Reviewer's report

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

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Reviewer: Ivano Riva

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This is an interesting clinical report about a case of angle closure glaucoma in a patient affected by Weill-Marchesani syndrome. Authors properly describe clinical features of this rare condition and explain their therapeutic approach. The paper is well written and the language is appropriate. However some points need to be addressed before publication.

Weill-Marchesani disease is a syndrome characterized by brachydactyly, short stature and joint stiffness. Microspherophakia and lens luxation are common features of this disease, as well as secondary angle closure glaucoma. This is a consequence of spherophakia and anterior luxation of the lens, which induce pupillary block. Angle closure chronicization brings to high IOP not responsive to medical therapy.

In the case described by Authors a young patient presented to their attention, with high IOP (49 mm Hg), extremely narrow anterior chamber and iris bulged forward. The patient had been already diagnosed with glaucoma and already underwent glaucoma surgery (which surgery?) 13 years before. Due to the clinical picture and patient's history, Authors suspected a malignant glaucoma and performed a trabeculectomy. After surgery, dilating pupil with atropine, Authors diagnosed patient's microspherophakia and lens anterior displacement, with endothelial damage. Phacoemulsification and IOL implantation guaranteed IOP control and corneal clarity.

Major Compulsory Revisions

#1: Malignant glaucoma is a challenging diagnosis. Generally, it nearly follows intraocular surgeries, such as cataract extraction or trabeculectomy, in predisposed eyes. The first approach to this condition is medical, with aqueous suppressants and mydriatic-cycloplegics. In phakic patients, atropine paralyzes the iris sphincter muscle and the ciliary muscle, tightening the zonula and helping to pull the lens posteriorly (Note well: atropine doesn’t induce zonular relaxation, but increases zonular tension, due to ciliary muscle palsy). In the case described by Authors, a first approach with atropine and aqueous suppressants should have been tried, in suspicion of malignant glaucoma, also if the time interval from first surgery was not prone to this diagnosis: pupil dilatation would have revealed the microspherophakia from the beginning, changing the surgical approach. Moreover, the use of dilatators would have probably resolved the pupillary block, creating a communication between the posterior and the anterior chamber, due
to the small dimensions of the lens and the superior dislocation.

#2: High-frequency ultrasonography biomicroscopy (UBM) is extremely useful for diagnosis of doubtful cases, like the one described in the paper. It allows identification of irido-corneal touch, appositional angle closure, anterior rotation of the ciliary body and ciliary body-iris. We consider UBM a fundamental step in diagnosis of malignant glaucoma. Examination of this case with UBM would have promptly revealed the microspherophakia and the lens displacement, allowing for a correct diagnosis.

#3: Trabeculectomy alone is a poor choice in case of malignant glaucoma: it doesn’t resolve the anterior displacement of the iris-lens diaphragm and has a high risk of failure. Lens removal, in combination with posterior capsulectomy and anterior vitreous removal, is the first surgical step in malignant glaucoma. Trabeculectomy is rationally performed in combination with/after these procedures, to guarantee IOP control in the long term, considering angle-closure chronicization and/or trabecular damage. So it is not clear why Authors opted at first instance for trabeculectomy alone, suspecting a malignant glaucoma.

Few reports have shown that lens extraction alone may be sufficient to manage glaucoma in spherophakia.(1-5) It is likely that lensectomy alone may be sufficient to control IOP in cases with acute pupillary block or lens dislocation, but may fail if there is significant synechial angle closure or there are angle anomalies. Because patient’s blurred vision and symptoms were persistent from at least 1 year, we think that phaco-trabeculectomy would have been the surgery of choice in this case (as stated by the same Authors in conclusions), allowing for optimal IOP control and minimizing the risk of complications.


Level of interest: An article whose findings are important to those with closely related research interests

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