Marantic Endocarditis Associated with Metastatic Fibrolamellar Hepatocellular Carcinoma in a Young Adult: About a Case

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Abstract

We describe the case of a 21-year-old patient presenting with marantic endocarditis of the aortic valve occurring in the context of underlying metastatic fibrolamellar hepatocellular carcinoma. The patient was admitted due to a pulmonary embolism complicating deep vein thrombosis in the right lower limb. Upon admission, the patient was hemodynamically stable and afebrile, with a poor general condition. Physical examination revealed a painful swelling in the right lower limb extending up to the thigh, hepatomegaly suggestive of a tumor, and moderate ascites. Cardiac auscultation did not reveal any murmurs or abnormal sounds. Laboratory tests showed a non-specific inflammatory syndrome, and blood cultures were negative. The electrocardiogram indicated regular sinus tachycardia. Echocardiography revealed a mobile, hyperchoic mass located on the ventricular aspect of the aortic valve, without regurgitation or stenosis. The patient was started on heparin therapy. Unfortunately, the patient passed away at home two weeks later, and no autopsy was performed.

Keywords

Marantic Endocarditis, Non-Bacterial Thrombotic Endocarditis, Hepatocellular Carcinoma, Young Adult

Introduction

Cancer is frequently associated with a state of hypercoagulability. This hypercoagulable state is multifactorial and can manifest in various ways, including venous thromboembolism, migratory superficial thrombophlebitis (Trousseau syndrome), arterial thrombosis, disseminated intravascular coagulation, thrombotic microangiopathy, and, rarely, non-bacterial thrombotic endocarditis (NBTE), also known as marantic endocarditis [1,2]. First described by Ziegler in 1888, marantic endocarditis involves sterile fibrin deposits on a native valve, typically with a high embolic potential [3-5]. We report the case of a 21-year-old male patient with metastatic fibrolamellar hepatocellular carcinoma, who presented with marantic endocarditis on the native aortic valve, along with a pulmonary embolism complicating deep vein thrombosis in the right lower limb.

Observation

This was a 21-year-old male patient admitted for the management of pulmonary embolism complicating...
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depth vein thrombosis in the right lower limb. The diagnosis of fibrolamellar hepatocellular carcinoma with peritoneal carcinomatosis had been made a few weeks earlier, and the patient was awaiting palliative chemotherapy. On admission, the patient had a poor general condition but was hemodynamically stable, with a blood pressure of 114/72 mmHg, a heart rate of 93/min, a respiratory rate of 18/min, oxygen saturation of 98% in ambient air, a temperature of 36.5°C, and a weight of 53 kg. Physical examination revealed a painful swelling in the right lower limb extending up to the thigh, hepatomegaly suggestive of a tumor, and moderate ascites. Cardiac auscultation did not reveal any murmurs or abnormal sounds. The electrocardiogram indicated regular sinus tachycardia and an S1Q3 pattern. Laboratory tests showed a non-specific inflammatory syndrome, with a white blood cell count of 11,300/mm³, hypochromic microcytic anemia with a hemoglobin level of 10.5 g/dL, and an elevated C-reactive protein at 162 mg/L. Blood cultures performed with a search for slow-growing organisms were negative, as was the cytobacteriological study of urine (Fig-1).

Echocardiography revealed a mobile, lobulated, hyperechoic mass located on the ventricular aspect of the right anterosuperior leaflet of the sigmoid valve, without regurgitation or stenosis. No other echocardiographic abnormalities were found. The patient was started on low-molecular-weight heparin as anticoagulant therapy. Unfortunately, the patient passed away at home two weeks later, and no autopsy was performed (Fig-2).

(a) The venous Doppler ultrasound at the crural level reveals a thrombus (indicated by the yellow arrow) within the vein. (b) Contrast-enhanced axial chest CT scan shows a bilateral endoluminal defect at the level of the right and left pulmonary lobes (marked by the arrows)

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Fig-2: The transthoracic echocardiography in the long-axis parasternal view during diastole (a) and systole (b) reveals a hyper-echoic mass (indicated by the arrows) attached to the ventricular aspect of the nonstenotic right anterosuperior leaflet of the sigmoid valve
Discussion

The exact incidence of marantic endocarditis (ME) is not well known. An autopsy series of 1640 adults revealed a prevalence of 1.25%. ME is more common between 40 and 80 years of age, although it can occur at any age [2,6,7]. The most commonly associated malignancies are lung adenocarcinomas, as well as those of the ovary, biliary system, pancreas, and stomach [8]. The pathogenesis is not fully understood, but the most significant factor in the formation of these friable vegetations composed of fibrin and platelets is the hypercoagulable state associated with malignancies. Elevated levels of circulating cytokines related to cancer, such as tumor necrosis factor or interleukin-1, can also contribute to local tissue damage, promoting vegetation formation. Blood flow likely plays a role in the localization and initiation of these valvular lesions [9]. Thus, ME more commonly affects the aortic valve, as seen in our patient, and sometimes the native mitral valve [10]. Rare cases involving the right-sided valves and prosthetic valves have been reported [4]. Vegetations typically localize to the free edge of the leaflets and do not significantly impair valve function. Consequently, clinical manifestations are primarily related to thromboembolic complications rather than valvular dysfunction. Arterial thromboembolic risk is high in ME, while cardiac valve dysfunction is rare [8]. In clinical practice, the diagnosis of ME in the context of neoplasia relies on the presence of vegetation detected by echocardiography and the exclusion of infection through negative blood cultures [7].

The management of these patients relies on treating the underlying cancer in conjunction with anticoagulation [3]. Additionally, many cases of marantic endocarditis (ME) occur in an underlying metastatic cancer, which limits curative options, as seen in our patient who was awaiting palliative chemotherapy. Nevertheless, these treatments offer palliative benefits and should be utilized.

Regarding anticoagulation, both unfractionated heparin and low-molecular-weight heparin have proven effective, while the impact of direct oral anticoagulants (DOACs) remains uncertain. Case reports of arterial embolic events in patients on DOACs for venous thrombosis suggest potential inefficacy [12]. Valvular surgery can be a therapeutic alternative when vegetations are large or responsible for valvular dysfunction, or in cases of recurrent embolism despite well-managed anticoagulation, provided the benefit-risk ratio is favorable [11,12]. The prognosis of marantic endocarditis remains poor, primarily due to the typically advanced stage of cancer and recurrent embolic events, particularly cerebral involvements [9].

Conclusion

Marantic endocarditis is a rare and often underdiagnosed complication of cancer. It should be considered when vegetations are detected on echocardiography, and blood cultures are negative in the context of metastatic neoplasia and associated venous or arterial thrombosis.

Conflict of Interest

The authors have read and approved the final version of the manuscript. The authors have no conflicts of interest to declare.

References

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