Reviewer’s report

Title: Juvenile elastic arteries after 28 years of renal replacement therapy in a patient with complete Complement C4 deficiency

Version: 2 Date: 30 August 2012

Reviewer: Joseph M Ahearn

Reviewer’s report:

This is a truly remarkable case report by Knoll et al. The authors clearly describe a patient with Henoch Schoenlein Purpura who had been on renal replacement therapy for 28 years. The extraordinary observation is that this patient, despite a full range of risk factors for vascular damage, demonstrated carotid artery intima media thickness below the normal range and normal pulse wave velocity. In contrast the patient's peripheral muscular arteries were heavily calcified. The authors attribute this unusual finding to an inherited complete lack of complement C4 protein due to genetic mutations. Normal elastic arteries despite prolonged renal replacement therapy in this patient is suggested to support the hypothesis recently proposed by Shields et al. that direct binding of C3- and C4- activation fragments to elastin and collagen fibers in the vascular wall of otherwise healthy individuals may contribute to the development of vascular stiffness.

The report could be improved by addressing the following points:

Discretionary Revisions

1) In the introduction, it is stated “no effective therapeutic strategy has been identified today until today”. This statement suggests that an effective therapy has now been identified. This is misleading and should be rephrased.

2) It would be interesting to learn more about the pathology reports from the patient’s renal tissues that may have been biopsied and/or removed. In particular it would be interesting to know the patterns of complement deposition that were observed both in the patient’s autologous tissue and in the failed allografts.

Level of interest: An exceptional article

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.