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CASE REPORT

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Occam's razor versus Hickam's dictum: two very rare tumours in one single patient

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Abstract

Occam's razor, the principle that a single explanation is the most likely in medicine, assumes that when a patient has multiple symptoms the clinician seeks a single diagnosis rather than diagnosing multiple and different ones. However, as proposed by Hickam's dictum, sometimes rare different diseases occurred in only one patient.
We present a patient with a simultaneous diagnosis of two rare tumours, a cardiac hemangioma (primary cardiac tumour, often misdiagnosed as myxoma) and an appendiceal mucocele (a lesion of the appendix that can be neoplastic or not).
A 71-year-old male presented with anorexia, asthenia, fever and weight loss for about one month. During the etiological investigation, a cardiac mass and an appendiceal lesion were detected and both lesions required surgical intervention. Cardiac and abdominal surgeries were uneventful and full recovery was achieved. The histological examination showed a cardiac hemangioma and a neoplastic appendiceal mucocele.

INTRODUCTION

Primary cardiac tumours are rare and usually benign. Cardiac hemangioma is one of the rarest, accounting for ~2.8% of all primary resected heart tumours. In the absence of specific symptomatology and despite established imaging features, most cardiac hemangioma are misdiagnosed as myxoma, the most frequent primary cardiac tumour. As such, pathologic confirmation is required.

Appendiceal mucoceles are also rare and characterized by a distended, mucus-filled appendix. The course and prognosis of appendiceal mucoceles relate to their histologic subtypes.

In this case report, we present the synchronous diagnoses of these two lesions in a patient that required urgent surgical intervention increasing the complexity of the clinical approach.

CASE REPORT

A 71-year-old male presented to the emergency department with a story of anorexia, asthenia, fever and weight loss (≅10 kg) for about one month. He also reported a single episode of a few seconds duration of sudden nausea and diaphoresis while standing, but with no loss of consciousness. He had medicated mild hypertension. He already had a Thoraco-abdominopelvic computed tomography (CT) showing (i). a mass with 29 × 25 mm in the distal inferior vena cava (IVC) lumen involving the right atrium (RA) (Fig. 1A and B), (ii) an aneurysm of the ascending aortic with 57 mm and (iii) a very enlarged appendix (60 × 25 mm) (Fig. 1C).

A grade III systolic murmur was audible throughout the precordium. Electrocardiogram showed first-degree atrioventricular (AV) block. Laboratory blood tests revealed leukocytosis (11.38 × 10^9 cels/L) and elevated C-reactive protein (6.8 mg/dL).

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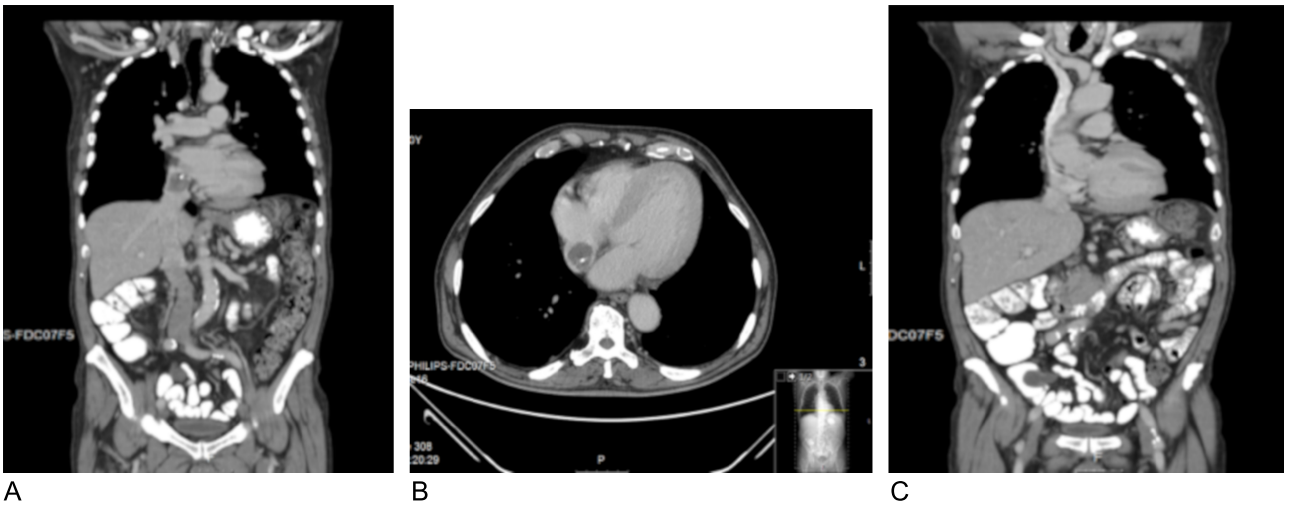

Figure 1: (A–C): Thoraco-abdomino-pelvic CT. Mass in the distal IVC (panels A and B) and a very enlarged mucinoid appendix (panel C).

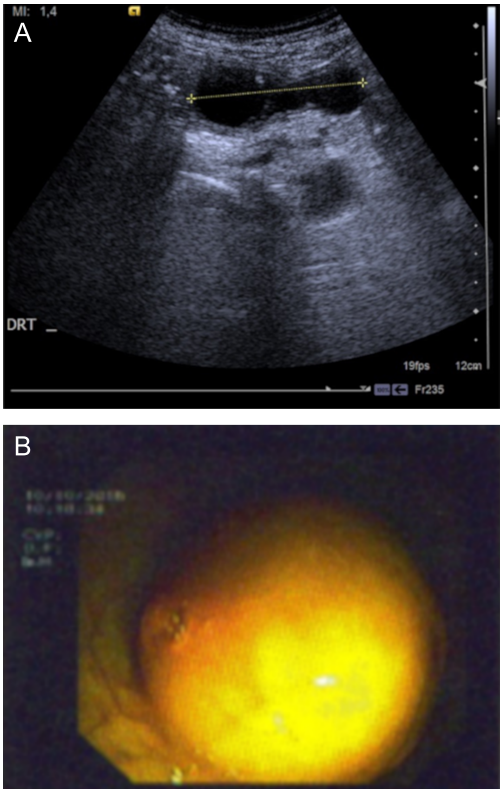

Figure 2: Abdominal US (panel A) and colonoscopy (panel B).

A transthoracic echocardiography was performed, which confirmed a mass in the IVC; this prolapsed into the RA and detected a severe stenotic bicuspid aortic valve.

The patient was admitted to an internal medicine ward. Blood cultures were collected and with suspicions of an abdominal infection, ceftriaxone was started. *Streptococcus gallolyticus* was recovered and antibiotic therapy was descalated to penicillin G.

An abdominal ultrasound (US) was performed that was suggestive of appendiceal mucocele (Fig. 2A), identified, in a colonoscopy, as a submucosal structure measuring roughly 6 cm (Fig. 2B).

With a transesophageal echocardiography, the mass that was previously identified as originating from the IVC was shown to originate from the RA. This was a mobile, appendicular mass, connected to the interatrial septum and protruding into the IVC. Rather suggestive of an RA myxoma (Fig. 3A). Aortic aneurism (>50 mm) as well as a stenotic bicuspid aortic valve were noted. A cardiac magnetic resonance was performed showing a heterogenous, irregular and rounded mass with an intermediate signal and a slight uptake of contrast. This reinforces the diagnostic hypothesis of a myxoma (Fig. 3B). A coronariography excluded coronary disease.

A surgical plan for the excision of the atrial mass and of the appendix was made. The patient completed 15 days of antibiotic therapy with clinical and laboratory resolution. A repeated set of blood cultures were negative.

Before the surgical procedure, a 2:1 and 3:1 AV block was developed. Therefore, cardiac surgery was anticipated. The atrial mass was resected; aortic aneurism was repaired and aortic valve was replaced ('Bentall' procedure). The patient had an uneventful recovery.

A second surgery was performed to resect the appendix, but because it was strongly adherent to the colon, a right hemicolectomy was performed. Again, an uneventful recovery was noted.

The histopathology of the atrial mass revealed a cavernous hemangioma. An appendiceal mucocele with neoplastic characteristics was found in the resected bowel (Fig. 4A and B).

DISCUSSION

We presented a patient with a clinical diagnostic of two very rare tumours, which were proven to be independent through a histological examination of the surgical specimens—a cardiac hemangioma and an appendiceal mucocele. The patient presented with constitutional symptoms, probably related to the bacteremia, which were believed to be secondary to bacterial translocation due to the appendix overdistension.

In a multidisciplinary conference, a decision to remove both lesions was taken. Because of the heart block rhythm, cardiac surgery was the priority. Both surgical interventions were uneventful.

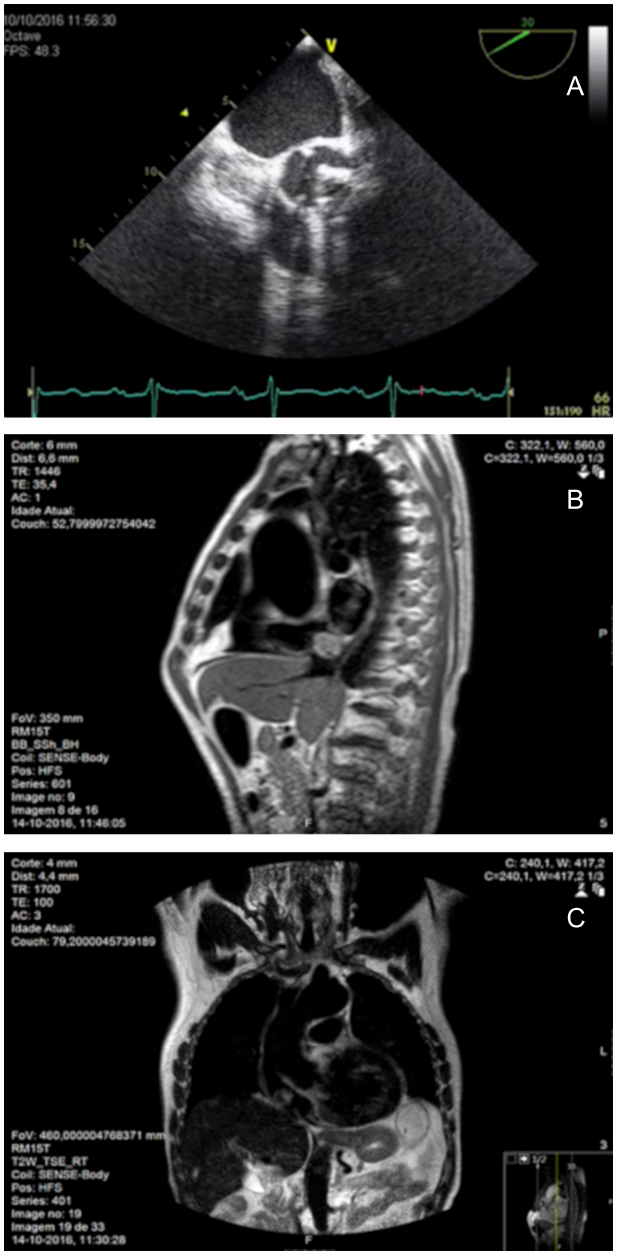

Figure 3: Transesophageal echocardiography (panel A) and cardiac magnetic resonance (panels B and C).

According to a comprehensive review, only 202 cases of cardiac hemangioma were described between 1950 and 2014 [1] constituting ~2.8% of the benign cardiac tumors. It can arise from any heart chamber, being the RA the predominant location. It can be pedunculated and mobile as atrial myxomas. Most are asymptomatic [1]. Echocardiography, both transthoracic and transesophageal, are the first-line imaging test. Coronary angiography is also important to map the feeding vessel, since tumour blush is a typical sign of cardiac hemangioma [1]. These tumours are usually misdiagnosed as atrial myxomas (65.9%) [1], as in our patient.

Cardiac hemangiomas may involute, stop growing or proliferate. Although these are benign tumours, they may metastasize or reoccur later in life [1, 2]. Surgical indication remains controversial in asymptomatic tumours, without structural or functional involvement, because of the surgical risks [1]. Yet, it should be removed when the tumor is confined to the septum (interatrial or interventricular), because of the involvement of the conduction pathways, which may lead to severe rhythm complications (as in our patient) or sudden death. The low incidence of adverse post-operative and long-term events suggests a more proactive surgical approach. This offers an extremely favourable long-term prognosis for the patient.

After the recovery of the cardiac surgery, our patient underwent abdominal surgery to remove the suspected appendiceal mucocele (associated with a rupture risk).

Again, appendiceal mucocele is a rare disease with an incidence of only 0.2%–0.7% of all excised appendices [3]. It can be a non-neoplastic or neoplastic is usually asymptomatic and may cause nonspecific symptoms. Ultrasonography is the first-line diagnostic method, but CT has higher accuracy [4]. On colonoscopy this tumour appear as a glossy, rounded protruding mass arising from the appendiceal orifice; a definitive diagnosis is made by histology.

Because of the risk of rupture with development of pseudomyxoma peritonei, with poor outcome, the appendiceal mucocele should be approached surgically. Given the great risk of developing an adenocarcinoma of the colon, colonic surveillance is warranted [5].

Our case illustrates one common dilemma of clinical practise. As referred to Hickam's dictum, sometimes a patient can have as many diseases as he pleases, and even rare ones should be looked for.

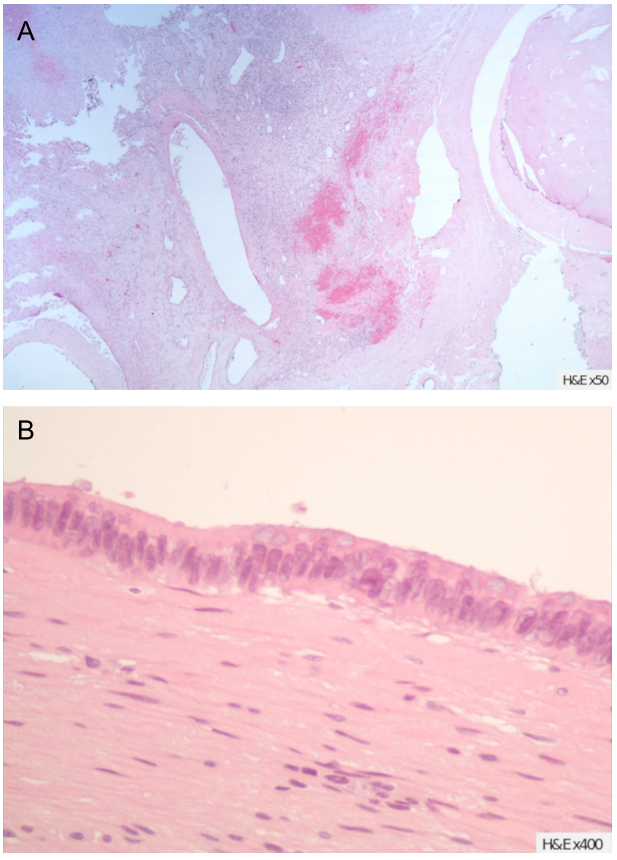

Figure 4: Histological findings. Microscopic viewing of the atrial tumour (panel A) and appendiceal mucocele (B).

CONFLICT OF INTEREST STATEMENT

No conflicts of interest.

FUNDING

Not applicable.

ETHICAL APPROVAL

None required.

CONSENT

We obtained written informed consent from the patient for the publication of this case report and the accompanying images.

GUARANTOR

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