Author's response to reviews

Title: Optic atrophy, necrotising anetrior scleritis and keratitis presenting in association with Streptococcal Toxic Shock Syndrome: a case report

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Author's response to reviews: see over
Dear Mr Bremner,

Thank you for reviewing our manuscript. Here are our responses to your comments:

**Case Report:**

1. **P3, Para 2, Line 6** - The “pupil” has been corrected to “iris” - although there was no transillumination the texture of the iris surface had the appearance of diffuse atrophy, it was also slightly paler in colour. This has now been amended in the text. We postulate that an ischaemic/vasculitic process may account for this change in the appearance of the iris.

2. **Figure 1** - There was no rubeosis on examination, this has been clarified in the text. The fundal view was clear with no evidence of intraretinal haemorrhages or pallor. The retinal vasculature appeared normal. There was subepithelial opacification in the cornea in three zones with no staining on fluorescein - we propose that this was an ischaemic effect. The epithelium was intact. This is now mentioned in the legend and in the text.

3. **P3 Para 3 Lines 8-9** - We are in agreement that a diagnosis of ION cannot be made as the patient did not report sudden onset of visual loss - she was for a period of time intubated and unconscious so there is a period when she was unable to report to the medical team of visual loss - and secondly the disc was not swollen (it may have been initially but we don’t know). We agree that “optic atrophy” is the correct term to use in this context and this has been amended in the title, abstract and text.
4. **P3 Para 3 Lines 8-9** - We have amended the term of “necrotizing anterior uveitis” to “post streptococcal uveitis”. We have re-evaluated the medical history and further reviewed the literature on the subject and consider that in this case the patient had a combination of processes such as an ischaemic vasculitis resulting in a degree of anterior segment ischaemia (hence an anterior necrotizing scleritis and keratitis) with evidence of post-streptococcal uveitis manifesting with extensive posterior synechiae and recurrent IOP elevations that required topical and systemic treatment. We also believe that as the patient was evaluated by the bed side with a hand-held slit lamp it is possible that a subtle cellular reaction in the anterior chamber was present but was not observed.

5. **P3 Para 2** – We have now amended the title, abstract and text and have included the term “necrotizing anterior scleritis” in the title, abstract and text.

6. **P3 Para 2 last line** - The eye retained PL vision at 6 months after presentation, the eye was comfortable and the anterior staphyloma was unchanged. This is now clarified in the text.

**Discussion:**

1. **P4 Para 2-3** - the “cotton wool spot” is now mentioned as part of the case report. The specific “immunological tests” are also part of the case report.

2. **P5 Para 2, Line 3** - We have removed the term DIC as we feel it has no relevance to the ocular findings discussed.

3. **P5 Para 2, Line 3** - The diagnosis of toxic shock syndrome was made by the medical team. The patient presented with pyrexia, shock, an erythematous rash of the extremities and multi-organ failure. She subsequently developed skin
desquamation of the lower limbs below the knees and ischaemic necrosis leading to amputation. These points are now clarified in the text

4. We agree that the most likely mechanism of disease is an immune complex mediated vasculitis affecting the anterior segment. This resulted in ischaemic keratitis and necrotizing anterior scleritis. We do not think that the patient developed an endogenous endophthalmitis as the vitreous was clear with no inflammatory membranes. This is now made clear in the text.

Having reviewed the medical notes and history we propose that post streptococcal uveitis may explain the inflammatory signs in the anterior segment such as the ciliary injection, posterior synechiae, iris “atrophy” and the intraocular pressure rise. The latter signs may suggest that the inflammatory component may have been present for some time before a referral was made to ophthalmology resulting in the above ocular changes.

Thank you for your time,

On behalf of all authors,

Dr K. Papageorgiou M.D.