Isolated endomyocardial fibrosis of the right ventricle

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Brief summary
We describe a case of tropical endomyocardial fibrosis isolated to the right ventricle (RV). Tissue characterization by CMR of a dilated RV with obliterated muscular part revealed diffuse sub-endocardial late gadolinium enhancement and by T1-mapping, myocardial extracellular volume was elevated on RV indicating myocardial fibrosis of the RV. This case highlights the importance of tissue characterisation to diagnose this condition.

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Case

In a 38-year-old African woman, cardiomegaly was incidentally diagnosed on a CT-scan. Transthoracic echocardiography (TTE) showed dilated right cardiac chambers with obliteration of the right ventricular (RV) cavity. A borderline blood eosinophilia (0.5 G/l; N≤0.3 G/l) was initially found, related to a parasitic infection (strongyloidiasis and filariasis), that completely reversed after treatment (0.23 G/l in 2011 and 0.05 G/l in 2019). The patient never reported typical symptoms of carcinoid syndrome, and presented with normal blood levels of chromogranine A, VIP, and pancreatic polypeptide. Only urine 5-HIAA was slightly elevated at 61 mcmol/24h (N<34 mcmol/24h), which could be attributed to a tryptophane-rich regimen. Repeated liver ultrasound and abdominal CT-Scan did not identify any neoplastic lesion, and TTE did not show any thickening or major restriction of the pulmonary or tricuspid valve supporting the diagnosis of carcinoid heart. Medical history was negative for any thrombo-embolic event, and coagulation tests (PT, aPTT, fibrinogen) and platelet count were normal. We proceeded to an endomyocardial biopsy, which demonstrated an important subendocardial and interstitial fibrosis without any inflammation, eosinophilic infiltration or granuloma (Fig 1AB). Therefore, a tropical form of endomyocardial fibrosis (EMF) was diagnosed.

The evolution over 10 years was characterized by transition to permanent atrial fibrillation and progressive dyspnea despite remarkably little peripheral oedema and no signs of left heart failure. At last visit, NT-proBNP was only moderately increased (1322ng/l) but cardio-pulmonary exercise test confirmed severely reduced aerobic capacity (peak VO_2 10.3ml/kg/min).

Repeat TTE and CMR (Figure 1C-G) showed a non-dilated left ventricle with an ejection fraction of 70%. RV inflow tract was dilated but its muscular part was almost completely obliterated. PW doppler on the tricuspid valve showed a restrictive inflow pattern. A 3cm-large thrombus was seen in a severely dilated right atrium. Tissue characterization by CMR revealed diffuse sub-endocardial late gadolinium enhancement and by T1-mapping, myocardial extracellular volume was 51% in the RV free wall, 36% in the septum and 31% in the LV free wall (N<28%) indicating bi-ventricular myocardial fibrosis with clear RV predominance.

Discussion

EMF in its chronic phase is characterized by obstructive endomyocardial fibrosis of the ventricular apex leading to restrictive cardiomyopathy. While it classically involves both ventricles, isolated RV involvement may occur. Postulated aetiologies include chronic infection, environmental and dietary factors.[1] Supportive treatment with diuretics is associated with a poor long-term outcome and surgical endomyocardial resection is linked to high immediate post-operative mortality (15-30%), which led us to discuss cardiac transplantation.[1]

Conclusion

Although EMF is rare in most parts of the world, it is one of the most frequent causes of heart failure in equatorial Africa. Patients are often asymptomatic until a late stage of the disease. At that time, survival and prognosis are usually poor.[2] This case highlights the importance of multimodality imaging and especially tissue characterisation to diagnose this condition.

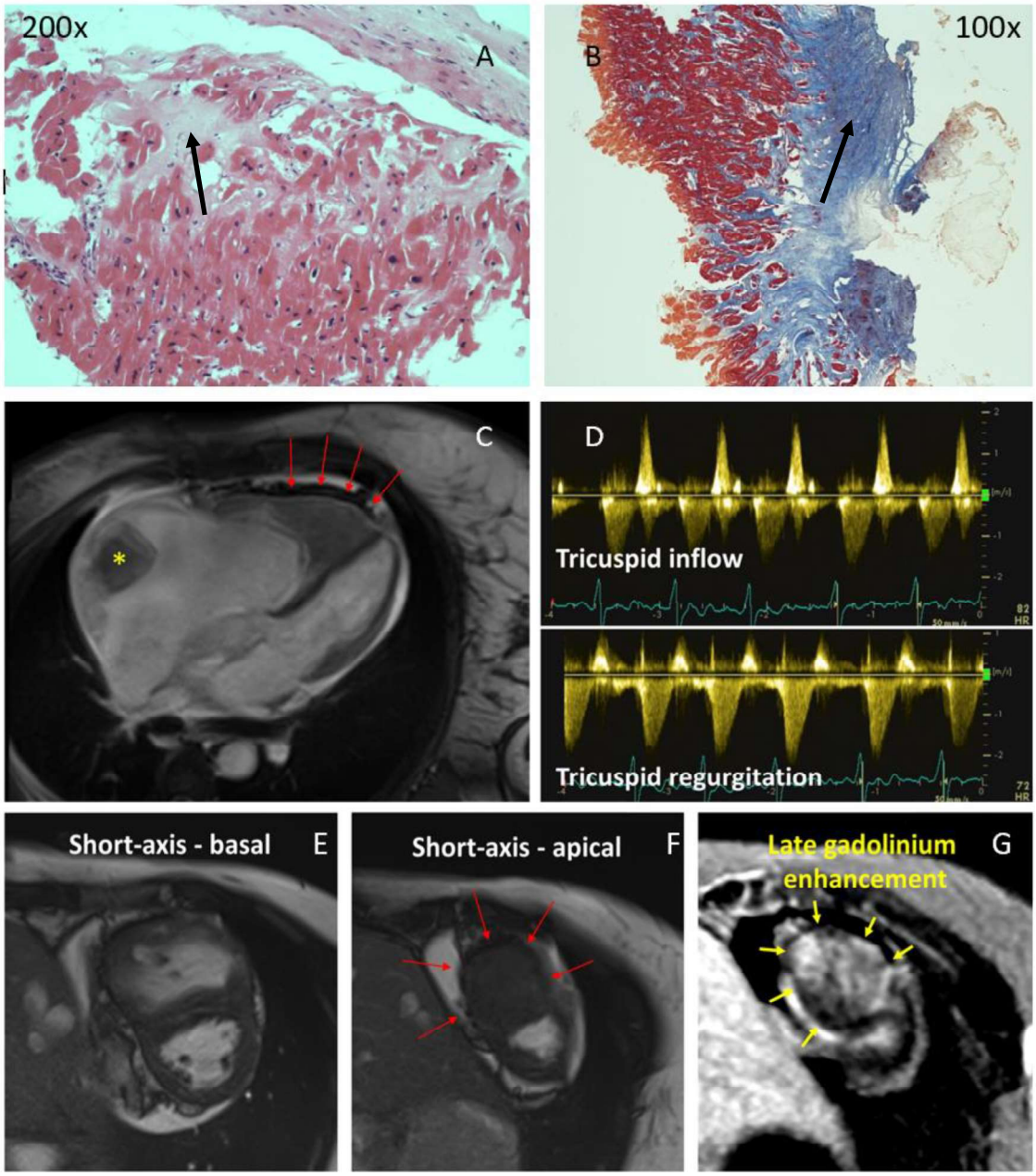

Figure 1 [A] Endomyocardial biopsy containing endocardium and myocardium with interstitial and subendocardial fibrosis (arrows) stained with H&E and [B] with Masson's trichrome (supplementary figures available with additional stains and magnifications). [C, E-G] CMR study demonstrating localized obliteration of the RV apex (red arrows) with associated fibrosis of the subendocardium (yellow arrows). A thrombus in the right atrium is seen (*). [D] TTE PW Doppler on the tricuspid valve showing restrictive inflow pattern and CW doppler through the tricuspid regurgitation showing a triangular shaped profile.

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