

Primary Cardiac Angiosarcoma

Intracardiac Echocardiography to Guide Biopsy of an Intracardiac Right-Sided Mass

Teodor Serban, Diego Mannhart, Otmar Pfister, Michael Kühne, Christian Sticherling, Patrick Badertscher

Department of Cardiology, University Hospital Basel, Switzerland; Cardiovascular Research Institute Basel, University Hospital Basel, Switzerland

Summary

Introduction: The diagnosis and characterisation of intracardiac tumours is challenging. Transoesophageal echocardiographic guided biopsy is an established method to confirm the pathology. Intracardiac echography (ICE) may help to increase the diagnostic outcome of biopsies, particularly in right-sided cardiac masses.

Case Report: We report on a 62-year-old patient who presented with recurrent pericardial effusions of unknown origin. Pericardial puncture did not reveal any significant findings. Positron emission tomography-computed tomography revealed a mass in the right atrium. Due to excellent nearfield imaging capacities for right-sided cardiac structures, ICE was used to guide the biopsy. Biopsy was performed safely and demonstrated a primary cardiac angiosarcoma.

Conclusion: ICE-guided biopsy can be a reasonable approach, particularly for right-sided structures, allowing a safe, minimally invasive way of diagnosing intracardiac tumours.

Keywords: Primary cardiac angiosarcoma; right atrial tumors; pericardial effusion; pericardial tamponade; intracardiac echography

Introduction

Primary cardiac tumours are rare and the majority are benign [1]. Primary cardiac angiosarcoma is the most common malignant cardiac tumour [2]. Affected patients are usually between 40 and 50 years old and are often symptom-free at the time of presentation, leading to diagnosis at a late stage. Various imaging modalities play a central role in diagnosis, such as transthoracic echocardiography (TTE), cardiac computed tomography (CT) or cardiac magnetic resonance imaging (cardiac MRI) [2]. The biopsy is classically an open approach procedure at the same time as the tumour resection, but lately other minimally invasive methods, such as biopsy using transoesophageal echocardiography (TOE) or intracardiac echography (ICE) for guidance have been described [3].

Case Report

We present the case of a 62-years-old patient presenting to the emergency department with exertional dyspnoea. The patient was found to have bilateral metastases in the suprarenal glands and multiple grouped metastases in the lungs, though the primary tumour remained unknown. He had been taking rivaroxaban for 8 years for recurrent deep vein thrombosis. On admission, the patient was tachycardic with a heart rate of 105 bpm and a blood pressure of 68/47 mm Hg. The electrocardiogram demonstrated PR segment depression in inferior leads and V3–V6. In lead aVR, PR segment elevation with ST segment depression was noted (fig. 1). Bedside echography was performed, which showed a large pericardial effusion with compression of the right atrium and right ventricle with swinging heart sign. The diagnosis

of pericardial tamponade was made and pericardiocentesis was performed. 550 ml of haemorrhagic pericardial fluid was drained. The analysis of the pericardial fluid showed no malignant cells. The patient underwent CT scan the following day, which revealed pulmonary embolism and a cardiac mass in the right atrium. For further examination, a subsequent positron emission tomography-computed tomography was performed, where a contrast-capturing infiltrative tumour of the right atrium was observed. On TTE, a transmural tumour of the free wall of the right atrium could be visualized. The tumour was adherent to the pericardium (fig. 2). Due to the location of the tumour in the right atrium, ICE was used to guide the biopsy. Under local anaesthesia, a steerable sheath Agilis NxT (Abbott, Chicago, Illinois, 8.5-Fr) and an ICE catheter (AcuNav, Siemens, Munich, Germany) probe were inserted via the femoral vein. Both the ICE probe and the steerable sheath were advanced into the right atrium. The Agilis sheath was used to provide precise control of the cardiac biotome. A 7.5-Fr 2.4 mm Jawz biotome (Argon Medical, Frisco, Texas) was inserted into the right atrium through the Agilis sheath and was directed toward the tumour (fig. 2). Multiple biopsy probes from different locations were taken. The histological examination showed a highly malignant angiosarcoma.

Discussion

With increasing complexity of interventional cardiac procedures, ICE has become a useful imaging modality with a broad scope of appli-

Case Report

cations. In this case report we demonstrate the use of ICE in the diagnosis of heart tumours by facilitating and guiding biopsy using a minimally invasive procedure. ICE was chosen to guide biopsy due to the location of the cardiac mass affecting the right atrial wall. In this location, precise biopsy is of utmost importance. By using ICE, biopsy was performed safely

with enhanced spatial detection leading to precise positioning of the biopsy forceps. ICE provides excellent nearfield imaging that is often superior to transoesophageal imaging for right-sided cardiac structures [4]. Better direct visualisation allows multiple biopsies to be taken from multiple targets, theoretically further increasing the diagnostic yield. Our case

extends and verifies findings from previous reports describing the use of ICE for the biopsy of tumours in several regions of the heart including the right atrium, the right ventricular apex, the right ventricle outflow tract and the left ventricle [3–5]. Due to the good quality of the TTE image in our case, guiding the biopsy procedure with TTE would have been possible.

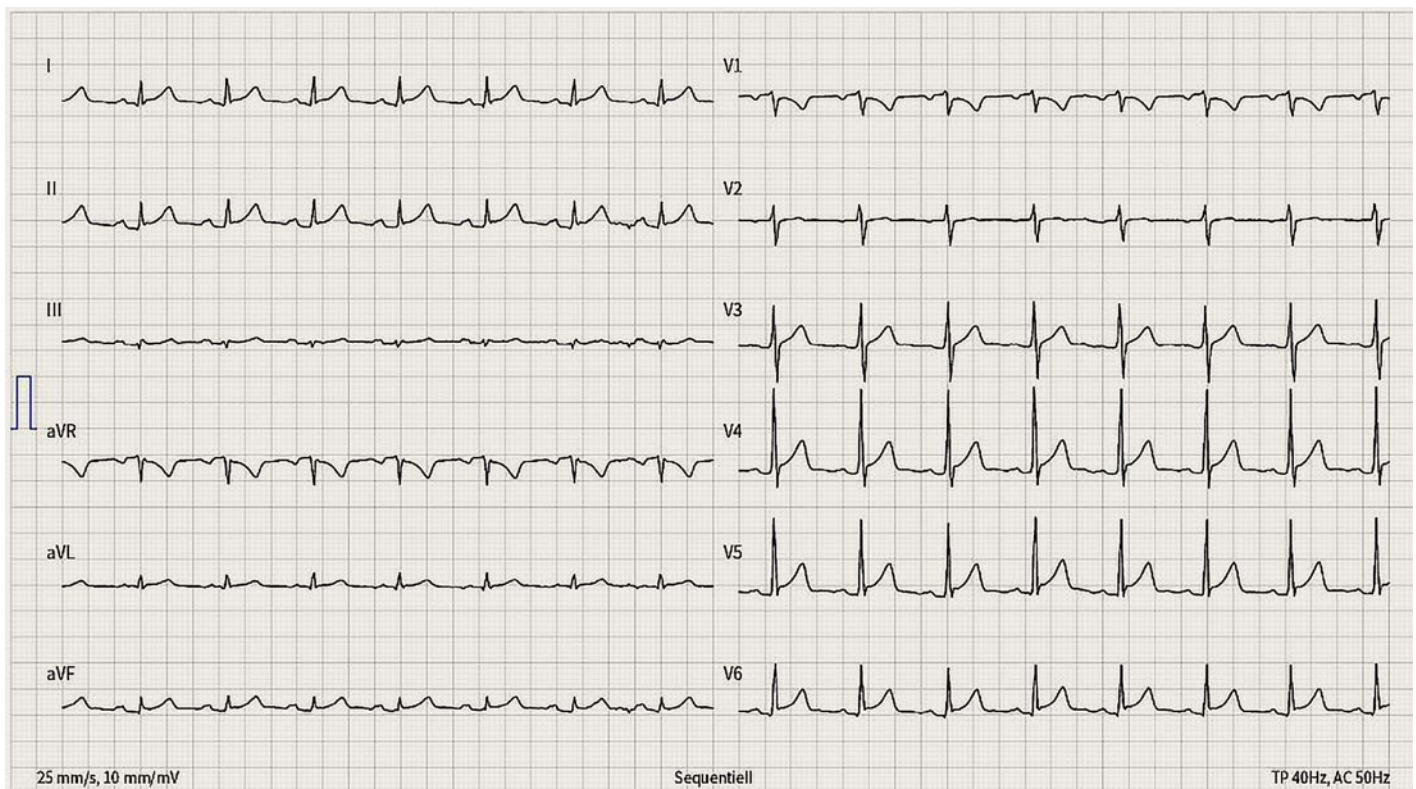


Figure 1: Electrocardiogram showing sinus rhythm with a heart rate of 90 BPM. Diffuse concave ST elevation without mirror ST segment depression were noted. Furthermore, a PR segment depression was seen in V4 and V5. Lead aVR shows a PR segment elevation with ST segment depression – also known as Knuckle Sign. These findings are consistent with acute pericarditis. Typical findings of cardiac tamponade include QRS alternans and sinus tachycardia.

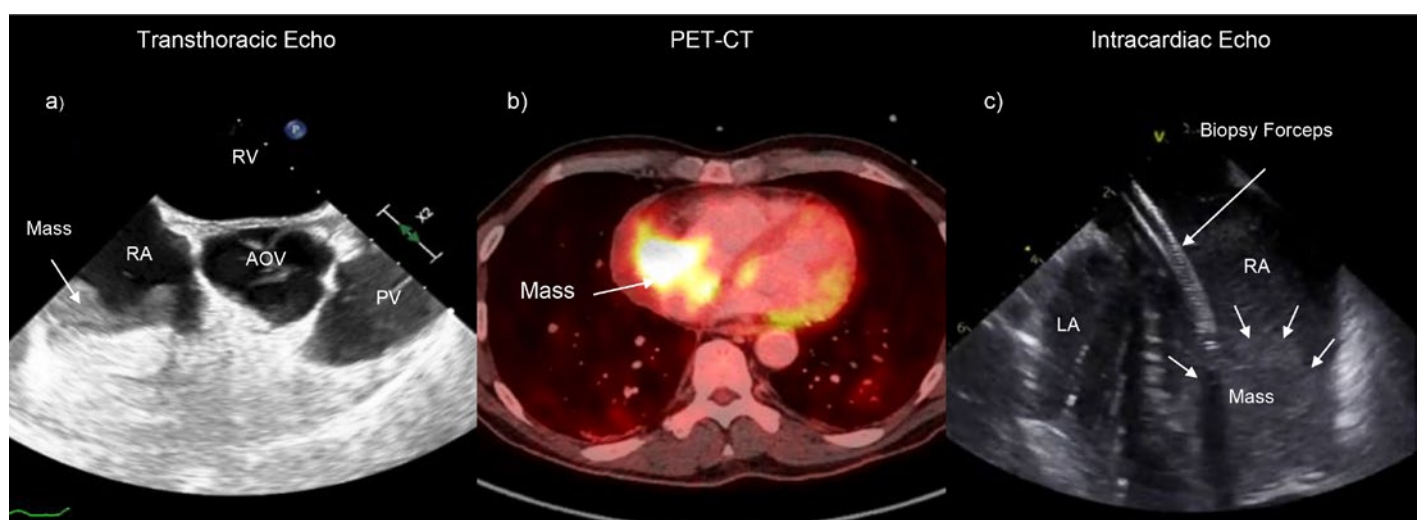


Figure 2: Panel encompassing several imaging modalities of the cardiac tumour. **a)** Transthoracic echocardiography (parasternal short axis view) demonstrating an unclear hyperechogenic mass in the posterior right atrial wall. **b)** Positron emission tomography–computed tomography (PET-CT) view – a contrast-capturing structure in the posterior free wall of the right atria can be observed. **c)** Intracardiac echography showing the biopsy forceps and the right atrial tumour. AOV = Aortic Valve, LA = left atrium, PV = Pulmonary valve, RA = right atrium, RV = right ventricle.

However, by using ICE for guidance, radiation exposure of the interventionalist and especially the cardiac sonographer can be minimised. An additional advantage of ICE is that the catheter can be introduced through the femoral vein, a standard vascular access site, and compared to TTE or TOE does not require additional set-up or staff members [5]. The main limiting factors are the restricted availability, operator's experience and the possible increased costs. In conclusion, multiple imaging modalities can be used as guidance for biopsy of cardiac masses. ICE should be considered in selected cases to guide biopsy given its ease of use and high definition visualisation of right-sided cardiac structures.

Correspondence

Dr Patrick Badertscher
University Hospital Basel
Petersgraben 4
CH-4031 Basel
[patrick.badertscher\[at\]usb.ch](mailto:patrick.badertscher[at]usb.ch)

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Dre Tomoe Stampfli, Chairwoman of the Jury
Fondation Cœur de la Tour
Av. Maillard 1 - 1217 Meyrin
info@coeurdelatour.ch