Clinical Case Reports

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CLINICAL IMAGE

Catheter-related right atrial thrombus in sickle cell disease

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Key Clinical Message

Catheter-related right atrial thrombus (CRAT) can occur in patients with sickle cell disease, particularly if additional risk factors for thrombosis are present. Cardiac MRI may differentiate thrombi from other types of atrial masses. Treatment should include anticoagulation and the timing of catheter removal should balance the potential risk of embolization.

Keywords

Anticoagulation, catheter-related right atrial thrombus, MRI, sickle cell disease.

A 27-year-old African American female with Hemoglobin-SS sickle cell disease presented as a new patient to clinic for an evaluation for eligibility for hematopoietic stem cell transplant. Her medical history includes frequent sickle cell vaso-occlusive crises requiring monthly exchange transfusions via bilateral in-dwelling catheters (port-a-caths). She has a history of provoked pulmonary embolism diagnosed during an acute chest syndrome episode in the setting of oral contraceptive use, catheter-associated thrombus, two-first-trimester miscarriages (no live births), ulcerative colitis, and primary sclerosing cholangitis. Prior hypercoagulability testing, including antiphospholipid testing, prothrombin time, and activated partial thromboplastin time, were within normal range. The patient was asymptomatic in clinic, but physical examination revealed a grade II/VI holosystolic blowing murmur throughout the precordium. Transthoracic echocardiogram demonstrated a right atrial, multilobulated, highly mobile mass (2.3 cm × 4.9 cm) (Fig. 1A) at the tip of a catheter (Fig. 1B).

The cardiac magnetic resonance imaging (MRI) revealed three separate right atrial thrombi. Thickening of the cross-sectional area of at least one of the catheters was visualized in the superior vena cava, which suggested fibrosis or possible layered thrombus. In addition, thrombus was attached to the tip of a catheter (Fig. 2A). Tissue characterization excluded lipoma (Fig. 2C). Additionally, there was less contrast uptake in the mass than myocardium, with only minimal late gadolinium enhancement, which is consistent with thrombus (Fig. 2C and D). Repeat antiphospholipid testing showed positive lupus anticoagulant. She was started on anticoagulation and the catheters were not removed due to concern of embolization risk during removal. Although direct oral anticoagulation is recommended to patients with catheter-related thrombosis, enoxaparin was started based on the patient's risk of embolization requiring emergent surgery. A repeat echocardiogram 1 week later demonstrated stable thrombus. She was discharged home with anticoagulation and cardiology follow-up for consideration of catheter removal.

Right atrial masses are rare, and the differential diagnosis includes thrombi, vegetation, or tumor, for example

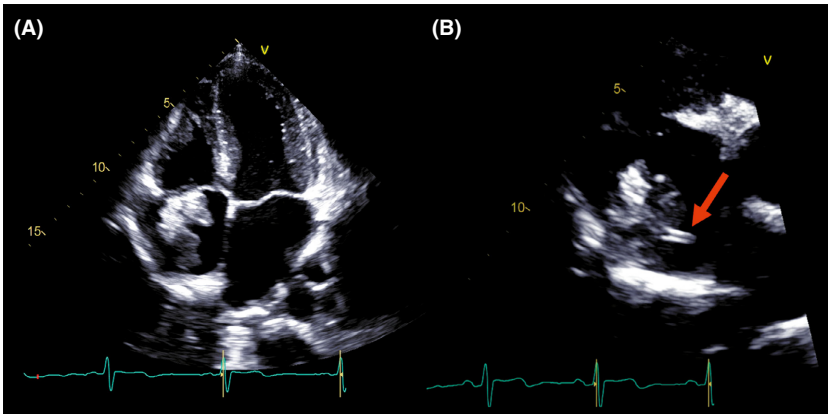

Figure 1. Transthoracic echocardiography. (A) Apical four-chamber view demonstrates a mass in the right atrium (yellow arrow) (B) Parasternal short-axis view with focus on the right atrium demonstrates a catheter tip is visible touching the mass (red arrow).

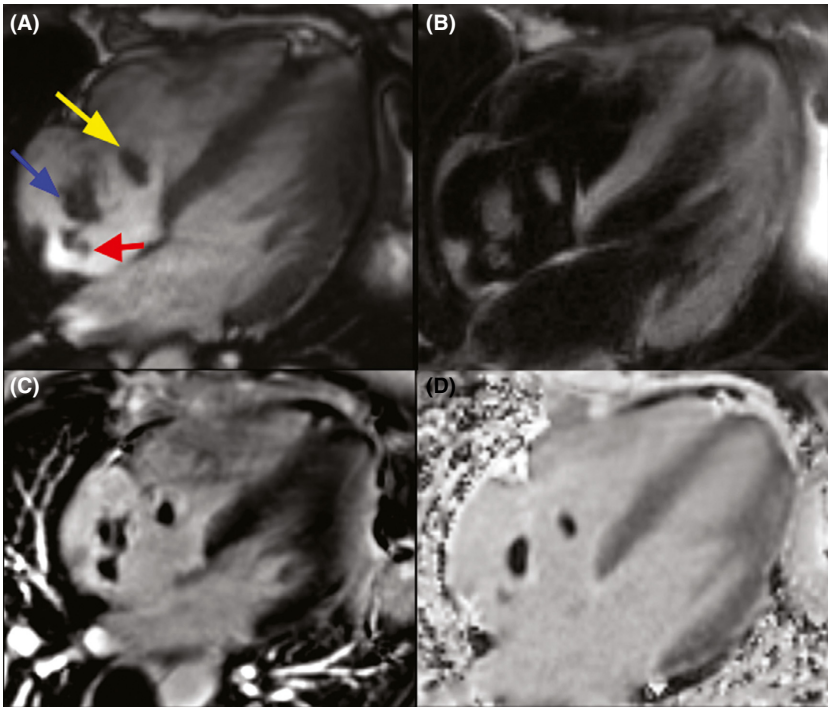

Figure 2. MRI characterization of right atrial thrombi. (A) A four-chamber Steady-State Free Precession (SSFP) cine shows three separate right atrial thrombi. There is thrombus associated with a catheter tip (red arrow). The largest thrombus is noted in the mid-portion of the right atrium (blue arrow). Thrombus is also noted close to tricuspid valve (yellow arrow). (B) A four-chamber fat-suppressed double inversion recovery image excludes lipoma as an explanation for the mass. (C) A 4-chamber perfusion image more clearly demonstrates low uptake of contrast compared to the blood pool. (D) Phase-sensitive inversion recovery imaging shows minimal late gadolinium enhancement when compared to normal myocardium. These cardiac MRI findings are consistent with intracardiac thrombus and are not typical of myxoma or vegetation.

myxoma [1]. Cardiac MRI has a high sensitivity and specificity for detection of intracardiac mass [2] and can differentiate thrombus, lipoma, inflammatory masses, and many solid tumors. Specific MRI methods can help characterize each of these masses but the setting of a nonlipomatous mass that shows very little contrast enhancement early and late after contrast is most consistent with

Catheter-related right atrial thrombus (CRAT). CRAT has been described in dialysis patients and can be associated with fatal complications, including arrhythmias and mechanical cardiac complications [3]. Sickle cell disease increases the risk of developing venous thromboembolism (VTE) [4], and right atrial thrombus has been reported in sickle cell disease patients [5]. However, the incidence of

CRAT is still unknown in patients with sickle cell disease and should be studied further. This patient had additional risk factors for VTE, including two in-dwelling catheters, a positive lupus anticoagulant, and ulcerative colitis [6]. Antiphospholipid testing, including anticardiolipin antibody, anti-beta2-glycoprotein, and lupus anticoagulant, should be repeated at least 12 weeks apart in order to diagnose antiphospholipid syndrome [7]. CRAT treatment is determined by thrombus size and a recent meta-analysis recommends that thrombi <6 cm can be safely managed by anticoagulation [8]. Patients should receive at least 3 months of anticoagulation regardless of whether the catheter is removed. If a catheter is not removed, then anticoagulation should continue as long as the catheter remains in place [9].

Authorship

CZ: identified clinical case, collected clinical data, performed literature, reviewed, drafted, and edited manuscript. BS: reviewed cardiac MRI and provided the figures. RNR: participated in patient care and reviewed manuscript. AEA: reviewed the MRI and provided the figures. AB: reviewed echocardiogram and provided the figures. SLT: participated in patient care, and reviewed and revised manuscript. APR: identified clinical case, revised manuscript, and approved final version to be published.

Conflict of Interest

None declared.

References

1. Cianciulli, T. F., M. C. Saccheri, H. J. Redruello, L. A. Cosarinsky, L. Celano, C. S. Trila, et al. 2008. Right atrial thrombus mimicking myxoma with pulmonary embolism in a patient with systemic lupus erythematosus and secondary antiphospholipid syndrome. Tex. Heart Inst. J. 35:454–457.
2. Freedberg, R. S., I. Kronzon, W. M. Rumancik, and D. Liebeskind. 1988. The contribution of magnetic resonance imaging to the evaluation of intracardiac tumors diagnosed by echocardiography. Circulation 77:96–103.
3. Salani, T. G., C. M. Borges, C. S. Urbini, P. Schincariol, K. R. Quadros, M. A. Ribeiro-Alves, et al. 2016. Patient in chronic hemodialysis with right atrial mass: thrombus, fungal endocarditis or atrial myxoma? J. Bras. Nefrol. 38:462–465.
4. Naik, R. P., M. B. Streiff, C. Haywood Jr., J. A. Nelson, and S. Lanzkron. 2013. Venous thromboembolism in adults with sickle cell disease: a serious and under-recognized complication. Am. J. Med. 126:443–449.
5. Yeghen, T., S. Benjamin, O. Boyd, C. Pumphrey, and D. H. Bevan. 1995. Sickle cell anemia, right atrial thrombosis, and the antiphospholipid antibody. Am. J. Hematol. 50:46–48.
6. Hudson, M., A. Chitolie, R. A. Hutton, M. S. Smith, R. E. Pounder, and A. J. Wakefield. 1996. Thrombotic vascular risk factors in inflammatory bowel disease. Gut 38:733–737.
7. Miyakis, S., M. D. Lockshin, T. Atsumi, D. W. Branch, R. L. Brey, R. Cervera, et al. 2006. International consensus statement on an update of the classification criteria for definite antiphospholipid syndrome (APS). J. Thromb. Haemost. 4:295–306.
8. Stavroulopoulos, A., V. Aresti, and C. Zounis. 2012. Right atrial thrombi complicating haemodialysis catheters. A meta-analysis of reported cases and a proposal of a management algorithm. Nephrol. Dial. Transplant. 27:2936–2944.
9. Rajasekhar, A., and M. B. Streiff. 2017. How I treat central venous access device-related upper extremity deep vein thrombosis. Blood 129:2727–2736.