Unusual echocardiographic features seen in a case of giant cell myocarditis

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The case of an 18-year-old college football player with a recent history of streptococcal pharyngitis who was experiencing progressive disabling dyspnea on exertion with easy fatigability and lack of stamina, and was taken to the hospital after a syncopal episode is described. The patient was initially diagnosed with heart failure and treated accordingly. However, because of a fulminant clinical deterioration, an endomyocardial biopsy was recommended, which showed focal giant cell transformation consistent with giant cell myocarditis. Treatment with methylprednisolone and cyclosporine was promptly initiated. Several apical clots were noted during treatment, but the patient attained full recovery with treatment.

Key Words: *Echocardiography; Giant cell myocarditis; Histology*

Des caractéristiques échocardiographique inhabituelles observées dans une myocardite à cellules géantes

Les auteurs décrivent le cas d'un joueur de football collégial de 18 ans qui avait récemment souffert d'une pharyngite streptococcique, qui présentait une dyspnée évolutive invalidante à l'effort accompagné de prédisposition rapide à la fatigue et de manque de vitalité et qu'on a amené à l'hôpital après une syncope. On a d'abord diagnostiqué qu'il souffrait d'insuffisance cardiaque et on l'a traité en conséquence. Cependant, en raison d'une détérioration clinique fulminante, on a recommandé une biopsie endomyocardique qui a révélé une transformation des cellules géantes focales évocatrice d'une myocardite à cellules géantes. On a vite entrepris un traitement à la méthylprednisolone et à la cyclosporine. On a remarqué plusieurs caillots apicaux pendant le traitement, qui a toutefois permis au patient de se rétablir complètement.

CASE PRESENTATION

An 18-year-old college football player with a recent history of streptococcal pharyngitis presented to the hospital after a syncopal episode. The patient had recently been experiencing progressive disabling dyspnea on exertion with easy fatigability and lack of stamina. On presentation, physical examination was remarkable for hypotension, basilar rales and jugular venous distention. An electrocardiogram showed sinus tachycardia with a left axis deviation and Q waves in leads III and aVF. Initial laboratory results showed elevated brain natriuretic peptide (8112 ng/L) and troponin I (16.8 µg/L) levels. As expected, an emergent cardiac catheterization showed elevated right-sided filling pressures with no evidence of epicardial coronary obstructive lesions. A nondilated, globally hypokinetic left ventricle (LV) was seen on transthoracic echocardiography (TTE), with an estimated ejection fraction between 20% and 25%.

Standard heart failure therapy was started while serologies for cardiotropic viruses were obtained. Lack of significant improvement during the next few days prompted another TTE, which showed further compromise in LV ejection fraction (estimated to be between 10% and 15%), with a significant amount of spontaneous echocardiographic contrast seen throughout the LV cavity consistent with a low flow state.

An endomyocardial biopsy was then recommended because of clinical deterioration and worsened TTE findings. The biopsy showed focal giant cell transformation consistent with giant cell myocarditis (GCM), as shown in Figure 1. In addition, no viral inclusions were seen, Grocott-Gomori methenamine-silver and acid-fast bacilli stains were negative and histological examination showed no Aschoff bodies. Treatment with methylprednisolone and cyclosporine was promptly initiated, resulting in significant clinical improvement. The patient was then discharged a week after initiating this aggressive immunosuppressive therapy.

Two weeks after discharge, the patient was examined because of complaints of a tingling sensation with dusky discoloration over the

Figure 1) Biopsy showing a florid inflammatory infiltrate with myocyte dropout as well as myocyte disruption (black arrows). In addition, scattered multilobulated or Langhans-type giant cells (white arrow) were also identified

fourth and fifth digits of his right hand. A TTE obtained at that time showed four distinct, very large and highly mobile homogeneous echodensities suggestive of clots in the LV apex with complete resolution of the previously seen spontaneous echocardiographic contrast (Figure 2). The patient was admitted and aggressive anticoagulation therapy with fractionated intravenous heparin was initiated with eventual resolution of the LV apical clots and no further clinical evidence of systemic embolization. He was then discharged home on maintenance cyclosporine, oral steroids and warfarin therapy. A repeat TTE three months later showed normal biventricular size and

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Figure 2) Transthoracic echocardiography. Two-chamber apical view showing left ventricular apical clots (arrows)

systolic function; the patient remained stable with no further evidence of heart failure.

DISCUSSION

GCM represents a specific form of myocarditis that generally has a more aggressive and fatal course than other forms of myocarditis. The most common presentation of GCM myocarditis is congestive heart failure (75% of cases), followed by ventricular arrhythmias (14% to 50%), acute myocardial infarction (6%) and complete heart block (5%). Compared with lymphocytic myocarditis, GCM has a markedly worse prognosis with a median survival of 5.5 months and a rate of death or cardiac transplantation of 89% (1,2).

Despite the variable use of endomyocardial biopsy in the diagnosis of myocarditis, there is a well-established criterion for the histopathological identification of GCM (2). Current guidelines suggest the use of immunosuppressive therapy to improve survival and suggest that this intervention is beneficial for GCM in both the native and transplanted hearts due to a high likelihood of recurrence (2).

At the University of Pittsburgh (Pittsburgh, Pennsylvania, USA), patients who present with new-onset heart failure do not routinely undergo endomyocardial biopsy. Given its inherent risks, a biopsy should be reserved for cases in which the results will influence the diagnosis and more importantly, the subsequent management. However, when younger patients present with progressive and fulminant acute heart failure, the diagnosis of GCM must be considered and excluded. Because GCM responds favourably to immunosuppressive therapy, an endomyocardial biopsy may be necessary to confirm the diagnosis and initiate prompt treatment, as in our case. Other clinical situations in which a biopsy may be warranted include heart failure in the presence of eosinophilia and a rash, as well as when there is suspicion for infiltrative diseases such as amyloidosis or sarcoidosis (3).

The combination of atrial fibrillation and a hypercoagulable state, as the result of enhanced platelet activation likely mediated by a combination of vascular endothelial dysfunction and reduced formation of the platelet inhibitor nitric oxide, are likely variables that increase the risk for thromboembolic events in patients with myocarditis and congestive heart failure (2,4). Two-dimensional echocardiography has been useful in the identification of thrombus in the LV.

Figure 3) Tissue Doppler imaging showing evidence of left ventricular dyssynchrony, as demonstrated by the strain curve differences between the left ventricular lateral wall and septum (A,B,C). Synchronicity was restored three months after aggressive immunosuppressive therapy was initiated (D)

In the present report, we described a case of fulminant GCM that not only developed a significant amount of spontaneous echocardiographic contrast but also apical clots. Tissue Doppler imaging, a well-known imaging modality that allows assessment of low-frequency myocardial velocities and strain (5), was useful in identifying LV dyssynchrony during the acute onset of heart failure and was instrumental in guiding therapy until LV synchrony was eventually restored, as seen in Figure 3.

CONCLUSION

We describe the appearance of spontaneous echocardiographic contrast with a lack of LV myocardial deformation signal and ventricular dyssynchrony. Clinical improvement paralleled restoration in myocardial deformation and resolution of the ventricular dyssynchrony. To the best of our knowledge, the present case is the first in which this noninvasive technique proved useful, not only in guiding anticoagulation therapy, but also in assessing the response to immunosuppressive therapy in a patient that developed GCM.

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