Cardiac Inflammatory Myofibroblastic Tumor in a Young Male Patient With Myopericarditis

Miriam Blanco, MD; Enrique Fulquet, MD, PhD, FECTS; Gregorio Laguna, MD; Gerardo Martínez, MD; Teresa Sevilla, MD, PhD; Salvatore Di Stefano, MD, PhD; Christian Ortega, MD

A 18-year-old man without a medical history was admitted to our hospital with pericarditic thoracic pain, fever, leukocytosis, and elevated cardiac enzymes. ECG showed a second-degree atrioventricular block, and transthoracic echocardiogram revealed a severe pericardial effusion (30 mm; Figure 1A). Because of signs and symptoms of cardiac tamponade, subxifoid pericardiocentesis was successfully performed, and a cloudy liquid was obtained. Empirical antibiotics were prescribed despite blood. Pericardial effusion and urine cultures were sterile because of the biochemical characteristics of the pericardial effusion. Viral serologies and Mantoux were also negative, and autoimmune disorders were ruled out. Cardiac magnetic resonance demonstrated an epicardial 21-mm-thick solid mass with homogeneous aspect, starting in the left ventricle and extending through the atrioventricular groove (Figure 1B and Movies I and II in the online-only Data Supplement). The patient had persistent fever and the thoracic pain did not disappear, so surgical intervention was decided on.

Sternum opening showed pericardial inflammation with multiple adhesions to the cardiac apex. In the left ventricle, from the atrioventricular groove to the apex, a solid heterogeneous mass with inflammatory aspect was observed, with moderate vascularization in some parts and frosted and necrotic aspect in others (Figure 1C and Movie III in the online-only Data Supplement). The mass seemed to infiltrate the myocardium, so it was not completely resected. Anatomopathological study revealed fascicular myofibroblastic proliferation (Figure 1D) without atypias and low mitotic activity, with cells expressing smooth muscle actin (α-actin; Figure 1E) and desmin (Figure 1F), supporting the diagnosis of an inflammatory myofibroblastic tumor (IMT). Anti-inflammatories were prescribed (600 mg ibuprofen every 8 hours and colchicine 0.5 mg every 12 hours), with clinical improvement. Six months after discharge, the patient was asymptomatic with descending doses of anti-inflammatories (colchicine 0.5 mg every 24 hours), and transthoracic echocardiogram shows progressive reduction of the inflammatory mass (Figure 2A and 2B).

IMT is a rare entity. Its definition includes a variety of injuries such as inflammatory pseudotumor, myofibroblastic sarcoma, and plasma cell granuloma. IMTs commonly present as a pulmonary lesion at an early age but have also been described in other organs. IMTs are frequently located in the heart, and in these cases, the tumor is located in inflow or outflow tracts conditioning obstruction or involving cardiac chambers. Exclusive epicardial location is exceptional in the literature. Anatomopathologically, IMT is a benign-tissue proliferative response including myofibroblasts and inflammatory cells. The spindle cells show no cytological atypia with a low mitotic activity.

There is no consensus on the optimal approach to IMTs. Surgical resection (when possible) followed by anti-inflammatories (including steroids) seems to be the most adequate strategy with heterogeneous results. Although the efficacy of steroids has not been demonstrated, several cases in the literature reported a decrease in tumor size with the use of steroids.

IMT is still an unknown entity. Only the search for and report of these cases will lead us to the right approach and treatment.

Disclosures

None.

References

Figure 1. A, Transthoracic echocardiogram shows severe pericardial effusion. B, Cardiac magnetic resonance reveals an epicardial solid mass. C, Surgery photo shows an inflammatory heterogeneous mass. Moderate vascularization and necrotic parts can be seen. D through F, Anatomopathological study: myofibroblastic proliferation, α-actin, and desmin expression, respectively.

Figure 2. A, Transthoracic echocardiogram at 6 months demonstrates reduction of inflammatory mass. B, Pseudotumor thickness has been reduced to 8 mm.
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_Circulation_. 2015;132:e386-e387
doi: 10.1161/CIRCULATIONAHA.115.018671

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/132/25/e386

Data Supplement (unedited) at:
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