

IMAGES

in PAEDIATRIC CARDIOLOGY

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Key words: thrombus, right atrium, t-cell lymphoma, imaging, lysis therapy.

Abstract

The case presents a wall adherent structure in the right atrium in a young patient with peripheral t-cell lymphoma followed by successful prolonged lysis therapy resulting in the resolution of the thrombus is presented. This case highlights the utility of multimodality imaging in an accurate assessment of the right atrium thrombus and the effectiveness of prolonged lysis therapy.

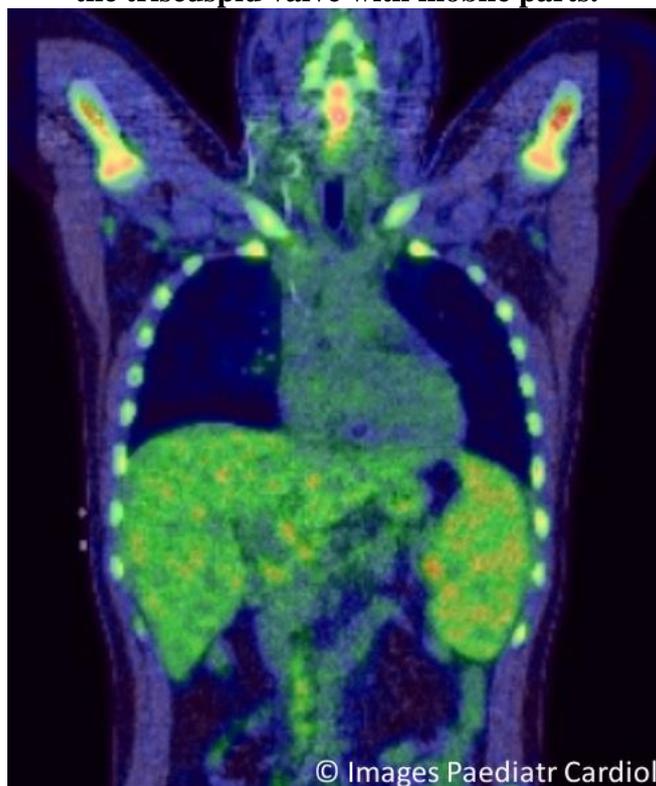
Introduction

The use of central venous catheters by patients with hypercoagulable state due to underlying malignancy could be associated with complications.¹ Especially the right atrial septic thrombi are a rare but feared complication and often associated with fatal clinical outcomes like migration toward the pulmonary artery.² The most promising treatment for wall adherent as well as free-floating massive right heart thrombi remains unclear and is open to debate.³ However the exact assessment of the structure in the right atrium is necessary for more accurate therapeutic decision.

Case Report

The case reports of an 18 year old man with the medical history of an Epstein Barr Virus infection in September 2012. In March 2013, he developed fever, night sweats with weight loss (- 15% of body weight) and coughing. Pneumonia was diagnosed in chest x-ray and confirmed in thorax CT scan with bronchoscopic detection of mycoplasmas in bronchoscopic lavage. Moreover swollen mediastinal and hilar lymph nodes were noticed; thus a lymph node biopsy was performed. The pathology unmasked the diagnosis of peripheral t-cell lymphoma (stage III B, WHO classification) without bone marrow infiltrates. Chemotherapy was started in May 2013 according to the guidelines of Studienzentrale Non-Hodgkin-Lymphome Berlin-Frankfurt-Münster (*NHL-BFM registry*) but had to be terminated because of neutropenia (1100/ μ l [$>1500/\mu$ l]) and elevated CRP (80 mg/dl [<5 mg/dl]) with fever of unknown origin (38.3°C). The resting ECG revealed a sinus tachycardia (115 bpm) with no signs of ischemia. On routine transthoracic echocardiography (TTE) (Fig. 1) a structure in the right atrium was suspected as a possible focus of infection, especially because a Hickman catheter (C. R. Bard, UT, USA) was placed in the right atrium.

Figure 1. TTE showed (9th August) a 4x3 cm wall adherent structure in the right atrium until the tricuspid valve with mobile parts.



To discriminate between thrombus and a malignancy a PET-CT (Fig. 2) was performed which detected hypermetabolic lymph nodes at the right hilus but no accumulation in the heart; thus an intracardial involvement of the lymphoma could be excluded.

Figure 2. PET-CT (5th August) revealed no pathologic nuclide distribution in the heart.



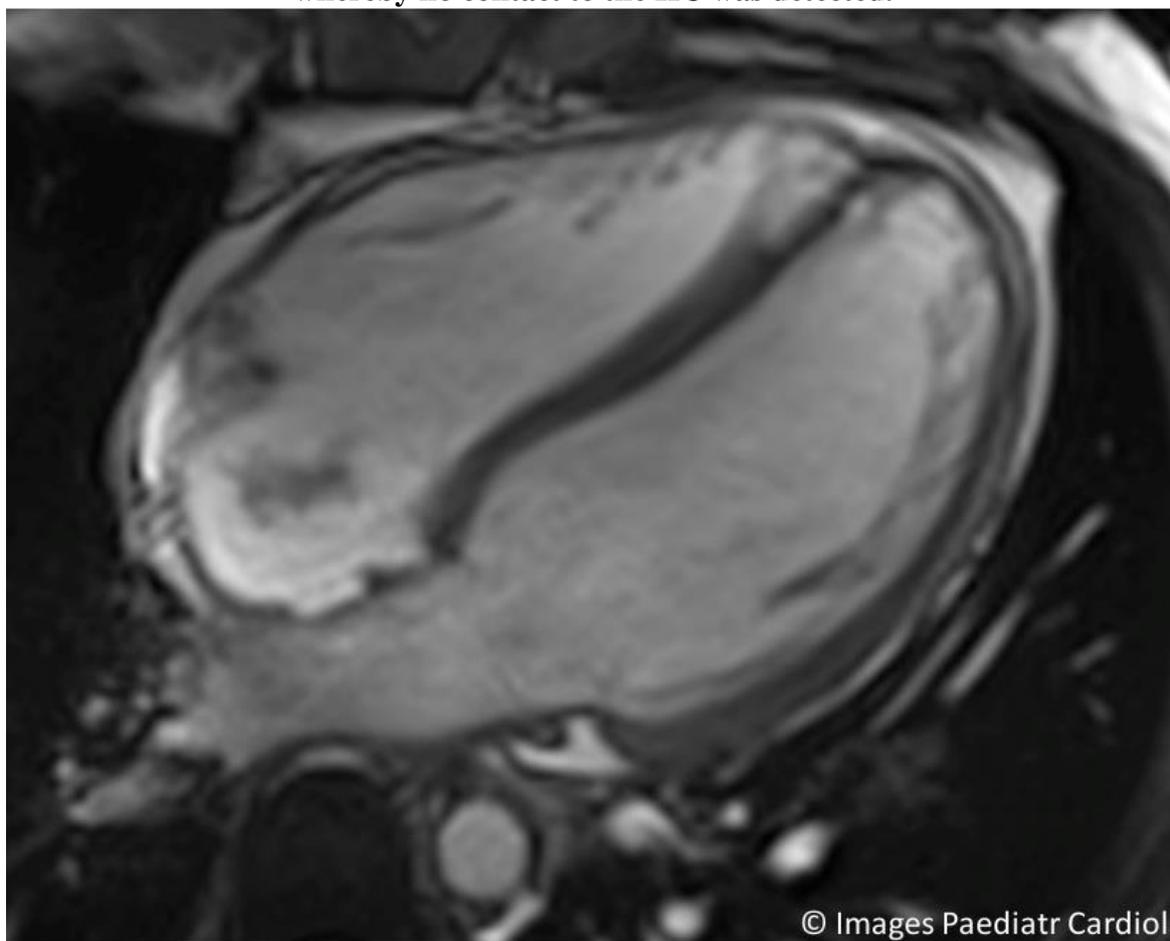
The hyperechoic structure in the right atrium was measured with 4x3 cm and floated in the right ventricle suspected with contact to the tricuspid valve whereas the left ventricular function was normal (Fig. 1). Additionally the distal end of the Hickman catheter (HC) was rough and has a thickened ending; thus the underlying mechanism could be an endocarditis. With the suspicion of an additional endocarditis, an empiric intravenous antibiotic therapy was started with vancomycin (2x1g). The blood cultures from peripheral blood and the HC remained negative; thus the antibiotic therapy was stopped three days later. The transesophageal echocardiography (TEE) (Fig. 3) revealed that the floating structure at the lateral wall of the right atrium has neither contact to the tricuspid valve nor to the HC.

Figure 3. TEE (16th August) A floating inhomogen structure a the lateral wall of the right atrium without contact to the tricuspid valve. Catheter was localist 9 mm in the right atrium with thickened ending.



These findings were confirmed by high-resolution Magnetic Resonance Scan (MRI) (Fig. 4). The entirety of these findings leads to the diagnosis of a thrombus and makes an endocarditis unlikely. The case was discussed with the cardiac surgeons to evaluate the possibility of an open heart operation with thrombectomy. Due to the comorbidities and the planned chemotherapy it was decided to try a conservative strategy with anticoagulation in this hemodynamically stable patient. The patient received a bolus unfractionated heparin 5000 international unit (IU) and afterwards two days continuously intravenous unfractionated heparin (750-1100 IU/h, target PTT 40-60 sec.). Despite of adequate concentrations, no change of size of the thrombus was seen in the follow-up TEE/MRI (Fig. 3-5). Following the literature for patients with prosthetic valve thrombosis it was performed a prolonged thrombolytic treatment with alteplase with 20 mg bolus followed by 80 mg/24h and unfractionated heparin 7.000 IU/24h during 6 days. The result was an effective thrombolysis with complete remission of the structure and a safe procedure without following neither pulmonary embolism detected by computed tomography or pulmonary arterial hypertension (Fig. 6).

**Figure 4. Cardiac-MRI (16th Aug). No Change the size under therapeutic anticoagulation
Due to the missing blood circulation a thrombus was assumed as the most probable diagnosis
whereby no contact to the HC was detected.**



**Figure 5. Bicaval view of TEE (16th August) revealed that the thrombus is opposite to the
superior cava vein and thus the HC near the Eustachian valve suggesting that the thrombus
might be due to trauma during placement of the HC.**



Figure 6. The follow-up TEE (19th Aug) under lysis therapy with remission with filamentary structures and wall-adherent remains.



Discussion

The case describes the utility of multimodality imaging approach for defining an intracardiac wall adherent structure for decision making and follow-up evaluation in an 18 year old patient with peripheral t-cell lymphoma. The incidence of venous thromboembolism in patients with malignancies is elevated and aggravated by the use of venous access catheters.⁴ Atrial masses were often first observed by routine TTE as a general screening test of cardiac structure and function for which thrombus is rarely the primary indication.⁵ However TEE added additional findings and is superior in elucidating the nature, significance and anatomical relationship of the masses.⁶ Moreover a MRI scan was performed of the heart and repeated in four days later for a close follow-up. The study of Srichai et al.⁶ suggested that contrast-enhanced MRI is superior at identifying intracardiac thrombi than echocardiography especially in patients with poor echocardiographic conditions such as intubated patients, obese patients as well as chronic obstructive lung disease. Contrast-enhanced MRI showed a high sensitivity and specificity of 88% and 99%, respectively, and was superior to both TTE and TEE for mural thrombus. Despite the fact that primary cardiac tumors,⁷ cardiac metastases^{8,9} or lymphoma¹⁰ are rare, a Positron emission tomography-computed tomography (PET-CT) was performed to rule them out. Therapy without any delay is necessary but the management strategy of right heart thrombi remains controversial. It was decided to keep the HC inside due to the elevated risk of thromboembolism resulting in severe right heart failure by

removing thrombi. Data from literature are controversial, there are reports on a thrombolytic therapy and a complete dissolution of a right atrial thrombus.^{11,12} However reported mortality was not influenced by the choice of treatment.¹² Another group concluded that in patients with severe pulmonary embolism and free-floating thrombi in the right heart emergency surgery is usually advocated.¹³ Nevertheless, thrombolysis is faster and seems promising either as the only treatment or as a bridge to surgery in emergent settings. Kula et al.¹ reported the case of a patient in the age of puberty with acute lymphoblastic leukemia and an atrial thrombosis in her right heart, who was treated with tissue plasminogen activator (t-PA) initially and underwent surgery because of an unsatisfactory response. Yoo et al. referred of the echocardiographic disappearance of a free-floating right heart thrombus after thrombolysis. However, the thrombus migrated and resulted in pulmonary thromboembolism and had to be removed by surgery.³ In patients with contraindications to surgery or thrombolysis therapy, percutaneous interventional techniques may be proposed.¹⁴ Low-dose, slow infusion of tissue-type plasminogen activator (t-PA) with repeated doses as needed provides an effective and safe therapy for the treatment of prosthetic valve thrombosis. It has a lower complication and mortality rates with no loss of effectiveness compared to higher doses or rapid infusions of streptokinase or t-PA.¹⁵ Nevertheless, before defining a treatment strategy, there is a need for defining the underlying origin. In our case, the concomitant use of different diagnostic tools leads to the right diagnosis and thus to a successful outcome.

Conclusion

The case emphasizes the need for high suspicion for thrombus when a right atrial mass is found in a patient with a hypercoagulable state due to underlying malignancy with a central venous catheter. As conclusion different types of imaging are required to confirm the initial hypothesis, to determine the exact location and anatomical relation of the thrombus and thus to guide treatment strategy.

Acknowledgments

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