Uncommon history of a giant cell myocarditis

C. Auf der Maur^a, Michel Zuber^a, Michael Jörg Mihatsch^b, Paul Erne^a

^a Division of Cardiology, Kantonsspital, Luzern

^b Department of Pathology, University Hospital, Basel

Figure 1

Parasternal long axis view with M-mode echocardiogram showing depressed contraction in the septal and posterior wall as a sign of global left ventricular systolic dysfunction.

RV = right ventricle; LV = left ventricle; IVS = interventricular septum; PW = posterior wall.



Figure 2

Apical two-chamber view shows an inferior aneurysm (\leftarrow) that has a thinned acinetic wall with possible proximal thrombus formation. The thickened proximal inferior wall has subendocardial sclerosis with bright echos. LV = left ventricle; LA = left atrium.



There is no conflict of interest.

Case report

A 48-year-old woman experienced an episode of cardiac decompensation due to a third-degree atrioventricular block. Normal systolic and diastolic functions of both ventricles were documented. A dual-chamber pacemaker was implanted and eliminated the patient's symptoms. Two years later, the patient was hospi-

Figure 3 Pacemaker follow-up.





Correspondence: Prof. Paul Erne Division of Cardiology Kantonsspital Luzern CH-6000 Luzern 16 Paul.Erne@KSL.CH

Figure 4

- A–F Different biopsy fragments showing extensive destruction of the myocardium by poorly granuloma-like granulation tissue composed of irregularly distributed giant cells, inflammatory cells and fibrous tissue.
- F A central areactive necrosis is present.



talised due to biventricular heart failure and wide QRS tachycardias. Pacemaker follow-up revealed a continuous deterioration of the pacing threshold and of the evoked action potential combined with a congestive car-

diomyopathy which was documented by echocardiography (fig.1-3). Granulomatous giant cell myocarditis (GCM) was diagnosed following endomyocardial biopsy (fig. 4-5). Since the patient declined evaluation for

Figure 5

Massive T-cell accumulation – in equal amounts helper (CD4) and suppressor (CD8) T-cells – in the periphery of the granuloma. No B-cell (CD20) infiltration (B), few histiocytes/macrophages (CD68) within the granulation tissue. The giant cells are of histiocyte/macrophage origin (CD68 positive).



Figure 6

The M-mode echocardiogram in the parasternal long axis view eleven months later documents normalisation of left ventricular function with good contractility in the interventricular septum (IVS) and the posterior wall (PW).

 RV = right ventricle; LV = left ventricle; IVS = interventricular septum; PW = posterior wall.



heart transplantation, immunosuppressive treatment with cyclosporine, azathioprine and prednisolone was established. This resulted in subjective improvement and regression of inflammation and granulomas, but was accompanied by progressive renal failure. Echocardiography showed a remarkable recovery of left and right ventricular function (fig. 6). The patient's condition has remained stable for the past 4 years.

References

- 1~ McCarthy RE $3^{\rm rd},$ Boehmer JP, Hruban RH, Hutchins GM, Kasper EK, Hare JM, et al. Long-term outcome of fulminant myocarditis as compared with acute (nonfulminant) myocarditis. N Engl J Med. 2000;342: 690–5.
- 2 Cooper LT Jr, Berry GJ, Shabetai R, for the multicenter Giant Cell Myocarditis Study Group Investigators. Idiopathic giant-cell myocarditis – natural history and treatment. N Engl J Med. 1997;336(26): 1860–6.