Author's response to reviews

Title: Weill-Marchesani syndrome with advanced glaucoma and corneal endothelial dysfunction: a case report and literature review

Authors:

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Author's response to reviews: see over
Dear Editor:

Thank you very much for allowing us the opportunity to submit a revision. We appreciate the reviewers detailed comments and valued suggestions. Our responses to the comments are listed as follows:

**Reviewer 1**

**Response to Q:**

Thanks for the comments. This patient had a 13 years history of glaucoma in both eyes and her right eye has been blind (no light perception) for over 10 years due to severe glaucoma. As she is a monocular patient, it brings a great challenge to us when making the treatment decision. When she came to our hospital, she presented with severe optic neuropathy (extremely small tubular visual field) and a quite high IOP (49mmHg) which need to be lowered as soon as possible to prevent further damage to the optic nerve. For this consideration, we gave the trabeculectomy at the next day after her admission. As she is a monocular patient, we tried to minimize the risk of complications and preserve vision in her sole seeing eye. The patient would have to take more risk of complications including extinction phenomenon or expulsive choroidal hemorrhage when performing lens extraction under such a high IOP. That’s why we did not choose lens extraction at the beginning. Sincerely hope this explanation can better justify our choice of surgery for this specific patient. We have further polished the language in this manuscript.

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**Reviewer 2**

**Response to Q1:**

Thank you so much for your detailed comments and professional suggestions. We
totally agree that the first appropriate approach to malignant glaucoma is to use mydriatic-cycloplegics and aqueous suppressants. In this case, the patient had a 13 years’ history of glaucoma and her right eye has been blind (no light perception) for over 10 years due to severe glaucoma. When she came to our hospital, she presented with severe optic neuropathy (extremely small tubular visual field) and a quite high IOP (49mmHg) which could not be lowered by mannitol. As she is a monocular patient, we tried to lower the IOP immediately to prevent further damage to the optic nerve and preserve her vision. For this consideration, we gave the trabeculectomy at the next day after her admission in order to lower the IOP as soon as possible. Atropine was applied at the same time in expectation to resolve the pupillary block.

Response to Q2:
Thank you for your suggestion of performing UBM examination for this case. Because the patient had a long history of glaucoma and showed severe optic neuropathy and a quite high IOP which need to be alleviated as soon as possible, thus we chose to lower IOP immediately to prevent further damage to the optic nerve. This is a great suggestion and we will keep in mind with the value of UBM for doubtful patients in future.

Response to Q3:
As she is a monocular patient, we tried to minimize the risk of complications and preserve vision in her sole seeing eye when we make the treatment decision. At the same time, the high IOP needed to be alleviated immediately. Base on above reasons, we decided to perform trabeculectomy, which is a relatively safe surgery, aiming to lower the IOP as soon as possible together with atropine. The patient would have to take more risk of complications including extinction phenomenon, expulsive choroidal hemorrhage or choroidal detachment when performing lens extraction under such a high IOP. If trabeculectomy had failed to control the high IOP, we might turn to lens removal and anterior vitreous removal. Fortunately, the IOP was lowered dramatically after trabeculectomy for this patient. But we totally agree with your
opinion that phaco-trabeculectomy would have been the surgery of choice for this case if we had diagnosed the microspherophakia from the beginning. The concept we would like to express and share in this manuscript is what we can do and what we can learn from such a special and rare condition. We would like to share our experience and lessons learned in treating this rare case. Especially, we are so glad that we have really learned a lot from your professional and valued comments.

Thanks again for your efforts and time spent on reviewing our manuscript. We sincerely hope that all the comments have been addressed to your satisfaction. Looking forward to hearing your positive decision.

Sincerely,
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