GIANT CELL MYOCARDITIS

MYOCARDITIS
FOUNDATION

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DEFINITION:

(What is Giant Cell Myocarditis?)

Giant Cell Myocarditis is a very rare, type of myocarditis that severely inflames the heart muscle. The exact cause is unknown, but researchers are exploring possible causes, such as infections, an abnormality of the immune system, or something in your genes that makes you more likely to get GCM. Since 20% of people who get GCM also have autoimmune disorders, those conditions may be another cause or risk factor.

The giant cells are abnormal masses produced by the fusion of inflammatory cells called macrophages. These giant cells that attack the heart muscle cause scarring. This can quickly lead to problems with how well your heart can:

- Pump blood
- Contract all of its parts at the same time during a heartbeat
- Allow heartbeat signals to take a normal, uninterrupted route through your heart.

Life expectancy from Giant Cell Myocarditis (GCM) depends on how soon you get a correct diagnosis and the treatment starts. Given both the low occurrence of the disease, and its often-severe consequences, it is imperative that physicians become more aware of its existence and clinical presentation and the latest information about methods of treatment. The disorder, unlike other myocarditis, affects males and females in equal numbers. It can affect individuals of any age, although most cases occur in young or middle-aged adults (median age 42 years). Because GCM is often fatal, many people who develop it may need a heart transplant.

SIGNS AND SYMPTOMS:

(When to Suspect Giant Cell Myocarditis)

The onset of symptoms of GCM is often rapid. Signs and symptoms of GCM myocarditis at initial presentation can be highly variable ranging from non-specific complaints to acute diffuse heart failure. Patients may recall a recent history of gastrointestinal illness or seek medical attention for mild, non-specific viral flu-like symptoms. Most commonly, the initial presentation is of sudden onset heart failure with severe chest pain, difficulty breathing, fatigue, palpitations, lethargy, decreased exercise tolerance and or syncope (dizziness).

Initial symptoms may also include swelling of the ankles, chest pain, heart palpitations, fatigue and shortness of breath especially upon exertion or lying flat. Affected individuals eventually develop irregular heartbeats (arrythmias) such as abnormally fast (tachycardia) or slow (bradycardia) heartbeats. Arrythmias may cause sudden episodes of lightheadedness or loss of consciousness. These symptoms occur due to congestive heart failure or heart block. These two heart abnormalities are progressive and eventually result in life threatening complications.

DIAGNOSIS:

(How is Giant Cell Myocarditis Detected?)

Accurate diagnosis of GCM myocarditis is challenging. A diagnosis of GCM is made by biopsy of heart tissue. A small tissue sample of the heart is surgically removed and studied microscopically looking for giant cells.

THERAPY:

(What are the Standard Treatment Options for GCM?)

Standard and supportive treatment options for cardiac failure and arrythmias are recommended. These treatment options may include the insertion of a pacemaker or implantable heart defibrillator. Affected individuals may require a heart transplant. Affected individuals are often evaluated for heart transplantation shortly after diagnosis. In 20-25% of patients, infiltration of giant cells has recurred after heart transplantation. Immunosuppression with drugs that include cyclosporin prolong survival free of transplantation. GCM can recur after therapy in both the native and allografted heart.

CONCLUSION:

(What is the Prognosis for GCM Patients?)

Unfortunately, most cases of GCM are not diagnosed until after death. From 1905 until 1987, all cases of GCM were described at autopsy, with survival usually less than 3 months from the initial onset of symptoms. We now know that with early diagnosis by heart biopsy and prompt immunosuppressive treatment, 90% of GCM patients survive at least one year. Physician and patient awareness of the symptoms of GCM is the first step in decreasing the tragic impact of this disease. The Myocarditis Foundation seeks to raise awareness of GCM, build a community of survivors, and support research that will lead to better treatments, longer survival, and ultimately a cure.

Myocarditis and the resulting disorder of idiopathic dilated cardiomyopathy are the cause of approximately 45% of heart transplants in the U.S.

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