An Atypical Clinical Presentation of Brown McLean Syndrome Progressing to Central Corneal Decompensation

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INTRODUCTION

A patient presents with chronic bilateral peripheral corneal edema secondary to Brown McLean Syndrome. Ocular examination reveals bilateral central corneal decompensation, which is atypical for this rare syndrome.

CASE HISTORY

Patient TC, a 73-year-old African American male, presented for a glaucoma follow-up and complained of mild blurry vision in the morning OU. His past ocular history is notable for congenital cataracts s/p CE in 1969 with resulting aphakia OU (performed at an outside office, reason for aphakia unclear), glaucoma secondary to longstanding aphakia with resulting angle abnormalities OU, Brown McLean syndrome OU (diagnosed 2019). His past medical history is notable for hypercholesterolemia and vitiligo. He is currently taking latanoprost qhs OU and dorzolamide-timolol bid OU.

OCULAR EXAMINATION

OD	Exam	OS
cc: 20/30+ OD, PH 20/25-	**VA**	cc: 20/50 OS, PH 20/30 OS
FTFC	**CVF**	FTFC
FROM	**EOMs**	FROM
longstanding fixed pupils with superior corectopia due to superior iridectomy	**Pupil**	longstanding fixed pupils with superior corectopia due to superior iridectomy
15 mmHg	**GAT**	11 mmHg
See Photos	**SLE**	See Photos
See Photos	**DFE**	See Photos

DIAGNOSIS AND DISCUSSION

This patient was diagnosed with corneal decompensation secondary to Brown-Mclean Syndrome (BMS). BMS is a rare, static annular corneal edema that affects the peripheral 2-3mm of the cornea. It is typically seen many (6-16) years s/p cataract surgery in eyes left aphakic and occurs more commonly in eyes s/p intracapsular cataract extraction (ICCE). There is no associated corneal neovascularization or anterior chamber inflammation. Central involvement is rare, and when present is usually transient. Corneal decompensation (a change in corneal cellular morphology) is rare, but present in this case. There is a possible correlation with high myopia, but it is not well understood. The etiology of BMS is unknown, but initially was believed to be caused by underlying endothelial dystrophy or a possible genetic component. The pathophysiology of BMS is also unknown. Endothelial trauma is not necessary to induce BMS; non-surgical cases can be caused by lens subluxation, spontaneous lens resorption, endotheliitis, keratoconus, angle closure glaucoma, or myotonic dystrophy. Potential complications include bullous keratopathy due to persistent corneal edema and infectious corneal ulcerations secondary to ruptured bullae.

FIGURE 1a
Slit lamp examination OD shows 2+ microcystic edema (MCE) over inferior 60% of cornea, superotemporal iridectomy, aphakia.

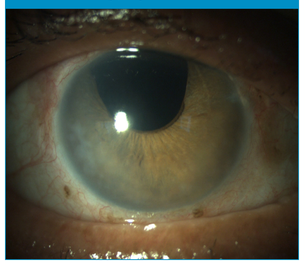


FIGURE 1b
Optic section of cornea OD shows stromal hyperreflectivity, indicating stromal edema.

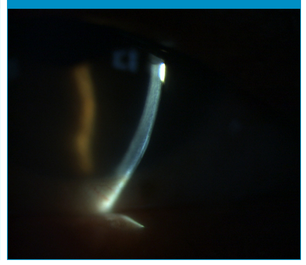


FIGURE 2a
Slit lamp examination OS shows 1-2+ MCE from 3:00-9:00, large spot of endothelial pigment superotemporally, superonasal iridectomy, aphakia.

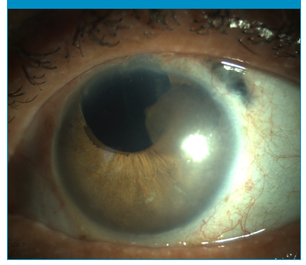
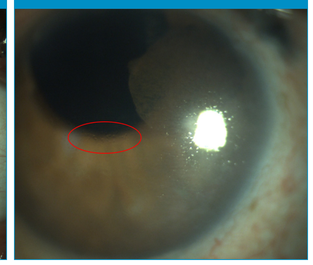
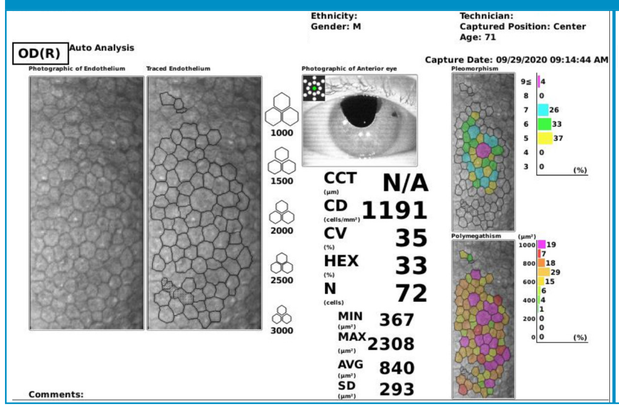
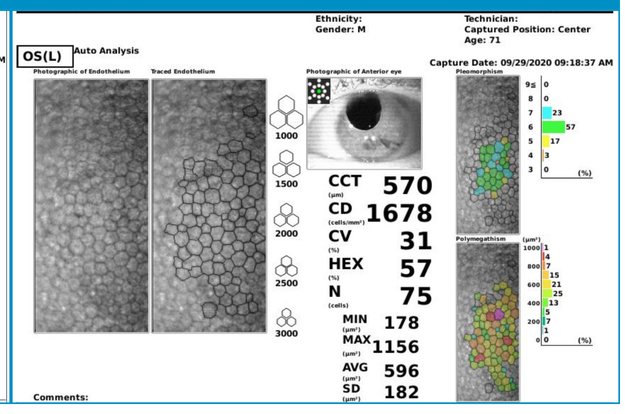


FIGURE 2b
Corneal guttata present centrally OS.



FIGURES 3a & 3b
Specular microscopy OU shows deceased cell density centrally (age norm 1800-2600 cells/mm2) with polymorphism and pleomorphism.

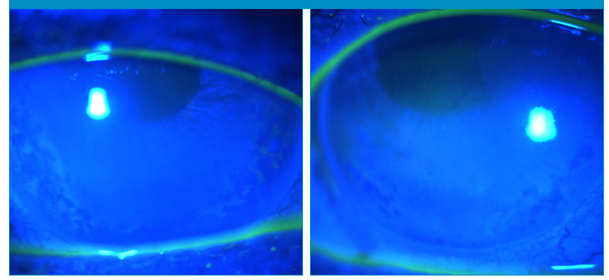




TREATMENT AND MANAGEMENT

Treatment with Muro 128 ung qhs OU was initiated to decrease blur in the morning. This patient's displaced pupils cause him to only have mild, transient symptoms since his visual axis is mostly spared. Symptoms will be monitored at follow up examinations (patient currently seen every 4-6 months for glaucoma monitoring) and specular microscopy will be repeated annually to monitor corneal decompensation. A further decrease in endothelial cell count or development of complications may warrant a corneal transplant (DSEK, DMEK, PK). Long term hypertonic use is warranted in some cases, but BMS can often remain untreated if the patient is asymptomatic and the central corneal endothelium is intact.

FIGURES 4a & 4b
At 4-week follow up examination, patient's symptoms resolved and NaFl staining shows resolved central corneal edema with persistent peripheral edema.



CONCLUSION

BMS is a rare corneal condition that does not typically lead to corneal decompensation. Due to ICCE falling out of favor in recent years, BMS is not commonly seen in the present day with the development of new cataract extraction methods such as phacoemulsification. Patients with BMS are usually asymptomatic and can be left untreated, but if symptoms develop the condition can be treated with over-the-counter hypertonic medications. However, it is important to counsel patients regarding transient visual symptoms and the possibility of developing complications (such as bullous keratopathy) that may warrant surgical intervention.

REFERENCES

References available upon request.

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