Author’s response to reviews

Title: Case report: Scleroderma with crescentic glomerulonephritis

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Version: 8 Date: 9 January 2008

Author’s response to reviews:

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Version: 7 Date: 24 December 2007

Author's response to reviews: see over

Comments to the authors by Dr. Andrew Short:

General: The authors have addressed many of the points raised. However it is not possible for them to determine whether the acute renal insult reported is due to ANCA related vasculitis because they do not have access to the techniques. Specialists in Nephrology and Rheumatology will be sceptical of the claim that this is a rare manifestation of scleroderma rather than a possible and much recognized ANCA related vasculitis. The authors do acknowledge this to some extent. The immune staining is interesting and does lend some support to
the authors' claim. The presence of a crescentic nephritis requiring specific treatment from that used for scleroderma renal crisis is important and it would be appropriate to highlight this in a general journal. Recognising the possibility of alternative pathology and investigating appropriately is crucial in this complex disease. I feel this should be the emphasis of the paper and then it can be published.

Reply: Text has been modified as suggested.
Specific comments: Authors use mg/dl for creatinine in page 3 and micro mol/L in page 4.
Reply: Corrected to Micromol/L.

2. Doppler study of native renal arteries is inadequate in our experience to exclude renal artery stenosis.
Reply: Accepted that doppler study is inadequate to exclude RAS. In our patient we did not proceed with other investigations like renal artery angiogram as we did not suspect renal artery stenosis in our patient.

3. Revisions necessary for publication: A slight change of emphasis to highlight the different causes of renal disease in scleroderma and the need to determine by appropriate investigation which disease is present in any individual patient.
Reply: The changes suggested in the text have been made.

Reviewer's report
Title: Case report: Scleroderma with crescentic glomerulonephritis
Version: 5 Date: 7 November 2007
Reviewer: PETER HEWINS
I am familiar with the literature and believe that this case meets one of the 7 criteria for evaluation in the journal: An unexpected association between diseases or symptoms
Has the case been reported coherently?: Yes
Is the case report authentic?: Yes
Is this case worth reporting?: Yes
Is the case report persuasive?: Yes
Does the case report have explanatory value?: Yes
Does the case report have diagnostic value?: Yes
Will the case report make a difference to clinical practice?: Yes
Comments to authors:
General
This manuscript is much improved.
Revisions necessary for publication

1) I do not agree with the statement that 'pulse methyl prednisolone .... can be considered the standard of care' for crescentic GN. In lupus nephritis (classes iii and iv), ANCA-associated GN and anti-GBM nephritis, for example, there is clear evidence that steroid only therapy is inadequate and additional therapy is needed. Cyclophosphamide is widely used and in the cases of anti-GBM nephritis and ANCA-associated GN with severe renal failure, plasma exchange would be the standard of care. For lupus nephritis, mycophenolate may be an alternative to cyclphoshamide. The text should be amended accordingly.

Reply: Text has been amended accordingly.

2) Moreover, recovery from dialysis dependent renal failure is possible (particularly for ANCA and lupus nephritides) and histopathological changes are often patchy. Accordingly, the statement that 'She did not respond to Methyl prednisolone also probably due to the presence of fibrous and fibro cellular crescents' is one which could be misinterpreted and should be amended. With the benefit of hindsight, a trial of additional immunosuppressive therapy would probably be have been appropriate.

Text has been amended suitably.

3) I would interpret the histopathology and immunofluorescence as suggesting a immune complex mediated disease - it would be appropriate to indicate this. I would still contend that the features and consistent with a lupus nephritis. If serum C3 and C4 levels were assayed, these should be recorded. If the authors consider that the pathologic findings some other particular etiology, they should state this.

Histopathology is interpreted as immune complex mediated disease. We do not consider lupus nephritis as underlying disease here because C3 C4 levels are normal and the immunoperoxide staining did not reveal a full house immunoglobulin deposits. Patient also did not have clinical features of SLE.

4) I recommend that the authors make specific reference to HCV assocaited MCGN. It is important to be mindful of HCV (and other infectious) causes of crescentic GN particularly in the context of features an of immune complex disease and before considering immunosuppressive therapy. In this particular case, the absence of purpura and other extra-renal manifestations, together with the absence of endoluminal thrombi make a cryoglobulinemic GN less likely but
these are not invariably present. It would be worth noting if the patient were rheumatoid factor +ve or had abnormal liver enzyme levels. Patient was negative for third generation anti HCV antibodies and had RA factor level of 8 IU /ml and did not have abnormal liver enzyme levels. Text is suitably amended.

What next?: Accept after minor revisions

Quality of written English: Acceptable