H51 Mal D'Afrique: The Mysteries of Endomyocardial Fibrosis in Western Countries

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Learning Overview: After attending this presentation, attendees will be aware of a very uncommon cause of death in Western countries: endomyocardial fibrosis.

Impact on the Forensic Science Community: This presentation will impact the forensic science community by demonstrating the features of an uncommon cardiac disease that is rather endemic in African men under the age of 30 years old.

Endomyocardial fibrosis is caused by fibrous thickening of the endocardium, involving one or both ventricles and the atrioventricular valves. Its first description dates back to 1948 in Uganda. Epidemiologic studies report endemic prevalence in Sub-Saharan Africa and other tropical areas. For Western countries, such as Italy, however, endomyocardial fibrosis is rare and somewhat mysterious in terms of clinical manifestations, etiology/pathophysiology, diagnosis, and therapeutic management.

This report presents the case of a 21-year-old man (height 166cm, weight 50kg) from the Republic of Gambia in West Africa who suddenly collapsed at a center for asylum seekers located in Apulia, in Southeastern Italy. One week earlier, he had presented to a local physician with high fever, chest pain, and asthenia; he was prescribed antibiotic therapy for a presumed influenza infection. The man was unable to be resuscitated and was pronounced dead. A judicial autopsy was requested. External examination was notable for Janeway spots, cachexia, and the absence of traumatic injuries. The heart weighed 350g and showed a slightly increased shape and volume. Coronary arterial sectioning revealed subcritical stenosis of the left coronary artery that was complicated by a small thrombus. The right coronary artery was patent. The right ventricular wall was partially substituted by fibrosis; the left ventricle likewise revealed fibrotic remodeling. Additionally, the left ventricular apex was completely occluded by endoluminal vegetation. Histologic examination confirmed that the left cardiac ventricular wall was replaced by diffuse endomyocardial fibrosis associated with an inflammatory infiltrate consisting of lymphocytes, plasma cells, and a rich eosinophilic component that was surmounted by a large vegetative fibrinous thrombus in the initial phases of organization. No histologic evidence of acute ischemia was detected. In accordance with these medicolegal investigative results, the cause of death was attributed to endomyocardial fibrosis leading to sudden cardiac death.

In Western countries, endomyocardial fibrosis was previously considered an "obscure" or "obsolete" pathology, scarcely investigated and without specific therapies. The clinical signs are not easy to evaluate, and this makes it difficult to determine the correct diagnosis clinically and, consequently, to administer appropriate therapy. The recent increase of this disease in Western countries is closely linked to migratory flows. According to the United Nations High Commissioner for Refugees (UNHCR) data, 119,247 refugees arrived in Italy between January 1–December 31 in 2017. For this reason, in industrialized countries like Italy, endomyocardial fibrosis—along with other previously obscure cardiovascular diseases such as Chagas disease—now represent novel and emerging diseases.

Currently, there is an epidemiologic transition occurring in which cardiac diseases previously deemed obscure at certain latitudes are now on the rise due to increasing numbers of immigrant citizens. Health care professionals—including forensic pathologists—are thus advised to develop familiarity with the features of these emerging cardiomyopathies. Creating a registry of rarer cardiac entities aimed at increasing knowledge of the diseases as well as diagnostic and therapeutic strategies is suggested.

Endomyocardial Fibrosis, Cardiovascular Diseases, Restrictive Cardiomyopathy