Multimodality imaging in Loeffler endocarditis as a rare cause of stroke

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INTRODUCTION

Loeffler endocarditis complicating hypereosinophilic syndrome is a rare restrictive cardiomyopathy caused by abnormal infiltration of eosinophils into the endomyocardium, with subsequent tissue damage from degranulation, eventually leading to fibrosis.

The patient may present with an intracardiac thrombus, arrhythmias, and/or acute heart failure which can be life-threatening, if not treated early.

The gold standard diagnostic modality for eosinophilic endocarditis was endomyocardial biopsy. Currently, clinicians prefer Cardiac imaging including echocardiography and cardiac MRI over endomyocardial biopsy as it is non-invasive and a more practical way to diagnose Loeffler endocarditis.

This case illustrates the importance of multimodality imaging in the diagnosis of hypereosinophilia.

CASE REPORT

A 63-year-old woman with no previous medical history, who presented with language disorder, praxis and rapidly progressive onset ataxia in the context of multiple strokes of cardio-embolic origin. Transthoracic echocardiography (TTE) and Transesophageal echocardiography (TEE) found LV wall thickening of the apex without thrombus (figure1), P2 perforation with thickening appearance of P2-P3(figure 3, 4), moderate to severe mitral regurgitation (MR)(figure 2). Cardiac MRI (CMR) showed subendocardial fibrosis of the LV(figure 5) with involvement of the posterior papillary muscle, severe MR, LV wall thickening of the apex(figure 6), no thrombus, no PFO. Complete microbiological check-up remained negative. The diagnostic of Loeffler endocarditis was then suspected. The only potential cause of Loeffler endocarditis, after eliminating secondary causes, and from the suggestive aspect of cardiac MRI as well us eosinophilia in biology test, was an hypereosinophilic syndrome. Our patient had a favorable clinical outcome and follow-up echocardiography showed stability of lesions.

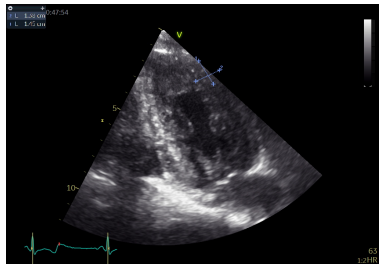


Figure 1: TTE showing LV wall thickening of the apex

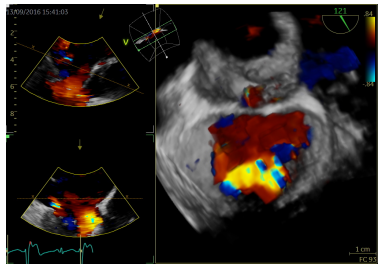


Figure 2: 3D TEE showing MR

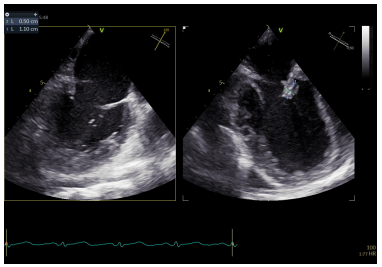
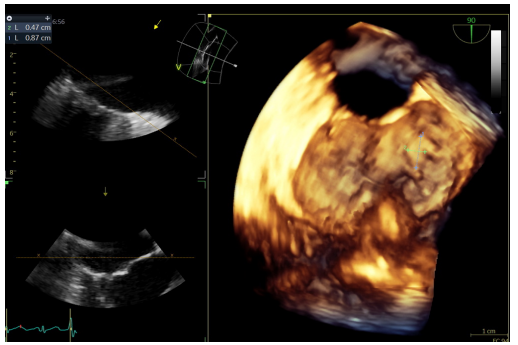


Figure 3: TEE simultaneous biplane image showing thickening appearance of P2-P3



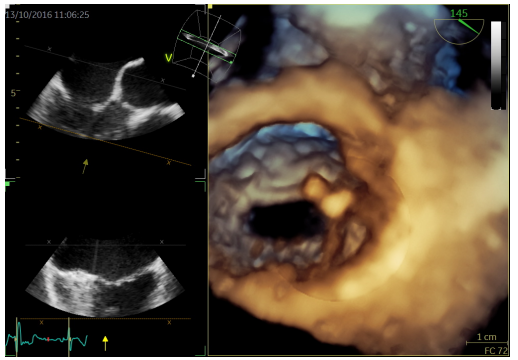
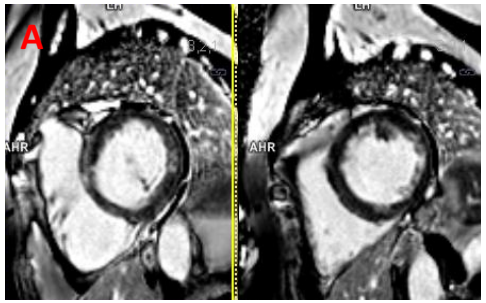


Figure 4: 3D TEE showing thickening appearance of P2-P3



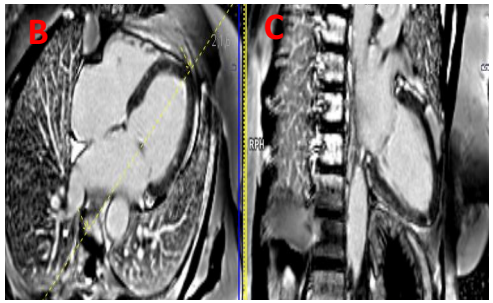


Figure 5: CMR imaging: Delayed gadolinium enhancement sequences demonstrating the left ventricular endomyocardial fibrosis (white arrow) in hypersignal. Midventricle short-axis slice(A), Horizontal long axis view(B) and vertical long axis view(C)

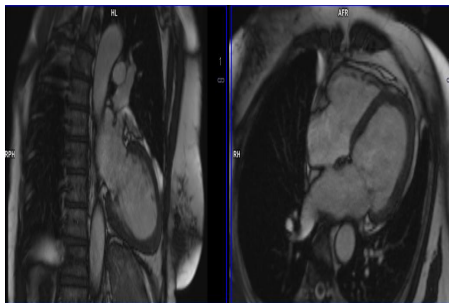


Figure 6: CMR imaging showing LV wall thickening of the apex without thrombus in cine sequences

DISCUSSION

- Hypereosinophilic cardiac disease is not rare.(1,2) Incidence and the prevalence is difficult to estimate (rarity of the disease, different degree of cardiac involvement, asymptomatic organ involvement).(3)

- It may have several presentations from acute myocarditis to endomycardial fibrosis and may involve the three cardiac layers. The most typical cardiomyopathy is represented by Loeffler's endomyocardical disease(3). TTE and cardiac MRI are clearly complementary.

- Loeffler endocarditis is characterized by hyper-eosinophilia and fibrous thickening of the endocardium, with generally large thrombi in the wall of the ventricles.

- Cardiac complications (especially intraventricular and subendocardial thrombus with risk of peripheral stroke and heart failure) are not uncommon and represent the first cause of mortality and morbidity(4).

- The diagnosis was based on cardiac biopsy, but since few years, multimodal cardiac imaging, including echocardiography and cardiac magnetic resonance imaging, is the gold standard of the diagnosis, especially for tissue characterization

→**In case of typical presentation in TTE and MRI, myocardial biopsy are not necessary in the diagnosis process of hypereosinophilic cardiac diseases**

- Echocardiography findings: mural ventricular thrombi at early stages, the acute necrotizing phase and endomyocardial fibrosis.
  - Thrombi may extend to the ventricular outflow tracts, to the subvalvular regions or even the inflow tract of atrioventricular valves inducing valvular regurgitation.
  - Obliteration of both right and left ventricles by thrombi corresponds to the typical description of the Loeffler endomyocardical disease.
  - Vegetations may be seen in advanced stages. They are made of a combination of organized thrombi, eosinophils and fibrosis and typically arise from the myocardial ventricular wall with a possible extension to the valvular leaflets. Embolic events or valvular dysfunction are their major complications(5)

- Contrast echo is obviously a very interesting technique in case of suspected apical thrombus and should be completed with cardiac MRI
- Strain analysis could provide useful information in case of a restrictive filling pattern to make the difference with infiltrative cardiomyopathy
- MRI is more specific and sensitive than TTE to differentiate thrombosis from fibrosis. It detects cardiac lesions not identified by other non-invasive modalities and may be considered as a surrogate to myocardial biopsies(6)

CONCLUSION

Loeffler endocarditis complicating hypereosinophilic syndrome is still a disease with significant morbidity and mortality. Usually diagnosed at an advanced stage, treatment is limited once fibrosis occurs, requiring heart failure medications or surgical intervention. Early and appropriate treatment is crucial to avoid the evolution toward irreversible cardiac lesions. Hence the importance of early diagnosis by multimodal cardiac imaging, including echocardiography and cardiac magnetic resonance imaging, especially for tissue characterization.

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