic artery and aneurismal dilatation of the splenic vein associated with an early opacification of the portal system. The dimensions of the aneurysm were: cranial-caudal 57 mm, latero-lateral 43 mm, and antero-posterior 24 mm. CTA also showed an intra-splenic venous aneurysm, which was presumed to be the location of the fistula (Figure 1A–1C).

Celiac and splenic arteriographies were carried out and showed a tortuous splenic artery with a large venous aneurysm combined with premature portal and splenic vein filling and marked opacification of the spleno-portal axis (Figure 2A, 2B).

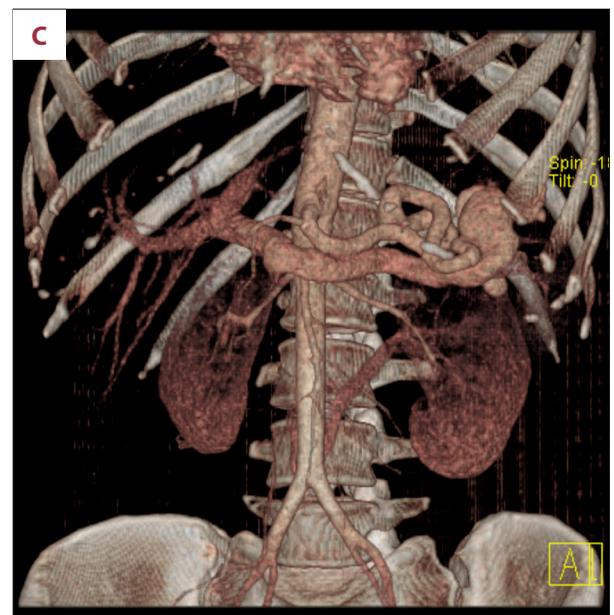
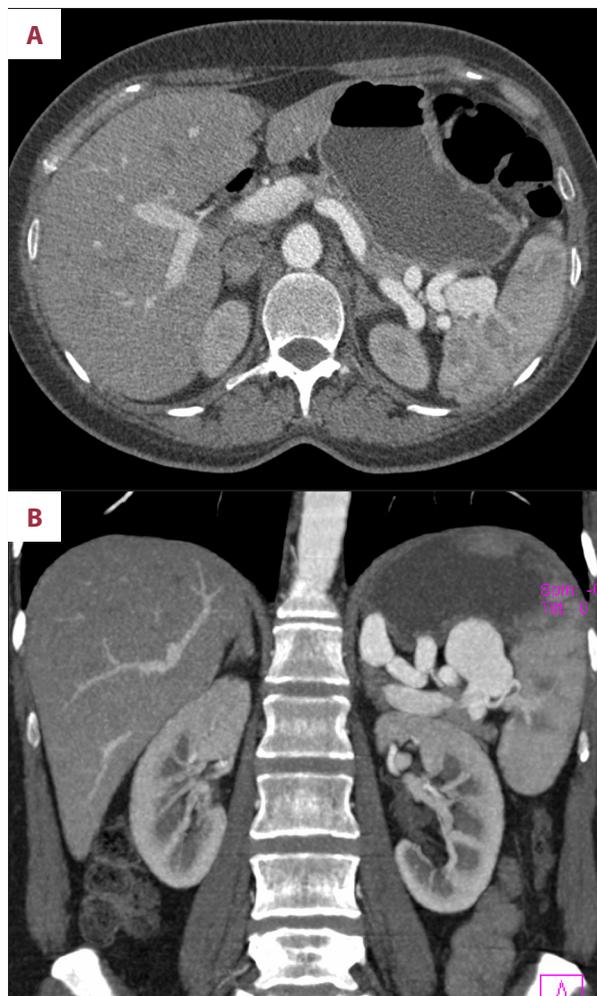


Figure 1. (A) Axial computed tomography in the arterial phase showed dilatation of splenic artery, aneurismal dilatation of the splenic vein, and small intra-parenchymal splenic aneurysm. (B, C) Reformatted maximum intensity projection (MIP) and volume rendering techniques (VRT) images in the coronal plane showed tortuous dilated splenic artery and multiple aneurismal dilatation of the splenic vein.

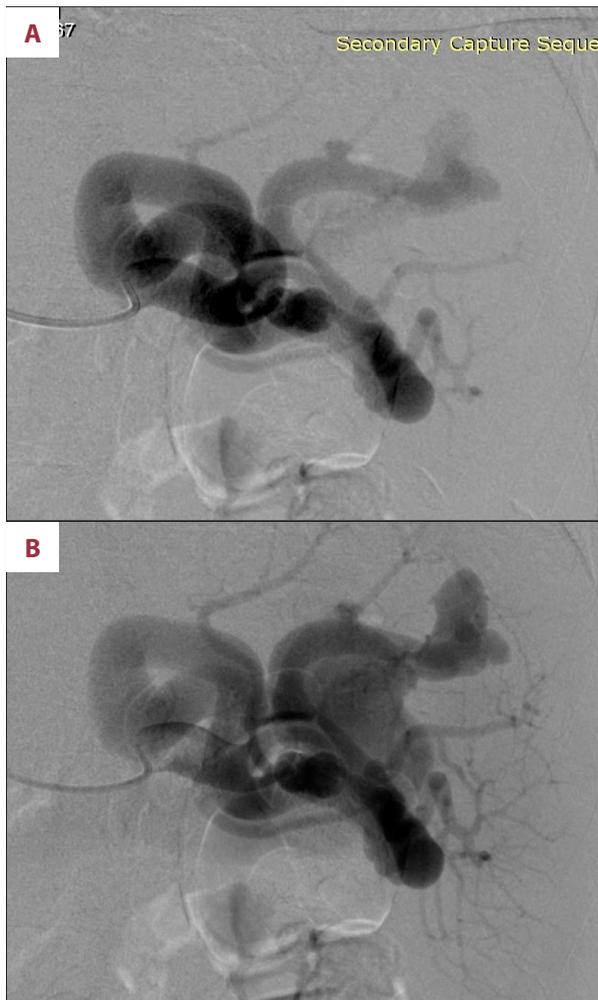


Figure 2. (A, B) Angiography showed a splenic arteriovenous fistula.

The patient was seen by the cardiologist to evaluate potential complications from SAVf. The cardiologist noticed that the patient had non-frequent palpitations, effort dyspnea, but no chest pain and normal cardiac function. Transthoracic echocardiography showed a large mass in the left atrium. Subsequently, the patient had a cardiac CT, which showed a large mass arising from the inter-atrial septum and protruding towards the mitral valve, consistent with atrial myxoma (Figure 3A, 3B). Based on the presence of both pathologies, surgical treatments were required.

Initially, the patient underwent a successful cardiac surgery. During the operation, a large mass was identified attached to the septum of the left atrium with a thin peduncle that measured approximately 1 cm. The mass was removed and the defect in the atrial septum was covered with an autologous pericardial patch. Several weeks after the cardiac operation, the patient underwent laparotomy with dissection of SAVf and splenectomy. The postoperative period was uneventful in both

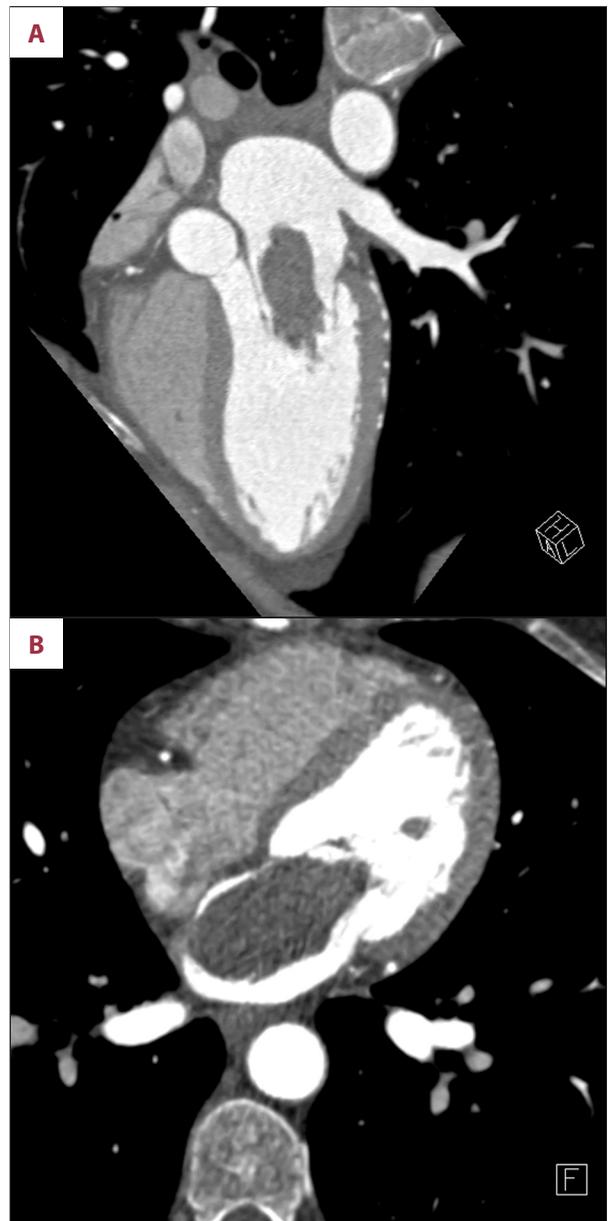


Figure 3. (A, B) Computed tomography showed a large atrial mass protruding towards the mitral valve.

cases. After several months of follow-up, the patient was doing well and did not present any complaints.

Discussion

SAVf was first described by Wiegert in the 19th century. Atrial myxoma is the most common type of benign cardiac tumor in adults. The literature does not reveal any connection between these 2 entities and we speculate that they are separate diseases, which were diagnosed at the same time. Further studies would be helpful to explore a possible connection between

these 2 pathologies. The mechanism of SAVf formation is an abnormal route of communication of a splenic artery aneurysm (SAA) with the corresponding vein. The predisposing factors for arterial aneurysm are increased blood flow and arterial wall damage associated with weakness of the smooth muscle layer. The cause can be congenital, infectious, due to pancreatitis, acquired after trauma or accidental penetrating injury, and prior surgery in the abdominal cavity. It is more common in multiparous women than in men, with a sex ratio 4 to 1 [1,2]. SAVFs may stay clinically silent for a long period of time. When the aneurysm is formed, the patient complains of discreet pain and bruit in the left flank, as observed in our patient.

Myxoma is mostly located in the left atrium, but in rare cases it is encountered in the right atrium. There is a recognized female predilection. The clinical presentation is variable; approximately 20% of myxomas are asymptomatic, or have general symptoms like weakness and weight loss. Depending on the degree of prolapse of the mass into the mitral valve, it could be associated with valvular obstruction, arrhythmia, or embolic diseases, particularly in the brain. The myxomas of the right side of the heart present with symptoms of heart failure. As the number of imaging diagnostic procedures is increasing, more myxomas are accidentally detected. They may be associated with multiple endocrine neoplasia (MEN) syndromes, known as the Carney triad [3,4].

The hemodynamic changes in SAVf may lead to a sudden increase of portal venous pressure, which is manifested with gastrointestinal bleeding, varices, and ascites. Portal hypertension due to chronic liver disease usually requires a longer period of time. Increased portal pressure is associated with splenomegaly and other gastrointestinal signs [5,6]. This patient had a normally sized spleen and liver. The liver had no parenchymal changes and the portal vein was normal.

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Cardiac symptoms have been reported by Gunther and include tachycardia, left ventricular dilatation, and ischemic disease of the myocardium [7]. Our patient had some cardiac symptoms that were more related to atrial myxoma.

Vague pain in the left flank was the most persistent symptom in our patient. She started complaining 3 years ago, but because of the vague nature and undetermined echographic findings, she had not been diagnosed. The estimated risk of rupture depends on the size of the aneurysm. There is an increased risk during pregnancy, but this was not the case for our patient [8].

Her past medical history was uneventful. Besides past pregnancies, we did not find any cause to speculate about the origins of this SAVf.

The CTA imaging findings were straightforward in both SAVf and atrial myxoma. The differential diagnoses of primary atrial mass include fibroelastoma, rhabdomyoma, fibroma, lymphoma, and sarcoma. Rhabdomyoma and fibroma mostly affect children, and the other tumors have different clinical and morphological features [9].

Nowadays, the treatment of SAVf is based on endovascular embolization [10]. Because of the complexity of this case, different stage operations were performed.

Conclusions

CTA is very accurate and helpful in delineating anomalies like SAVf and atrial myxoma and in prompt management. Further study need to be done to explore any connections between them.