Patient in chronic hemodialysis with right atrial mass: thrombus, fungal endocarditis or atrial myxoma?

Paciente em hemodiálise crônica com massa em átrio direito: trombo, endocardite fúngica, ou mixoma atrial?

We present the case report of a 19-year-old patient with chronic kidney disease due to chronic glomerulonephritis, in hemodialysis (HD) by central catheter, with the incidental finding of a mass of 28x16 mm in right atrium (RA). The diagnosis of thrombus, infective endocarditis or myxoma were considered. Given the context of immunosuppression and difficult access vascular therapeutic practice has proved complex. Although Doppler echocardiography suggested thrombus in RA, nuclear magnetic resonance imaging (MRI) indicated for the diagnosis of myxoma in RA. In both conditions, the proposed surgical approach was limited by intense immunosuppression history and the risk of infectious complications. Throughout the treatment, the general state of K.M.F. remained satisfactory and revealed no signs or symptoms related to atrial dysfunction. The absence of fever and negative blood cultures excluded infective endocarditis. Prior echocardiogram report without masses in the RA decreased the chance of cardiac myxoma. The therapeutic response to anticoagulation confirmed the diagnosis of thrombosis. After 180 days of anticoagulation, there was significant reduction in mass. The patient developed asymptomatic. The diagnosis of mass in RA can be a challenge and only the evolution of the case was able to guide the appropriate conduit. While MRI has high sensitivity and specificity for the diagnosis of cardiac myxoma, the interpretation of images can be subjective. Controversial point is the removal of the catheter in such cases, which is subject discussed throughout the report.

Keywords: catheters; endocarditis; myxoma; renal dialysis; thrombosis.

Resumo

Apresentamos o relato de caso de uma paciente de 19 anos com doença renal crônica devido à glomerulonefrite crônica e em hemodiálise (HD) por cateter central, com o achado incidental de uma massa de 28x16 mm em átrio direito (AD). Foram considerados os diagnósticos de trombo, endocardite infecciosa ou mixoma. Devendo ao contexto de imunossupressão e dificuldade de acesso vascular, a condução terapêutica revelou-se complexa. Apesar de Ecodoppler cardiograma sugerir trombo em AD, imagens de ressonância nuclear magnética (RNM) apontaram para o diagnóstico de mixoma em AD. Nas duas condições a proposta de abordagem cirúrgica esteve limitada pelo histórico de imunossupressão intensa e o risco de complicações infecciosas. Ao longo do tratamento, o estado geral de K.M.F. manteve-se satisfatório e não foram observados sinais ou sintomas relacionados a disfunção atrial. A ausência de febre e hemoculturas negativas excluíram endocardite infecciosa. O relato de ecocardiograma prévio sem massas em AD tornou menor a possibilidade de mixoma cardíaco. A resposta terapêutica à anticoagulação confirmou o diagnóstico de trombo. Após 180 dias de anticoagulação, houve redução significativa da massa. A paciente evoluiu assintomática. O diagnóstico de massa em AD pode ser um desafio e somente a evolução foi capaz de guiar a conduta apropriada. Apesar da RNM ter elevada sensibilidade e especificidade para o diagnóstico de mixoma cardíaco, a interpretação de imagens pode ser subjetiva. Ponto controverso é a retirada de cateter nesses casos, que é assunto discutido ao longo do relato.

Palavras-chave: cateteres; diálise renal; endocardite; mixoma; trombose.
INTRODUCTION

A significant portion of the individuals enrolled in hemodialysis (HD) programs without a definitive vascular access (e.g.: arteriovenous fistula) use long-term indwelling catheters. Prolonged use of indwelling catheters has been associated with severe mechanical complications and infection.1-3

A right atrial mass in an individual on hemodialysis is usually one of three possibilities: (a) a thrombus, (b) vegetation as a sign of infectious endocarditis, or (c) a heart tumor, most likely a myxoma.4 Once these diseases call for different therapies, improper diagnosis may lead to unnecessary invasive procedures, delayed start of treatment, and increased morbidity and mortality.

This case report illustrates the diagnostic challenges physicians face when finding a right atrial mass located contiguously to the HD catheter on a transthoracic echocardiogram (TTE) taken as part of the annual examination protocol in effect at the dialysis center. The report also aims to show the key role proper follow-up and therapy play in the production of a good clinical outcome.

CASE REPORT

K.M.F., female, 19, had systemic vasculitis with renal and pulmonary involvement. Despite immunosuppressant therapy with steroids and cyclophosphamide, the patient progressed to renal failure. She was started on HD with a short-term catheter at the Hospital of the University of Campinas (UNICAMP) in Brazil. The patient had to undergo four catheter changes due to catheter malfunction or infection. Once the infection was eradicated, the patient had a long-term indwelling catheter implanted.

A TTE was ordered seven months after the implantation of the long-term catheter, and a mass measuring 28 x 16 mm was found in the patient’s right atrium. The mass was initially believed to be a thrombus (Figure 1A). The patient was on prednisone 10 mg/day and did not have fever or systemic symptoms, and her general condition was good. Heart auscultation was normal. The patient was not fatigued and did not have hepatomegaly, peripheral edema or signs of pulmonary embolism. A nuclear magnetic resonance (NMR) scan of the heart was ordered to verify the initial diagnostic suspicion. The scan showed a solid round non-moving pedunculated mass on the floor of the right atrium close to the inferior vena cava, measuring 25 x 24 x 20 mm and suggestive of an atrial myxoma.

No microorganisms were detected in her blood cultures. The patient was fever-free and generally in good shape. The thoracic surgery team assessed her and the risk of removing the mass led the attending physician to choose anticoagulation with warfarin as a therapeutic test for the diagnosis of a thrombus. The long-term catheter was kept in place, as finding a new HD vascular access would not be easy. A control transesophageal echocardiogram (TEE) performed 100 days after the start of treatment (Figure 1B) showed the right atrial mass had shrunk to 12 x 10 mm.

The patient was symptom-free and was kept on warfarin for another 90 days. TEE images produced after six months of anticoagulation revealed a thickening of the Eustachian valve, which corresponded to remnants of the thrombus measuring 7 x 8 mm. She was offered an AVF and the long-term catheter was removed. The patient is asymptomatic and in good general condition. She had no warfarin-related adverse events, and was kept on anticoagulation therapy for another three months.

DISCUSSION

The following diagnostic possibilities were considered for the patient’s right atrial mass: thrombus, infectious endocarditis, and atrial myxoma. Infectious endocarditis was ruled out based on the patient’s overall satisfactory progression, the absence of fever, and blood cultures negative for bacteria and fungi. Although the NMR scans suggested she had an atrial myxoma, her previous TTE images did not show cardiac anomalies, which made a cardiac tumor a more remote possibility. However, atrial myxomas are known to develop within short periods of time due to their composition with scattered cells within a mucopolysaccharide stroma.5 The lack of constitutional symptoms and her response to warfarin therapy supported the diagnosis of a right atrial thrombus.

Although low in the general population, the incidence of right atrial thrombi increases in individuals using central venous catheters.1,2,6,7 The patient described in this case had an HD catheter implanted in her right internal jugular vein. The complications more commonly associated with right atrial thrombi are pulmonary embolism, septic
Right atrial mass in a patient on hemodialysis

Figure 1. A. Transthoracic echocardiogram (TTE) showing right atrial mass measuring 28 x 16 mm consistent with thrombus. B. Control TTE showing a significant reduction on the mass to 12 x 10 mm.

embolism, endocarditis, arrhythmia, mechanical cardiac complications, and systemic embolism in individuals with a patent foramen ovale.

The possible pathophysiological mechanisms involved in the formation of right atrial thrombi associated with HD catheterization include the activation of the coagulation cascade secondary to trauma to the atrial wall by the tip of the catheter. Local conditions are exacerbated by hemodynamic changes, which lead to the stagnation of blood flow in the right atrium in the area around the catheter. The location of the tip of the long-term indwelling catheter inside the right atrium - as per the guidelines of the National Kidney Foundation: Dialysis Outcomes Quality Initiative - has been strongly associated with the formation of thrombi.

TTE and TEE are the most commonly used tests to diagnose and follow the progression of right atrial masses. The specificity and sensitivity of echocardiography in the diagnosis of intracavitary masses are 86% and 95%, respectively. However, NMR is the preferred method for the differential diagnosis of right atrial masses, as it is highly specific (100%) for the detection of heart tumors.

In our case, the diagnoses suggested by the imaging tests differed (TTE for thrombus and NMR for myxoma), thus forcing the assisting team to make a call based on clinical findings. This shows that despite highly specific tests the interpretation of the scans still plays a significant role in achieving a diagnosis.

Treatment options range between offering anticoagulants or thrombolytic agents associated or not to catheter removal. Anticoagulation therapy for patient with kidney disease can be challenging. Elliott et al. found that warfarin is associated with significant risk of bleeding in patients on HD. Additionally, the use of vitamin K antagonists such as warfarin has been associated with vascular calcifications caused by the inhibition of vitamin k-dependent proteins such as the matrix-GLA protein.

Thrombectomy is indicated in cases suspected for infection or concomitant cardiac anomaly. A study showed that individuals with autoimmune disease and recent use of immunosuppressants are assigned higher levels of surgical risk and develop greater numbers of post-op complications, including myocardial infarction, heart failure, pneumonia, sepsis, and wound dehiscence, particularly those on high-dose oral prednisone before surgery. This finding supported the decision to treat our patient conservatively.

A meta-analysis with 71 patients looked into the prognostic factors for death and the treatment options for catheter-induced thrombi in patients on HD. The authors found that the factors associated with poorer outcomes were age, time on dialysis, and permanence of the catheter after diagnosis. The authors proposed a clinical management algorithm in which first-line therapy included anticoagulants and the use of a guide wire in catheter removals and changes. The authors of the meta-analysis also stated that catheters should be removed after the start of anticoagulant therapy and patients should be kept on anticoagulants for six months or until their condition is entirely resolved.
Surgery was recommended as the best option for patients suspected for infection, cardiac anomalies, or with contraindications to anticoagulation treatment. Another systematic review with 177 individuals diagnosed with right heart (atrial and ventricular) thrombi reported death rates of 28.6%, 23.8%, and 11.3% for patients offered anticoagulant therapy, embolectomy, and thrombolysis, respectively. All individuals not offered treatment died. The authors indicated that thrombolytic therapy was associated with lower death rates when compared to other treatments.

**Conclusion**

Diagnosing right atrial masses can be challenging. However, patient clinical progression may be used to guide the choice of either conservative or surgical treatment, as shown in the case reported herein. There is no consensus in the literature as to the best time to remove long-term indwelling catheters or the treatment of thrombi associated with catheterization. The recommendations described in the literature suggest care should be individualized and based on the information published in prospective randomized trials.

**References**

11. Forauer AR, Theoharis C. Histologic changes in the human vein wall adjacent to indwelling central venous catheters. J Vasc Interv Radiol 2003;14:1163-8. DOI: http://dx.doi.org/10.1097/01.RVI.0000086531.86489.4C