

Left ventricular endomyocardial biopsy guided by intracardiac echocardiography via a trans-septal approach

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Abstract

We describe our use of intracardiac echocardiography (ICE) to guide endomyocardial biopsy of the left ventricle in a patient with chronic heart failure secondary to endocardial fibroelastosis. Under ICE guidance, we advanced the biptome through the middle of the mitral annulus, within a trans-septal sheath, and guided the biptome to the correct location on the mid-posterior wall of the left ventricle. This was performed without adverse events, such as injury to the chordae tendineae or mitral valve regurgitation. Therefore, ICE-guidance is possible for left-sided heart procedures and could lower the risk of complications and improve the biopsy rate for diagnosis.

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Key words : atrial septal approach, endocardial fibroelastosis, intracardiac echocardiography, left ventricular endomyocardial biopsy

Endocardial fibroelastosis (EFE) is a unique form of *de novo* fibrosis that forms in the subendocardial layer encapsulating the myocardium and stunting its growth. EFE is typically associated with congenital heart diseases of heterogeneous origin, such as hypoplastic left heart syndrome.¹ Herein, we describe the presentation and diagnostic process for EFE in a 29-year-old woman referred to our hospital for catheter ablation (CA) due to symptomatic atrial fibrillation (AF), after 2 failed CA attempts at another hospital. Her relevant past medical history was as follows. She had presented with left ventricular dysfunction at the age of 1 year, with a diagnosis of EFE made at that time. She had undergone endomyocardial biopsy (EMB) of the right ventricle at the age of 18 years, with no histopathological evidence of EFE. Therefore, a biopsy of the left ventricular myocardium was now requested, to be performed at the time of CA, for further examination.

The echocardiographic findings at the time of the present admission revealed a left ventricular spherical dilation and wall motion abnormality (**Figure 1A**). Cardiac magnetic resonance imaging revealed a thinning

of the inferoseptal aspect of the left ventricle, with an increased extracellular volume fraction (**Figure 1B**). Heart palpitation during AF negatively impacted her quality of life. Of note, her heart failure status was associated with left ventricular dysfunction, although without evidence of pathological EFE findings. After careful consideration, we decided to perform left ventricular EMB for histopathological diagnosis and CA for AF, sequentially, under general anesthesia.

After trans-septal puncture, via the femoral vein, an 8-Fr ultrasound catheter (SOUNDSTAR, Biosense Webster, Diamond Bar, CA, USA) and a 8.5-Fr deflectable sheath (Agilis Sheath, St. Jude Medical, St. Paul, MN, USA) were placed in the left atrium. After bending the deflectable sheath to the ideal position through the middle of the mitral valve annulus, the biptome (Technowood Biopsy Forceps, Tonokura Ika Kogyo Co Ltd, Tokyo, Japan) was inserted into the sheath. Under intracardiac echocardiography (ICE) guidance, the biptome was inserted through the left atrium and provided real-time images of the left ventricle. Subsequently, it was advanced to the high echoic lesion in the thinned region of the inferoseptal wall of the left ventricle. However, specimens could not be obtained from the site of the lesion, even with repeated attempts, due to slippage of the biptome (**Figure 2A**). The biptome was then advanced to the mid-posterior wall of the left ventricle where EMB was successfully performed, with 2 specimens obtained for diagnostic purposes (**Figure 2B**). The entire procedure was performed under ICE guidance, with direct visualization of the anatomy of the left ventricle, with no complications, including mitral valve regurgitation. After EMB, CA was performed without any complication. The patient had no symptoms postoperatively and was discharged after a normal sinus rhythm was maintained for 7 days after the procedure.

Pathological examination of the specimens revealed an endocardial fibrous thickening and interstitial fibrosis of the left ventricle (**Figure 3**). These findings were not observed in the previous specimens from the right ventricle and were indicative of possible secondary EFE.

The overall complication rate for EMB is low, estimated at 1-2% for experienced surgeons.² When complications do occur, cardiac perforation and valve regurgitation due to tearing of the chordae tendineae are lethal. It is noteworthy that sampling errors do occur frequently in cases with a localized lesion, such as cardiac sarcoidosis.³ ICE-guidance for EMB can be helpful in this regard, with previous studies having described its benefit for EMB from the right atrium and ventricle.^{4,5} However, to our knowledge, this is the first report of ICE-guided EMB from the left ventricle, via a trans-septal approach, under ICE-guided navigation, with the ICE probe inserted into the left atrium. This technique could permit left ventricular biopsy when the transaortic approach is difficult, such as after aortic valve replacement. We note that although ICE guidance from the right ventricle may provide left ventricle information, more detailed information of the left-sided heart could be obtained, including clearer imaging, using ICE inserted into the left atrium. The technique also enables EMB of the left ventricle to be performed without increasing the risk of valve regurgitation as it enables visualization of the surrounding heart structures and, thus, avoiding injury to the chordae tendineae. ICE-guidance also allowed us to navigate the biptome to the ideal location and to predict success in obtaining the biopsy specimen; this could improve the rate of success of diagnostic biopsy for cardiomyopathies.

Disclosures : The authors have no conflicts of interest to declare.

Ethics : The patient provided consent for the publication of this case report.

Figure legends

Figure 1

1. Transthoracic echocardiography showing spherical dilation and wall motion abnormality (long axis view). LV = left ventricle; LA = left atrium; Ao = Aorta
2. Cardiac magnetic resonance imaging showing thinning of the inferoseptal aspect of the left ventricle (small arrows) and an increased extracellular volume fraction (red lines).

Figure 2

Intracardiac echocardiography (ICE) images obtained during the procedure.

1. The tip of the biptome is shown next to the high echoic area of the thinned inferoseptal aspect of the left ventricle where repeated biopsy attempts failed (small arrowhead).
2. The chordae tendineae were visualized and the biptome manipulated to avoid injury, with subsequent advancement to the mid-posterior wall where endomyocardial biopsy (EMB) was performed.

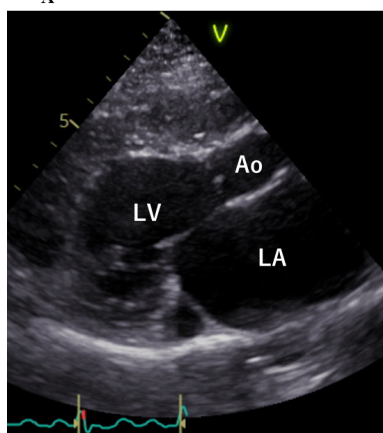
Figure 3

Pathological examination confirming endocardial fibrous thickening and interstitial fibrosis of the left ventricle (Masson's trichrome stain). Scale bar, 100 μ m.

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Figure 1 A



B

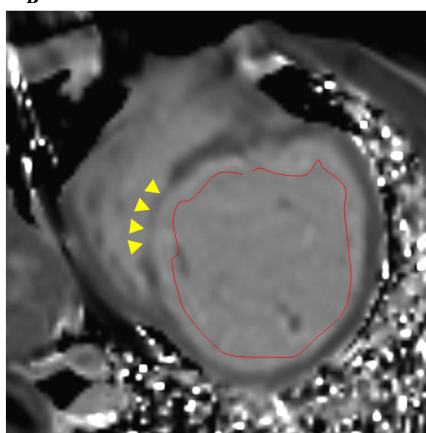


Figure 2

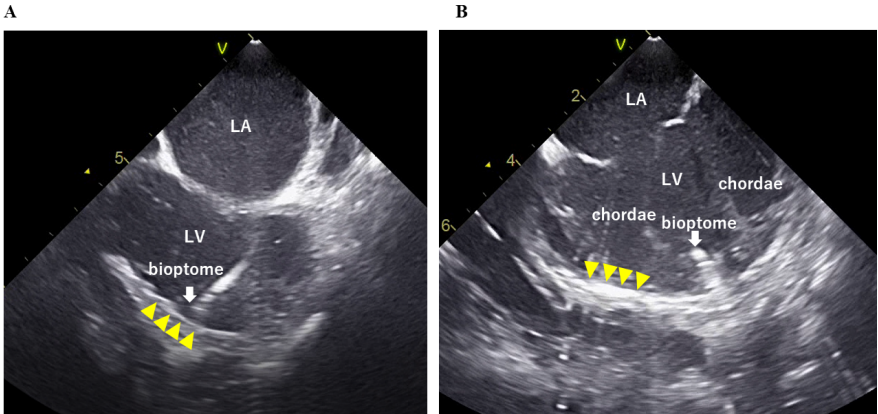


Figure 3

