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CASE REPORT
Valvular heart disease

Embolic myocardial infarction secondary to marantic endocarditis in a patient with ulcerative colitis: case report

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Background Nonbacterial thrombotic endocarditis (NBTE) is an uncommon and often underdiagnosed condition characterized by a state of hypercoagulability. We present a case of a patient with ulcerative colitis who experienced a non-ST-segment elevation myocardial infarction as a rare complication of NBTE.

Case summary We report a case of a 16-year-old male with a history of ulcerative colitis who presented to the emergency department with chest pain at rest. He was admitted to the cardiology ward with an initial suspected diagnosis of acute myocarditis. Transthoracic echocardiography revealed inferior septobasal hypokinesia and a mobile pedunculated mass attached to the aortic valve, although the valve's function was not compromised.

Given the suspicion of an acute coronary syndrome of embolic origin, cardiac magnetic resonance imaging (MRI) with intravenous contrast was performed. The MRI revealed an evolved infarct in the territory of the right coronary artery. Biopsy of the material removed during surgery revealed findings consistent with thrombotic material.

Discussion We present a case of acute coronary syndrome as an unusual manifestation of NBTE in a patient with ulcerative colitis. Although we have not identified any reported cases in the literature involving ulcerative colitis, it is crucial to consider NBTE in patients with underlying conditions that predispose them to a procoagulant state. Furthermore, it is necessary to define criteria or guidelines for its diagnosis or treatment.

Keywords Nonbacterial thrombotic endocarditis • Ulcerative colitis • Non-ST-segment elevation myocardial infarction • Case report

ESC curriculum 2.1 Imaging modalities • 2.2 Echocardiography • 3.2 Acute coronary syndrome • 4.11 Endocarditis • 7.5 Cardiac surgery

Learning points

- Nonbacterial thrombotic endocarditis (NBTE) is a rare, often underdiagnosed condition associated with malignancies and autoimmune diseases; our case shows that it can also occur in ulcerative colitis patients.
- Although stroke is the most common embolic event, acute myocardial infarction can also manifest in NBTE patients.
- Treatment typically involves anticoagulation therapy, with surgical intervention indicated for larger masses.

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Introduction

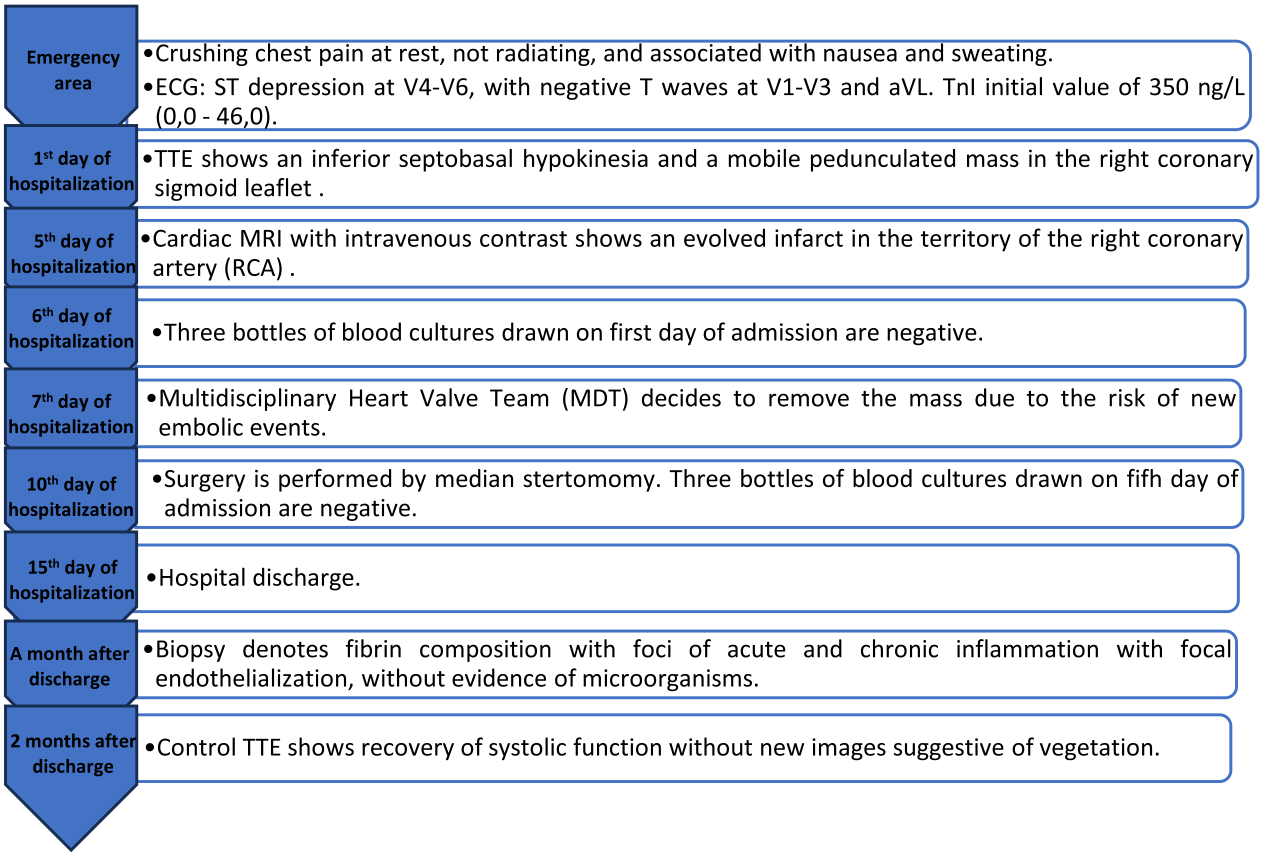
Nonbacterial thrombotic endocarditis (NBTE) is a rare condition, with a reported prevalence ranging from 1.1% to 1.6% in autopsy series.[1] Malignant neoplasms, coagulopathies, and autoimmune diseases are mainly responsible for its pathogenesis, which is based on a hypercoagulability state.[2–5]

The most common clinical manifestation of NBTE involves embolic events, typically affecting the central nervous system.[1–3] Advances in imaging techniques, particularly echocardiography, have led to an increased rate of diagnosis. Transoesophageal echocardiography (TEE) has demonstrated superior diagnostic accuracy[2,4] compared to transthoracic echocardiography (TTE), although the latter remains the initial screening tool.

Although many aspects of its therapeutic management remain unclear, addressing the underlying pathology is crucial. Anticoagulation[1,3,4] should be administered in the absence of contraindications, always assessing the risk–benefit ratio, despite the lack of guidelines regarding its duration.

We report the case of a 16-year-old male with a history of ulcerative colitis who suffered a non-ST-segment elevation myocardial infarction secondary to NBTE.

Summary figure



Case presentation

A 16-year-old Caucasian male with a history of ulcerative colitis on treatment with mesalazine 4 g and subcutaneous adalimumab 40 mg presented to the emergency department for chest pain at rest, not radiating, and associated with nausea and sweating, lasting a few minutes. He also reported a recent fever, which had triggered a flare-up of his inflammatory bowel disease.

On physical examination, the patient displayed significant pallor of the skin and mucous membranes, secondary to iron deficiency anaemia. At the time of evaluation, he was asymptomatic, normotensive, afebrile, and eupnoeic. Cardiorespiratory auscultation was unremarkable.

The electrocardiogram (ECG) showed a ST depression at V4–V6, with negative T waves at V1–V3 and aVL (*Figure 1*). Laboratory tests revealed an initial troponin I (TnI) level of 350 ng/L (0.0–46.0), which increased to 13 489 ng/L (0.,0–46.0). Based on these findings, the patient was admitted to the cardiology ward with a preliminary diagnosis of acute myocarditis.

During hospitalization, a TTE revealed inferoseptal basal hypokinesia with left ventricular ejection fraction (LVEF) of 52% and a mobile, pedunculated mass of 10× 7 mm, attached to the right coronary sigmoid leaflet, with no evidence of valvular dysfunction (*Figure 2*). This was confirmed by TEE (*Figure 3*). Three bottles of blood cultures drawn on first

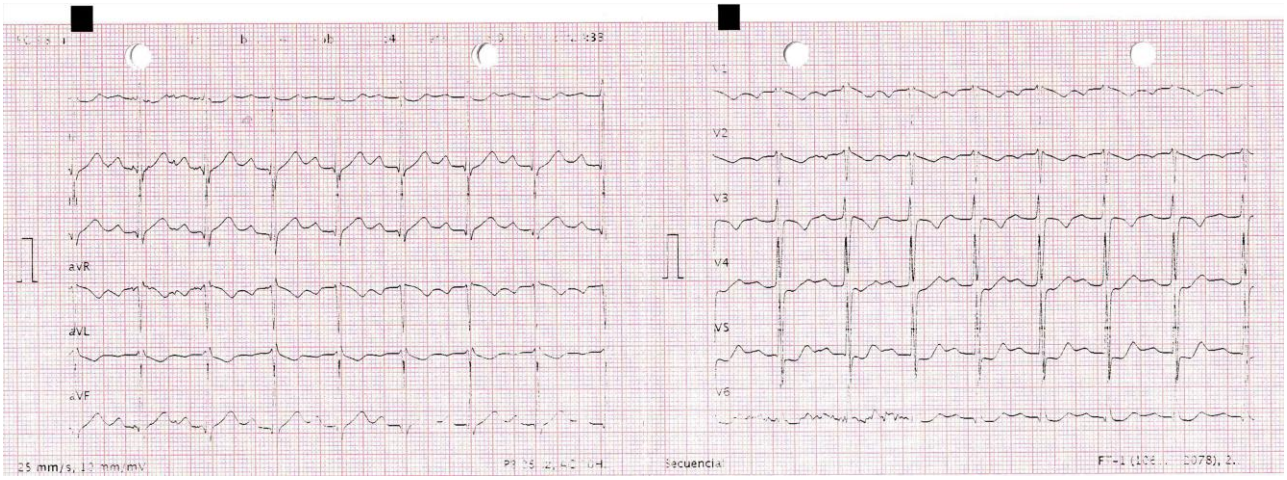


Figure 1 ECG performed in the emergency department. ST-segment depression was observed from V4 to V6, with negative T waves from V1 to V3 and in aVL.

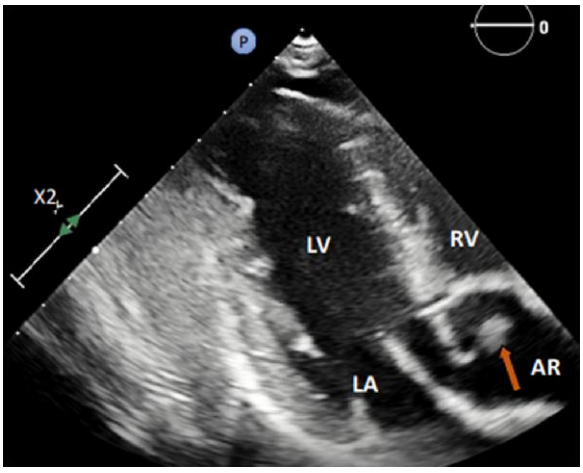


Figure 2 Transthoracic echocardiogram. Parasternal plane long axis. Nodular pedunculated mass dependent on the right coronary sigmoid valve leaflet (arrow). LV, left ventricle; RV, right ventricle; LA, left atrium; RA, aorta artery.

and fifth days of admission were negative. Given the patient's underlying predisposition, marantic endocarditis was suspected, and treatment with enoxaparin 60 mg every 12 h was initiated. Thrombophilia study came back negative. Other differential diagnoses considered included infective endocarditis and fibroelastoma.

Given the suspicion of acute coronary syndrome of embolic origin, a cardiac magnetic resonance imaging (MRI) with intravenous contrast was performed. It revealed inferobasal wall thinning with late transmural enhancement, findings consistent with a small area of chronic infarction in the right coronary artery (RCA) territory (*Figure 4*).

The Multidisciplinary Heart Valve Team recommended surgical removal of the mass via median sternotomy due to the risk of recurrent embolic events. A 2 cm mass was resected and sent for histopathological examination. After stabilization, the patient was returned to the cardiology ward, where he evolved favourably and was discharged.

Biopsy of the specimen showed a fibrinous tissue fragment with acute and chronic inflammation with focal endothelialisation (fli1+), with no evidence of signs of microorganisms (*Figure 5*).

A follow-up TTE two months after the intervention showed a recovery of systolic function (LVEF 66%), with no new images suggestive of vegetation.

Discussion

Nonbacterial thrombotic endocarditis is a rare condition, with a reported prevalence ranging from 1.1% to 1.6% in autopsy series.[1] It is most frequent between the fourth and eighth decades of life, with a slight female predominance attributed to its association with autoimmune phenomena.[2–4]

Although its pathogenesis is not well-defined, it is associated with a predisposing factor that induces a hypercoagulability state characterized by immune-inflammatory phenomena, leading to endothelial damage and the formation of an organized sterile vegetation of granulation tissue.

In susceptible populations, the prevalence rises to 15%,[3] with malignant neoplasms[5] (marantic endocarditis), particularly pulmonary adenocarcinoma, being the most frequent underlying condition.[1] A retrospective cohort study conducted at a single centre between 1999 and 2019 found that among cases of NBTE, 40.5% were associated with malignancies, 35.7% with antiphospholipid syndrome, and 33.3% with systemic lupus erythematosus (SLE; Libman–Sacks endocarditis).[2] To date, one case of NBTE has been reported in a patient with Crohn's disease,[6] although we have found no records in ulcerative colitis.

The typical clinical presentation of NBTE involves embolic phenomena, primarily in the form of stroke, which occurs in ~60% of cases.[1–4] This incidence is higher than in infectious endocarditis due to the extreme friability of the aseptic vegetations.[3] In our case, the presentation appears to be an acute coronary syndrome, an infrequent complication in the reported cases.[5,7,8] In contrast, valvular complications are generally milder and less common compared with infective endocarditis. The mitral valve is the most frequently affected (61.9%),[2] followed by the aortic valve (23.8%).[2]

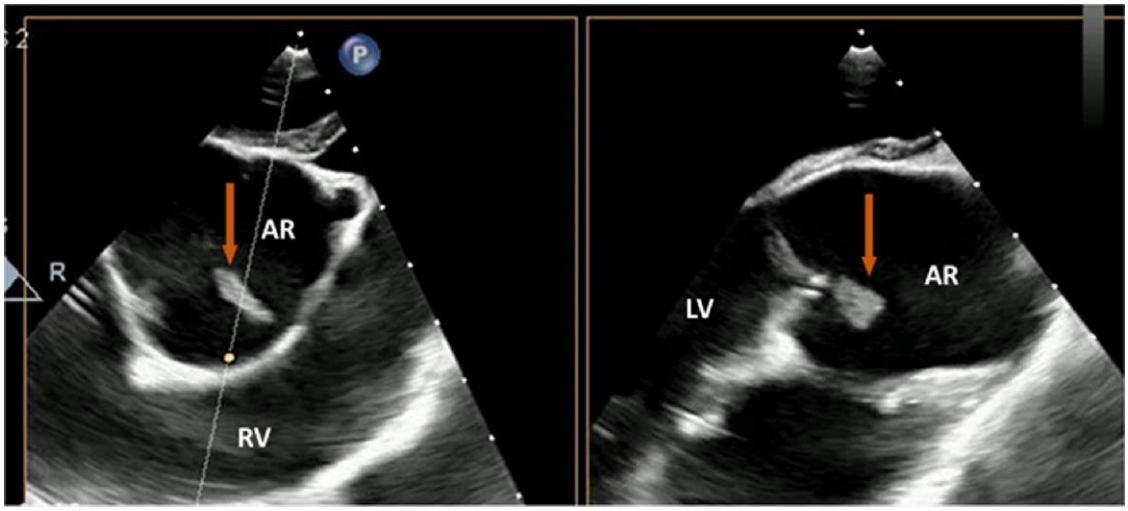


Figure 3 Transoesophageal echocardiogram. Nodular pedunculated mass dependent on the right coronary sigmoid valve leaflet (arrow). LV, left ventricle; RV, right ventricle; AR, aortic artery.

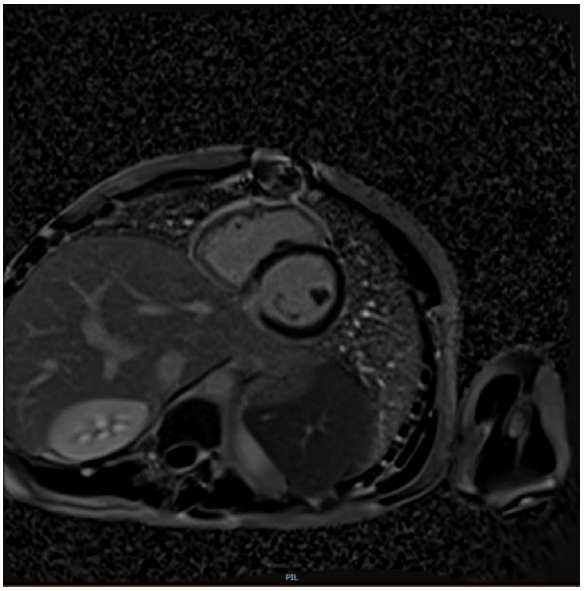


Figure 4 Cardio-MRI. Late gadolinium enhancement imaging reveals late transmural enhancement in the inferobasal region, indicative of an evolved infarction in a small area of the right coronary artery (RCA) territory.

The diagnosis of NBTE is challenging and should be considered in the presence of compatible clinical and imaging findings in a patient with an underlying condition. Echocardiography is the standard diagnostic tool, while exclusion of infectious agents through microbiological techniques is crucial. In this context, TEE is superior (2.4) to TTE, which has lower sensitivity (63%) and specificity (58%).[1] Nonbacterial thrombotic endocarditis presents heterogeneously in terms of echogenicity and morphology.

Differential diagnoses to consider include infective endocarditis and primary valve tumours.[3] In our patient, infective endocarditis was excluded based on negative blood cultures and anatomopathological findings.

The primary treatment strategy for NBTE involves addressing the underlying disease. However, initiating anticoagulation therapy with low-molecular-weight heparin, vitamin K antagonists, or unfractionated heparin is essential,[1,3,4] always assessing the risk–benefit ratio.[9] There is a current need for further evidence to establish clear guidelines on the optimal duration of anticoagulation therapy, the role of novel oral anticoagulants, and whether maintaining prophylactic doses is essential to prevent clinical recurrence. The role of cardiovascular surgery remains controversial, but it should be considered in cases with large vegetations, recurrent embolism despite anticoagulation, or severe valvular dysfunction.

In the most comprehensive review to date, in-hospital mortality rates are as high as 36%,[5] particularly in cases associated with malignancy, although this rate has decreased significantly in recent decades.[10]

Conclusion

Nonbacterial thrombotic endocarditis is an underdiagnosed condition that requires high clinical suspicion for effective screening. We have not identified any cases described in the literature involving ulcerative colitis, underscoring the importance of considering NBTE in patients with underlying conditions that predispose them to a procoagulant state. Additionally, while acute coronary syndrome is an infrequent presentation of NBTE, it should be evaluated as a potential embolic target.

Even though advances in echocardiographic imaging have favoured its identification, there are no established criteria or guidelines for the diagnosis or treatment of NBTE. Additionally, specific markers to distinguish NBTE from infectious forms are not available.

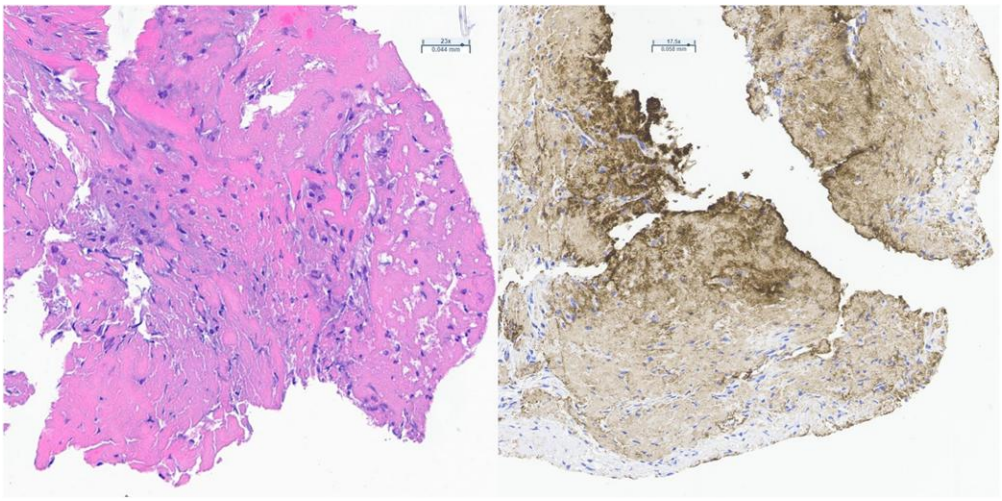


Figure 5 Biopsy of the aortic valve mass. (*A*) Haematoxylin–eosin staining showing fibrinothrombotic material with foci of mixed inflammation (acute and chronic) with no evidence of microorganisms. (*B*) Immunohistochemistry with CD61 allowing the assessment of platelet aggregates in the thrombotic phase.

Lead author biography

Diana Ladera Santos graduated in 2016 at University of Seville, Seville, Spain. She is currently working as cardiology resident at Hospital Universitario Reina Sofía, Córdoba, Spain.

Consent: The authors confirm that the patient gave written consent for the submission and publication of this case report, in line with the Committee on Publication Ethics guidelines.

Conflict of interest: None declared.

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Data availability

The data will be shared on reasonable request to the corresponding author.

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