

4 September 2018

Dr Ruo-Yu Ma

Science editor

World Journal of Nephrology

Manuscript number: 41582

Thank you for giving us the opportunity to resubmit our revised manuscript entitled “Awakening the sleeping kidney in a dialysis dependent patient with fibromuscular dysplasia” to the *World Journal of Nephrology*. We thank all the reviewers for their feedback, which we have carefully considered and have revised the manuscript in accordance to their recommendations. We hope that the manuscript is now suitable for publication in the *World Journal of Nephrology*.

Sincerely,

Yazied

Dr MY Chothia, MBChB, MMed, FCP(SA), Cert Nephrol (Phys) SA

Divisions of General Medicine and Nephrology, Department of Medicine, Faculty of Medicine and Health Sciences, Stellenbosch University and Tygerberg Hospital, Cape Town, South Africa.

Reviewer #1 (00503339):

The planned management of the Left Kidney (shrunken) should have been included to complete the recounting of what appears to be brilliant management.

Response: We would like to thank the reviewer for their comments. Regarding the shrunken left kidney, we did not attempt revascularization or any other intervention because we considered it to have negligible contribution to overall kidney function.

The following sentence has been added to the 8th paragraph of the case description: 'Due to the negligible contribution to overall kidney function, no interventions were planned for the left kidney.'

Reviewer #2 (00503279):

This is an interesting manuscript for some cases we face in clinical practice. It is well written and comprehensive.

Response: We would like to thank the reviewer for their comments.

Reviewer #3 (02888410):

1. The summary needs to be improve. It do not explain the importance of the case report, specially, that the patient was in hemodialysis.

Response: We humbly disagree with the reviewer regarding these points. We have clearly highlighted the importance of evaluating patients with renal artery stenosis and dialysis dependence for kidney viability and restoration of kidney function with a revascularization procedure. Also, the title of the manuscript, the abstract and the conclusion all clearly highlights that the patient was receiving dialysis.

2. Introduction is too short.

Response: We agree with the reviewer on this point. We have therefore added more detail regarding fibromuscular dysplasia. The following has been added to the 1st paragraph of the introduction: “The latter is regarded as a non-atherosclerotic, non-inflammatory vascular disease which most frequently affects the renal arteries (60-75%) followed by the carotid arteries (25-30%), however; involvement of many other vascular beds have been described [1]. The pathological classification of FMD is based on the arterial layer primarily affected. The most common pathological type is medial fibroplasia, which has a characteristic “string-of-beads” appearance on renal angiography. The cause of FMD remains unknown but may have a genetic component since the disease tends to affect first-degree relatives of affected individuals [2]. The disease is frequently asymptomatic and may only be identified coincidentally. Studies have reported that FMD represents < 10% of cases of renovascular hypertension [3]. Even more uncommon is renal failure due to FMD despite studies reporting that up to 63% of patients have loss of kidney volume [4].”

3. Case report lack of data about treatment; specifically, the use of renin-angiotensin-axis blockers and the timeline of its use with the evolution of the renal function.

Response: Renin-angiotensin aldosterone antagonists were avoided pre- and post-operative due to concerns regarding the effect that this class of drugs may have had on renal function. We have added the following sentence to the 5th paragraph of the case description: “...Her antihypertensive regimen included atenolol 50 mg daily, furosemide 160 mg daily, amlodipine 10 mg daily and minoxidil 5 mg daily. Antagonists of the renin-angiotensin system were avoided due to the concern of the effect that this class of drugs may have had on renal function.”

4. Was a microscopic study of renal artery sample performed? Explain the results.

Response: Unfortunately, no specimens were sent for histological assessment.

5. How do the authors explain the obstructive uropathy after surgery? Could it be a main cause of renal function?

Response: The exact cause for the post-operative obstructive uropathy was not identified but our urology colleagues suspected that there may have been a kinked ureter related to the perinephric hematoma. In the case description, the first sentence of the 9th paragraph has been changed as follows: “Approximately 1 week post-operatively, she complained of right flank pain. An ultrasound examination revealed a right perinephric haematoma and *suspected kinking of the ureter causing hydronephrosis.*”

6. Discussion needs a thoroughful description of previous cases.

Response: To the best of our knowledge there has only been one other case of suspected FMD-associated, dialysis dependent renal failure described in the literature (reference 20). This patient received dialysis for 6 months before a spleno-renal bypass procedure was attempted. He remained dialysis-free for 17-months at the time of writing the case report. We amended the 6th paragraph of the discussion as follows: “..., a patient who was dialysis-dependent for 6 months became dialysis-independent following surgical correction that involved a *spleno-renal bypass procedure*”.