Intravascular hemolytic anemia and renal failure caused by myxoma

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ABSTRACT

Background: Cardiac myxoma is the most frequent "benign" tumor of the heart and presents an important diagnostic challenge.

Case presentation: A 51-year-old woman was admitted at the hospital for anorexia, fatigue, nausea, and vomiting for the last 10 days. Laboratory results showed hemoglobin 5.26 g/dl, platelets 83× 10^9 µl, Lactate Dehydrogenase (LDH) 348 U/l, bilirubin 2.0 mg/dl, haptoglobin 100 mg/dl, negative Coombs test, blood smear with schistocytes and urea 327 mg/dl, creatinine 8.56 mg/dl. Non-autoimmune hemolytic anemia and acute kidney injury was assumed. After seven plasma exchange treatments, she went into acute pulmonary edema. Body tomography was performed, and revealed a mass in the left atrium. She underwent atriotomy and after surgery hemoglobin values stabilized but kidney function did not improve, and she became dialysis dependent.

Conclusion: Myxomas can mimic other diseases which may delay diagnosis and compromise prognosis. In this particular case, the myxoma manifested with intravascular hemolytic anemia. This is an, especially, interesting case because a myxoma caused kidney failure without recovery.

Keywords: Cardiac tumor, myxoma, embolization, renal failure, hemolytic anemia.

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Background

Primary tumors of the heart are rare but they are one of the major causes of patient morbidity and mortality and occur in as many as 3 in 1,000 patients [1].

Cardiac myxoma is the most frequent "benign" tumor of the heart and presents an important diagnostic challenge. Myxomas may resemble many cardiovascular or systemic diseases. There are no pathognomonic signs and symptoms that suggest the presence of a myxoma [2]. The cardiac myxoma may be silent until it has the volume to obstruct a heart valve or make a complaint and lead to emboli.

Case Presentation

A 51-year-old woman without previous history was admitted at the hospital for anorexia, fatigue, nausea, and vomiting for the last 10 days. The patient denied epidemiological context. Three days earlier she was prescribed symptomatic medication without improvement. At admission she was hemodynamically stable, apyretic, pale, and dehydrated.

Laboratory results showed hemoglobin 5.26 g/dl, platelets 83× 109 µl, LDH 348 U/l, bilirubin 2.0 mg/dl, haptoglobin 100 mg/dl, negative Coombs test, blood smear with schistocytes and urea 327 mg/dl, creatinine 8.56 mg/dl. Renal ultrasound evidenced normal-sized kidneys without changes.

Non-autoimmune hemolytic anemia and acute kidney injury was assumed. She did several red blood cell units, started hemodialysis, and due to the possibility of thrombotic microangiopathy she started therapeutic plasma exchange.

During the first week of treatment, she needed daily red blood cell units and thrombocytopenia progressed (84,000 × 109 µl). After seven plasma exchange treatments and absence of renal recovery, a renal biopsy was performed. During the procedure, she went into acute pulmonary edema. There was no improvement with medical measures or ultrafiltration. She was mechanically ventilated and transferred to the intensive care unit. Body tomography was performed, and revealed a mass in the left atrium with 5 cm and a splenic hematoma of about 10 cm (Figure 1).

On transesophageal echocardiography the image was very suggestive of myxoma in the left atrium. She underwent atriotomy and splenectomy. The specimen's analysis of the cardiac mass identified an atrial myxoma. After surgery, hemoglobin values stabilized but kidney function did not improve and she became dialysis dependent.

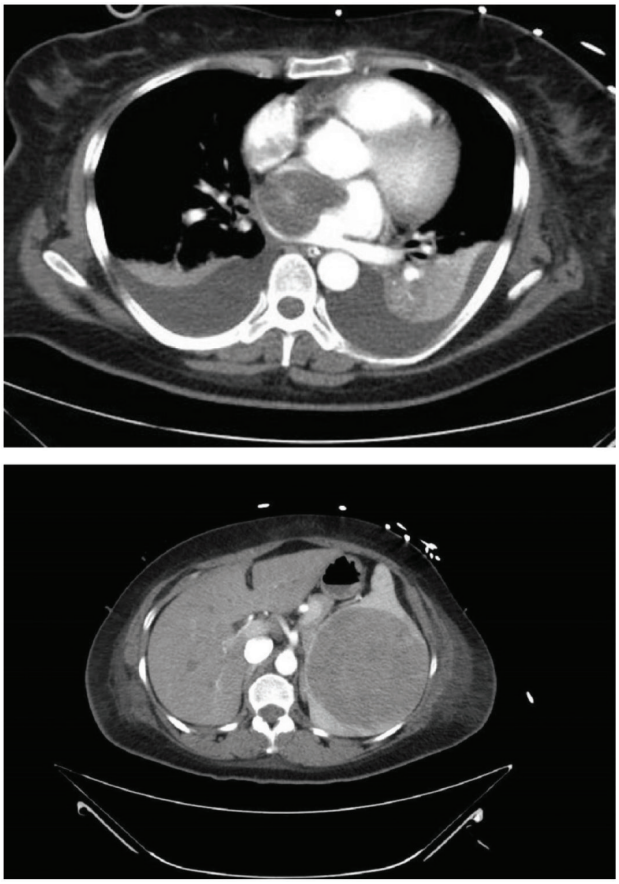

Figure 1. *Mass in the left atrium and a splenic hematoma.*

Discussion

Cardiac myxoma is the most frequent "benign" tumor of the heart and presents an important diagnostic challenge. Myxomas may resemble many cardiovascular or systemic diseases. There are no pathognomonic signs and symptoms that suggest the presence of a myxoma [2]

Histologically, these tumors are composed of scattered cells within a mucopolysaccharide stroma [3]. Macroscopically, they are pedunculated and gelatinous in consistency; the surface may be smooth, villous, or friable. Tumors vary widely in size, ranging from 1 to 15 cm in diameter, and weigh between 15 and 180 g [4].

The specific signs and symptoms of cardiac tumors generally are determined by the location of the tumor in the heart and not by its histopathology [5]. Approximately, 35% of myxomas are friable or villous, and these tend to present with emboli. Larger tumors are more likely to have a smooth surface and to be associated with cardiovascular symptoms. The cardiovascular manifestations depend upon the anatomic location of the tumor. Approximately, 80% of myxomas originate in the left atrium and most of the remaining are found in the right atrium [5]. In addition to their cardiovascular effects, patients with myxomas frequently have constitutional symptoms (e.g., weight loss and fever) [6]. Although the etiology of these symptoms is not fully understood, the production of various cytokines and growth factors by the tumor may contribute to these clinical [7].

Complete surgical removal of the myxoma and its cardiac attachment is usually curative; however, myxomas can recur, especially in patients with a familial myxoma syndrome [8].

The cardiac myxoma may be silent until it has the volume to obstruct a heart valve or make a complaint and lead to emboli [10]. When left atrial myxomas become symptomatic they can obstruct the mitral valve, embolize peripherally, or cause systemic effects [9].

In this particular case, the characteristic triad was verified: 1) Obstructive symptoms: unusual, rapidly progressive congestive cardiac failure (dyspnea); 2) Systemic emboli: renal failure and a splenic hematoma; and 3) Constitutional signs: fatigue and anorexia.

Embolism occurs in about 30%–40% of patients with myxomas. The majority of the emboli migrate to the central nervous system, but any arterial bed may be affected, leading to a great variety of symptoms and signs [2].

In this particular case, the myxoma manifested with intravascular hemolytic anemia, splenic hematoma and renal damage by multiple renal embolization's leading to renal failure.

In a young individual with no history of atrial fibrillation and evidence of systemic embolization, myxoma should be excluded. The gold standard exam is the echocardiogram. However, systemic embolization in most cases causes non-specific symptoms, such as those observed in this patient. Splenic hematoma was a finding on CT, as it was asymptomatic.

Although cases of kidney embolization have been reported, renal failure due to myxoma is rare, and fortunately, it is not common for patients to be dependent on dialysis as in this patient.

In summary, we present this case to remember that myxomas can mimic other diseases which may delay diagnosis and compromise prognosis. The clinical challenge is to consider the possibility of a cardiac tumor so that the appropriate diagnostic test(s) can be conducted. In addition, this is an especially interesting case because a myxoma caused kidney failure without recovery.

What is new?

This is an interesting case because a myxoma caused kidney failure without recovery. Although cases of kidney embolization have been reported, renal failure due to myxoma is rare. And fortunately, it is not common for patients to be dependent on dialysis as is the case with our patient.

Consent for Publication

Informed consent to publish this case report was obtained from the patient.

Ethical approval

Ethical approval is not required in Centro Hospitalar de Setúbal to publish an anonymous case report.

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Summary of the case

1	**Patient (gender, age)**	Woman, 51 years old
2	**Final diagnosis**	Cardiac myxoma
3	**Symptoms**	anorexia, fatigue, nausea and vomiting
4	**Medications**	Hemodialysis, Plasma exchange
5	**Clinical procedure**	Atriotomy and splenectomy
6	**Specialty**	Nephrology