Reviewer's report

Title: Myocardial fibrosis as the first sign of cardiac involvement in a male patient with Fabry disease. Report of a clinical case and discussion on the utility of the magnetic resonance in Fabry pathology.

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Reviewer: Ali Yilmaz

Reviewer's report:

Sechi et al. describe an interesting case of a male Fabry patient who underwent a CMR study and was found to be LGE-positive in the LV inferolateral wall “without” any signs of LV hypertrophy. Moreover, the presence and pattern of LGE did not change in this patient following ERT.

Comparing their findings in this patient with previous data (particularly from Niemann et al, JACC Cardiovasc Imaging 2011) that suggested that the development of myocardial fibrosis does not necessarily require myocardial hypertrophy in “female” patients compared to “male” patients, Sechi et al. convincingly argue that the development of myocardial fibrosis does not require myocardial hypertrophy in “both” (!) male and female patients with Fabry disease.

Major compulsory revisions:

- Recently, Thompson et al. (Circ Cardiovasc Imaging 2013) published T1-mapping results in patients with Fabry disease. Amongst others, these authors stated that “sex differences in non-contrast T1 and ECV were independent of wall thickness” and concluded that “reduced non-contrast myocardial T1 values are the most sensitive and specific cardiovascular MRI parameter in patients with Fabry disease irrespective of sex and LV morphology and function”. Hence, Sechi et al. are right when arguing that “presence of LGE” (!) does not require myocardial hypertrophy in “both” (!) male and female patients with Fabry disease. However, this finding does not disprove the notion that there are some sex differences in the development/pattern of cardiomyopathy in patients with Fabry disease. The authors should address this important issue and the study of Thompson et al.

Minor essential revisions:

- Figures: If possible, the authors should show comparative CMR images (cine as well as LGE images) of this patient prior to and post ERT.
- There are some minor typos that should be corrected throughout the manuscript …

Level of interest: An article of importance in its field
Quality of written English: Acceptable

Statistical review: No, the manuscript does not need to be seen by a statistician.

Declaration of competing interests:
I declare that I have no competing interests