Fibrosis Endomiocárdica Asociada a Infarto de Miocardio

Autor
Nicolás Ariza Ordóñez
Antonia Pino Marín

Director
Gabriel Salazar Castro
Claudia Jaimes
Daniel Isaza

Cardiología adultos
Medicina interna

Escuela de Medicina y Ciencias de la Salud
Cardiología-Medicina interna
Universidad del Rosario

Bogotá - Colombia

2025

Abstract:

- **Objective**: Endomyocardial fibrosis (EMF) is the most common cause of restrictive cardiomyopathy in Africa and South America. We present a case of a 55-year-old male who experienced an anterior myocardial infarction due to thrombotic occlusion of the LAD artery. Multimodality imaging revealed LV apical obliteration, apical thrombus and subendocardial fibrosis, without significant systolic dysfunction, consistent with EMF.
- **Key Steps:** EMF should be suspected in patients presenting with heart failure symptoms due to restrictive physiology and/or thrombotic manifestations with typical echocardiographic findings. CMR imaging can provide valuable information regarding typical scar patterns.
- **Potential Pitfalls:** Similar features can be observed in other forms of cardiopathy, including:
    - Apical hypertrophic cardiomyopathy
    - LV thrombosis
    - Excessive LV trabeculation
    - Ebstein anomaly
- **Take-Home Messages:**
    - EMF is a significant cause of restrictive cardiomyopathy that primarily affects vulnerable populations in South America and Africa.
    - Multimodality imaging is essential for identifying typical features and facilitating differential diagnosis.

Introduction

EMF is a unique form of restrictive cardiomyopathy that usually presents in resource-limited settings. Several theories have been proposed to explain the origin, geographical distribution, and pathological findings of EMF [1,2]. Proposed mechanisms include parasitic infections endemic to tropical and vulnerable areas, eosinophilia, and potential dysregulated immune responses [2]. Additionally, environmental factors such as malnutrition, magnesium and protein deficiencies, and linamarin toxicity from cassava consumption have been suggested as contributing to its development [3]. These factors, combined with suspected genetic susceptibility, are believed to initiate an inflammatory process leading to endomyocardial fibrosis.

Diagnosis is based on multimodal imaging, with characteristic findings in echocardiography and cardiac magnetic resonance (CMR) related to apical filling with fibrotic tissue in one or both ventricles [3]. These findings typically translate to restrictive physiology, thrombotic complications, and valvular disease [4]. Despite limited treatment strategies, early diagnosis is crucial for identifying complications and managing them, as well as for developing follow-up strategies for advanced heart failure treatment and endocardectomy surgical planning.

We present a case of a 55-year-old male with acute myocardial infarction in which coronary embolism was suspected after angiographic findings and left ventricular apical obliteration with thrombosis documented via 2D echocardiogram. CMR revealed normal LV volume and systolic function, two LV scar patterns on LGE imaging, apical thrombosis and biatrial enlargement. The criteria proposed by Mocumbi et al. were applied fulfilling the diagnosis of moderate EMF [4].

Case summary

A 55-year-old man with no prior history, from a rural area in Colombia, presented with six hours of acute, crushing chest pain that began during brisk walking. Initial attention was provided at a primary care facility, where an ECG on admission revealed anterolateral Q waves and elevated conventional troponin-I levels. The following day, the patient was transferred to our center with a working diagnosis of acute myocardial infarction.

On admission, vital signs were unremarkable. Physical examination revealed a grade 3/6 systolic murmur at the left sternal border and grade I bilateral ankle edema. On systems review he reported intermittent episodes of shortness of breath with physical activity over the past few years, of variable duration, with no other symptoms. No prior imaging or laboratory data were available to contextualize his presentation.

Transthoracic echocardiogram revealed mildly reduced left ventricular ejection fraction (LVEF: 50%) and left ventricular apical obliteration, making regional wall motion assessment difficult. Biatrial moderate enlargement (left atrial volume 44 mL/m2) and mild mitral regurgitation were also noted. Diastolic assessment indicated grade II diastolic dysfunction (Fig. 1 and 2, video 1, video 2).

The patient also underwent coronary angiography, which showed partial recanalization of thrombosis in the left anterior descending (LAD) artery, with no atherosclerotic components in the LAD or the remaining coronary arteries (Fig. 2, video 3), fulfilling the criteria proposed by Shibata et al for coronary embolism [5].

A cardiac magnetic resonance (CMR) imaging study was performed to further characterize myocardial tissue and apical obliteration. CMR showed normal left ventricular systolic function (LVEF 60%) and end-diastolic volume (102 mL/m2), LV apical wall thickening with apical obliteration and biatrial enlargement. Early gadolinium enhancement (EGE) sequences showed an apical thrombus; and LGE sequences revealed two patterns of LV scar distribution: a transmural scar involving the apical segment of the anterior wall; and a subendocardial scar involving the other apical segments (Fig 4, video 4, 5 and 6).

The diagnosis of EMF was based on three major criteria: obliteration of the left ventricular apex, endomyocardial plaques >2 mm and ventricular thrombus without severe systolic dysfunction, and one minor criteria: enlarged atria with normal-sized ventricles. Complementary tests included a complete blood count, which showed no eosinophilia (30 cells/μL), and a stool microscopic examination that revealed no protozoa, helminths, or fecal leukocytes.

Management of the acute myocardial infarction was performed according to both international and national guidelines. Anticoagulation was maintained during hospitalization with low molecular weight heparin, and antiplatelet therapy was initiated with aspirin and clopidogrel. Other medications included extended-release metoprolol, losartan, and high intensity atorvastatin. A follow-up coronary angiography demonstrated a substantial reduction in LAD thrombotic load.

Acute parasitic infections and autoimmune disorders were ruled out. During hospitalization, a nutritional assessment was conducted to reduce cassava consumption, increase intake of vegetables, fruit, and protein; and improve food handling. The patient was discharged on a regimen of warfarin, clopidogrel, atorvastatin, losartan, and metoprolol.

The patient is being followed up with anticoagulation and cardiology outpatient clinics. Over the past 3 months, there have been no new episodes of chest pain or congestive symptoms.

Procedural steps

EMF diagnosis needs a high suspicion index that raises in patients from sub-Saharan Africa or tropical Latin-America presenting with heart failure symptoms related to restrictive physiology, valvular disease or thromboembolic complications. 2D Echocardiography is the first line evaluation modality because its wider availability and lower cost compared to other imaging modalities. Echocardiogram findings include:

- Obliteration of ventricular apices or valvular recesses (Fig 1, video 1 and video 2).
- Endocardial plaques or thickening (Fig 1, video 1 and video 2).
- Ventricular thrombosis or spontaneous contrast without severe ventricular dysfunction (Fig 1, video 1 and video 2).
- Atrioventricular valvular regurgitation due to leaflet restriction to movement.
- Atrial enlargement.
- Diastolic dysfunction (Fig 2).
- Right ventricular retraction and reduction of cavity volume.

Once the echocardiogram is obtained, the criteria proposed by Mocumbi et al. can be applied to establish a diagnosis [4]. When available, CMR provides valuable information on cardiac morphology and ventricular function. Additionally, tissue characterization techniques offer insights into the presence and pattern of myocardial inflammation, injury, and fibrosis [2]. When performed, CMR should follow a nonischemic cardiomyopathy protocol that includes:

- Anatomic SSFP, fast spin echo (FSE) or gradient recovery echo (GRE) single-shot images.
- Short and Long axis SSFP or GRE cine images (two-chamber, three-chamber, four-chamber) for ventricular systolic function assessment.
- Tissue scaring with LGE acquired 10-15 minutes after an intravenous bolus of contrast at the short- axis, 2-chamber, and 4-chamber views.

Other CMR modalities that can add information include:

- FSE T1- and T2-weighted imaging with or without fat saturation.
- Parametric T1 and T2 mapping, and extracellular volume at short axis.

When made, CMR findings may include:

- Apical obliteration of the left ventricle (LV) or right ventricle (RV) and atrial enlargement (Fig 4, video 4, 5 and 6) identified in cine imaging.
- Normal or mildly dilated ventricular cavity volumes and systolic function (Fig 4, video 4, 5 and 6).
- A typical scar pattern: subendocardial, not confined to a specific coronary territory, primarily affecting the apical segments of the affected ventricle, with potential extension to the ventricular septum, free wall, and atrioventricular leaflet insertion (Fig 4, video 4, 5 and 6).
- Ventricular thrombi usually located on the apical subendocardial surface (Fig 4, video 4, 5 and 6).

Potential pitfalls

Similar imaging findings can be found in other conditions different to EMF with potential confusion, those include:

- **Apical hypertrophic cardiomyopathy:** in this condition apical obliteration occurs predominantly during systole while EMF exhibits this feature during all the cardiac cycle. Apical aneurysm and thrombosis can also be potentially found. LGE pattern usually presents as patchy subendocardial or mid-wall distribution.
- **LV apical thrombosis:** conditions different to EMF associated with apical thrombosis include ischemic, dilated and Chagas cardiomyopathy, among others. CMR imaging is helpful for thrombus identification, ventricular morphological and functional as well as scar distribution assessment.
- **Excessive trabeculation of LV:** this condition can potentially coexist with ventricular thrombosis and systolic dysfunction. CMR imaging is useful for detailed morphological and scar assessment.
- **Ebstein Anomaly:** this congenital heart disease can be confused with right ventricular (RV) EMF due to the apical displacement of the tricuspid valve and the “shrunken” appearance of the RV, accompanied by dilated right atria. Distinguishing features that differentiate it from EMF include the absence of left ventricular (LV) compromise and scar on CMR.

Conclusions

Endomyocardial fibrosis (EMF) remains a neglected disease, predominantly affecting rural and low-income populations in tropical regions. Diagnosis is based on both echocardiographic and CMR findings, this last one providing tissue characterization, valuable for differential diagnosis.

Thromboembolic events, including those affecting the coronary arteries, can complicate the disease course. Prompt and accurate diagnosis, along with appropriate referral to experienced centers, can significantly improve patient management and prognosis.

Take home messages

This case highlights the importance of multimodality imaging for EMF diagnosis. CMR provides useful information for differential diagnosis.

Thromboembolic complications are among the clinical manifestations of EMF, including the coronary circulation.

References

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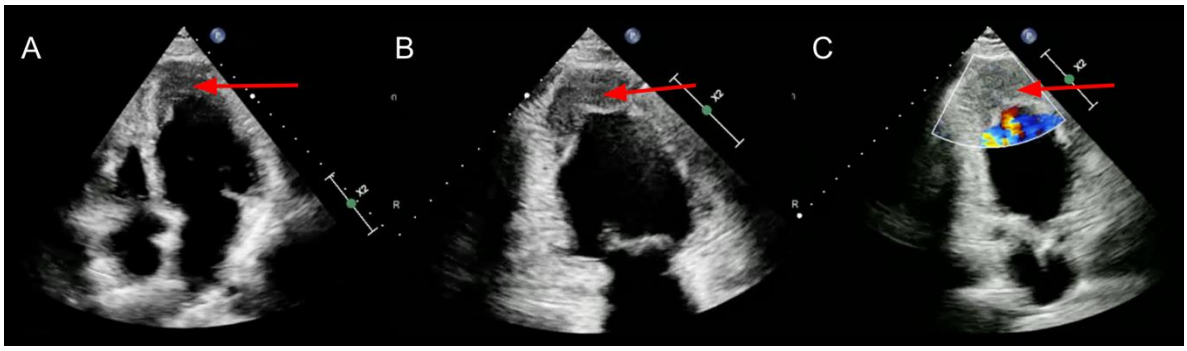

Figure 1. 2-Dimensional Transthoracic Echocardiogram

(A) Apical 4-chamber view. (B) Apical 2-chamber view. (C) Left ventricular apical obliteration and left apical thrombus (red arrows), with flow restriction shown in color Doppler assessment.

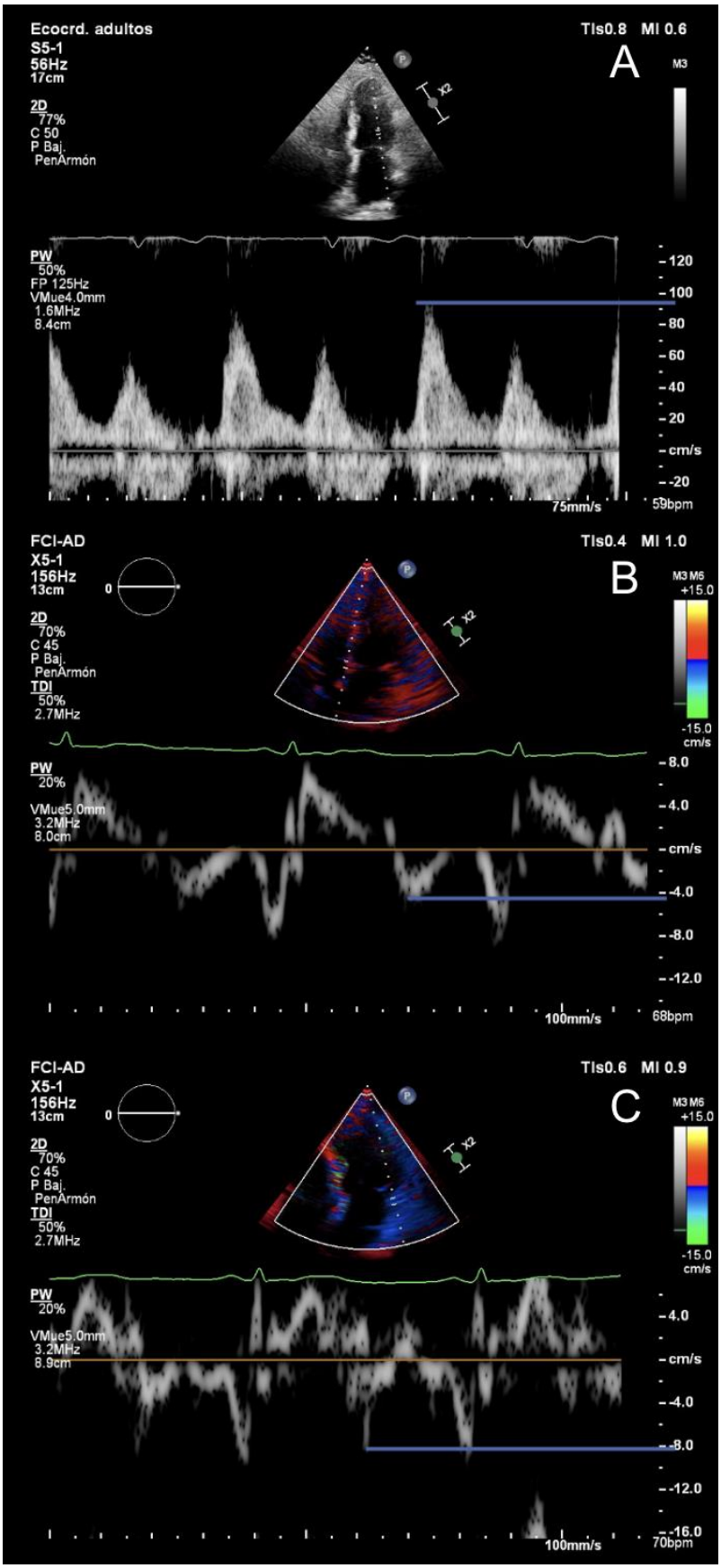
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Figure 2. Diastolic function assessment on 2-Dimensional Transthoracic Echcocardiogram

(A) Transmitral inflow pulsed wave doppler with E/A relation of 1.53 (B) Tissue doppler septal e' wave 4 cm/s (C) Tissue doppler lateral e' wave 8 cm/s and mean E/e'relation 17.7

Figure 3. Coronary angiography

View shows partially recanalized thrombosis in the mid third of the left anterior descendant artery (red arrow).

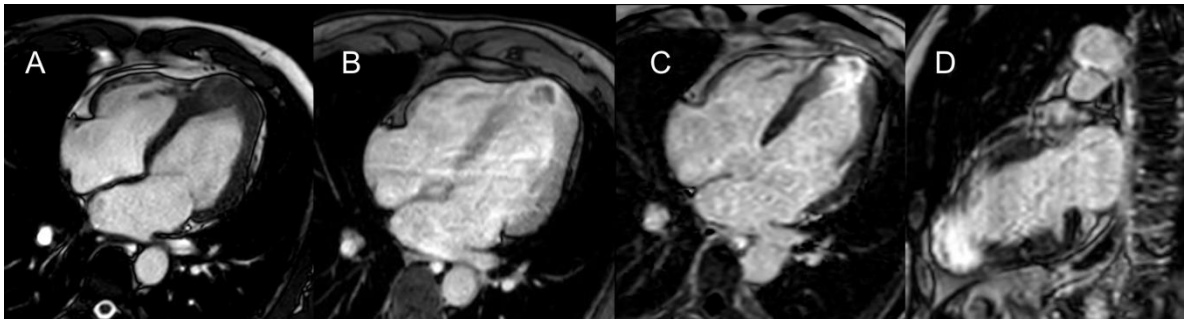

Figure 4. Cardiac magnetic resonance

(A) Cine SSFP four-chamber view showing apical obliteration and left atrial enlargement. (B) Early gadolinium enhancement imaging demonstrates apical thrombus. (C, D) Late gadolinium enhancement reveals two LV scar patterns: transmural on the anterior apical segment and subendocardial in the other apical segments.