This is the published version of a paper published in *International Cardiovascular Forum Journal*.

Citation for the original published paper (version of record):

Unusual arrhythmogenic myocardial disease.
http://dx.doi.org/10.17987/icfj.v1i4.52

Access to the published version may require subscription.

N.B. When citing this work, cite the original published paper.

Permanent link to this version:
http://urn.kb.se/resolve?urn=urn:nbn:se:umu:diva-107225
Unusual arrhythmogenic myocardial disease

Christer Backman MD, Bengt Johansson MD, PhD, Erik Tossavainen MD, Michael Henein MD, PhD

Umeå Heart Centre and Department of Public Health and Clinical Medicine, Umeå University, Umeå, Sweden

A twenty-two-year-old lady, with a 2-year history of recurrent tachyarrhythmia, presented with an episode of arrhythmia preceded by sore throat followed by chills, cough and exertional breathlessness, while abroad. She was commenced on penicillin V for 10 days with no improvement and because of worsening cough with purulent sputum she was prescribed broad-spectrum antibiotics and her chills disappeared. Two months later she had a relapse when she felt extremely tired, when a casualty ECG showed right bundle branch block and anterior T-wave inversion. An echocardiogram showed normal size cardiac chambers but the left ventricular basal posterior wall was hypokinetiic with a bright echodense 2x3 cm subendocardial segment suggestive of localized fibrosis (Figure 1). The same finding was confirmed with Gadolinium enhanced CMR scan (Figure 2) which suggested a prior myocardial injury. The ejection fraction was normal at the time of CMR.

On telemetry she had periods of supraventricular tachycardia. Chest-x-ray showed normal cardiothoracic ratio and clear lung fields. High sensitivity CRP was mildly raised to 5 (<3), as was high sensitive Troponin-T to 33 ng/l (<14.5) and pro-BNP to 474 pg/l. Electrolytes and electrophoresis were normal. Serological tests were all negative including CMV, EBV, parvo B 19, enterovirus, mycoplasma, Clamydia pneumoniae and Borrelia. Only Adenovirus titre was slightly raised. Three blood cultures as well as urine, sputum and pharynx cultures were all negative. PCR from the pharynx (Clamydia pneumoniae, Mycoplasma pneumoniae) and nasopharynx (Enterovirus, Influenza A and B, Metapneumovirus, Clamydia pneumoniae) as well as PCR from stool, (Enterovirus) were all negative. Also DNA tests for clamydia pneumoniae, mycoplasma and RNA, enterovirus and influenza A virus were all negative. The patient received heart failure treatment including ACE-inhibitors and beta blockers and was discharged on broad-spectrum antibiotics. Serum-ACE levels were normal, thus excluding sarcoid disease.

Three months later both Troponin-T and pro-BNP remained unchanged and Troponin-I was significantly raised to 296 (<16), despite progressive recovery of the chest leads T wave inversion (Figure 3). A recent exercise ECG showed a maximum work load of 160 Watt and was stopped because of fast self terminated SVT at a rate of 250 bpm when she developed breathlessness of the same nature as that of the original presentation.

Discussion: the main findings of this case suggest an evidence for localized myocardial damage involving the basal posterior wall of the left ventricle with established subendocardial fibrosis. The progressive improvement of electrical changes is consistent with recovered inflammatory process affecting the anterior wall, this is despite the continuing troponin rise. The localized organized segmental fibrosis of the basal posterior wall raised questions. Firstly, whether it was related to the current presentation of 3 months history of illness or more likely a subclinical episode more than 2 years ago when the patient started experiencing tachyarrhythmias. Myocardial fibrosis takes 6 months, at least, to fully develop to the extent of what was confirmed by imaging in this lady. The remaining raised troponin suggests the possibility of an ongoing myocardial inflammation.

Figure 1: Echocardiographic long axis view (left) and short axis basal view (right) showing basal posterior fibrosis

...
of some sort which needs to be followed. Finally, the recurrent similar episodes of tachyarrhythmia and SVT coinciding with the patient’s breathlessness raises the possibility of arrhythmogenic myocardial disease.

**Correspondence:**
Bengt Johansson MD PhD
Umea Heart Centre, Umea, Sweden
Bengt.johansson@medicin.umu.se

**Figure 2:** CMR apical views (left) and short axis view (right) showing basal posterior wall myocardial fibrosis

**Figure 3:** ECG changes over a follow up period.

*ECG at admission* *ECG at follow-up*