

The clinical presentation and outcome of Mooren's ulcer at Ruharo Eye Centre, Southwestern Uganda; a hospital based retrospective study

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ABSTRACT

Objective: To describe the clinical presentation, management, and treatment outcomes of Mooren's ulcer in Uganda.

Methods: We reviewed records of all patients diagnosed with Mooren's ulcer at Ruharo Eye Centre from January 2013 to December 2015. We collected data from fifty-two patient charts on demographic characteristics, history and presenting complaints, eye affected, clinical presentation, modes of medical and surgical treatment, and post treatment visual acuity.

Results: The median age was 24.5 years (IQR 16.5 years). The ratio of males to females was 8:1. Thirty two (82.1%) patients presented with pain, 13 (33.3%) with redness, and 10 (25.7%) with tearing. Forty seven (90.4%) patients had unilateral disease. Four (80%) of those with bilateral disease were aged 30 years and above while one (20%) was below 30 years. Twenty nine (55.8%) of the patients received only medical treatment and 23 (44.2%) received various forms of surgical management. Forty one (78.8%) still had active ulceration on their last review date, 7 (13.5%) perforated after admission, 3 (5.8%) of the patients were marked as healed on their last review date, and one (1.92%) was eviscerated. At the last follow up, most patients ended with worse visual acuity (n=21, 40.4%) while 20 (38.5%) had no change and only 11 (21.2%) improved.

Conclusion: This primary study provided background information on presentation of Mooren's ulcer in Uganda; our data indicated that younger males were the most affected group; severe disease occurred in older patients.

INTRODUCTION

Mooren's ulcer is a chronic and painful ulceration of the cornea that often starts in the periphery and may gradually progress centrally or circumferentially to eventually involve the entire cornea¹. The ulcer sometimes involves the full thickness of the cornea and leads to perforation². This disease commonly presents with severe ocular pain and in late stages, there is destruction of the peripheral corneal stroma. This destruction often results in vascularisation of the cornea and corneal perforations may occur in some cases. The exact pathogenesis remains unclear but it is generally agreed upon that Mooren's ulcer is an autoimmune disease which targets the cornea without other systemic disease association³. Recent work in Mooren's ulcer patients identifies antibodies specific to the antigen Calgranulin C which is found uniquely in corneal stroma⁴.

Mooren's ulcer is a disease that is often seen in healthy adult men with no evidence of systemic disease though a case has been documented in a 3 year old child⁵. It may occur in any sex and age. Studies

done in India and China show that median age ranges between the 6th and 8th decades⁶ even as the prevalence is 0.03%¹.

Mooren's ulcer often presents with a red, tearing, painful eye, and decreased vision. The ulcer begins peripherally and moves circumferentially often involving the limbus if allowed to progress. Severe ulceration may lead to perforation¹. Diagnosis depends on the exclusion of any potential infective or rheumatological causes. This means extensive blood and radiography investigations are required.

There is no trial evidence for any of the interventions used to treat Mooren's ulcer¹. However, treatment is in a stepwise manner and progress from one step to another depends on patient's response. Initial therapy involves intensive steroid use, both topical and oral. If the patient's response is not favourable within 7 – 10 days, conjunctival resection up to bare sclera is the next step. The conjunctiva adjacent to the ulcer is thought to have cytokines which increase inflammation and antibodies against the cornea⁷. Multiple resections may be required. Cases, which fail to improve with steroids and conjunctival resection,

require immunosuppressive therapy. Surgical intervention with superficial lamellar keratectomy can reduce inflammation, tissue adhesives used for small perforations and corneal grafts for larger ones. Corneal grafts in active Mooren's ulceration have a poor prognosis though⁸. Despite the stepwise process, immunosuppressive therapy should be reserved for more severe forms though it's often better to start it sooner than later⁹. At Ruharo Eye Centre (REC), there is currently no standardized method of management of Mooren's ulcer and while various ophthalmologists choose different medical and surgical interventions, no studies exist that show if some work better than others do. Although rare, Mooren's ulcer tends to be more common in Africa and India worldwide, which implies a geographic and genetic predisposition. Africans tend to have a worse prognosis than Caucasians⁴.

MATERIALS AND METHODS

In this descriptive hospital based case series chart audit, data were extracted for all patients who were diagnosed with Mooren's ulcer at REC from January 2013 to December 2015. REC is an associate training institution for ophthalmology residents from Mbarara University of Science and Technology (MUST).

Demographic characteristics such as sex, age, address; and clinical variables, which included history, clinical presentation, treatment, and follow-up, tabulated. We collected data using a questionnaire, entered into Epi-Info version 3.5.4, and STATA version 12.0. We tabulated the results. We sought ethical permission from the head of department.

RESULTS

There were fifty two patients' charts collected and all were analysed for baseline characteristics and demographic data. However, only 39 of the charts were analysed for history and the rest excluded due to missing data. Table 1 demonstrates the demographic data of all 52 patients. The age ranged from 11 years to 72 years. There were 46 males enrolled and the male to female ratio was 7.67:1, the median age was 24.5 years; 24 (46.2%) patients enrolled first presented in 2015, and 15 (67.3%) patients lived within less than 100 km distance from REC.

Table 1: Demographic data of patients with Mooren's ulcer attending REC from 2013 to 2015 (n=52)

Gender (Male : Female)	7.67 : 1	(%)
Age (years)		
Youngest	11	
Oldest	72	
≤ 30 years	34	65.4
> 30 years	18	34.6
Median	24.5 (IQR 16.5)	
Mean	29.1	
Year of first presentation	Frequency	
2013	14	26.9
2014	14	26.9
2015	24	46.2
Distance from REC	Frequency	
≤ 100 Km	35	67.3
> 100 Km	17	32.7

Of the 39 charts that were analysed for history, 32 (82.1%) patients presented with pain, 13 (33.3%) patients with a red eye, and 10 (25.7%) patients with tearing. Of the 52 patients, 31 (59.6%) of them presented with Mooren's ulcer in the left eye, 16 (30.8%) with disease in the right eye, and 5 (9.6%) presented with bilateral disease. Of the five cases with bilateral disease, 4 (80%) of them were in patients 30 years and older. Nine (17.3%) cases presented with a perforated ulcer while six (11.5%) cases presented with an impending perforation (Table 2).

Table 2: Baseline characteristics and clinical presentation of patients with Mooren's ulcer attending REC from 2013 to 2015 (n=39)

Presenting symptom	Frequency	(%)
Pain	32	82.1
Reduced vision	8	20.5
Tearing	10	25.7
Foreign body sensation	10	25.7
Redness	13	33.3
Use of TEM	7	17.9
History of trauma	6	15.4
Affected eye		
Right eye	16	30.8
Left eye	31	59.6
Both	5	9.6
Impending perforation	6	11.5
Perforated	9	17.3

Of the 52 charts that were analysed for management, 28 (53.9%) patients received a form of antibiotic treatment while 13 (25%) patients got prednisolone tablets and 8 (15.4%) patients got prednisolone eye drops. Thirty two (61.5%) patients had their initial treatment changed due to poor progress. Table 3 displays the various forms of initial treatment that were given; 29 (55.8%) patients received only medical

treatment while the rest received some form of surgical management.

Table 3: The different forms of initial treatment administered to patients with Mooren's ulcer at REC from 2013 to 2015 (n=52)

Medical management	Frequency	(%)
Antibiotic drops	16	30.8
Antibiotic cream	28	53.9
Atropine	20	38.5
Prednisolone drops	8	15.4
Prednisolone tablets	13	25
NSAID	9	17.3
Change in treatment	32	61.5
Surgical management		
No surgery	29	55.8
Conjunctival flap	15	28.9
Conjunctival resections	5	9.6
Corneal graft	2	3.9
Cryotherapy	1	1.9

We took the outcome of treatment for the 52 charts reviewed on the date last reviewed. Three (5.8%) patients healed, seven (13.5%) patients got perforated ulcers after admission, and one (1.92%) patient got an eviscerated.

Table 4 shows the various outcomes. Of the 24 patients that came for review at least 3 times after date of presentation, 18 of them lived in areas less than 100 km from REC.

Table 4: Outcomes of treatment of Mooren's ulcer in patients at REC from 2013 to 2015 (n=52)

Clinical outcome	Frequency	(%)
Healed	3	5.8
Perforation after admission	7	13.5
Eviscerated	1	1.92
Change from baseline V/A		
Improved	11	21.1
No change	20	38.5
Declined	21	40.4
Number of reviews		
At least 3 times	24	46.2
No review	16	30.8

There was no statistical association with the decline in visual acuity ($p < .002$), whether the patients received only steroid therapy, only antibiotic therapy or both.

DISCUSSION

The median and mean ages of the patients were much lower than studies done in different races like in China where the mean age was 48.8 years¹ or in India where the median and mean ages were 65 and 61 years. A case series published by Kietzman¹⁰ in Nigeria showed an aggressive form of Mooren's ulcer being

more prominent in young males between the ages of 20 to 30 years. This led to the belief that a malignant form of the ulcer occurs more in young, black males⁵. Another study done in Nigeria showed the mean age to be 30.9 ± 16.6 years¹¹ which is very similar to the mean age found in this study, possibly indicating that there's a different type of Mooren's ulcer occurring in Africa.

Similar to previous studies, males outnumbered females in this study however, the male to female ratio was 7.67:1. This value is much higher than previous studies done in Caucasians which showed that men were only 1.6 times more likely to get Mooren's ulcer than women⁵. The study done in Nigeria by Fasina *et al*¹¹ had the likelihood closer at men being 3.6 times more likely to contract the disease than women.

Lewallen and Courtright¹¹ attributed this increased risk to factors like men having more ocular trauma than women but this may be different in other regions. The study also showed that 4 of the 5 cases of bilateral disease were in older people, which is in line with previous findings⁵. The authors found more left eye involvement than right eyes but did not find any causative reasons.

There are two types of clinical Mooren's ulcer described. The first type is benign, occurring in older patients and is bilateral in 25% of the patients while the second type is malignant, occurring in younger patients and is bilateral in 75% of the cases¹². The study found however that even though most of the cases occurred in younger patients below 30 years of age, the bilateral disease happened in the older patients, which is what we consider the malignant form.

The rate of perforation of the ulcers was 17.3% which is much lower than the rate reported by Kietzman¹³. Mooren's ulcer is often found to occur after a history of trauma or ocular surgery⁶ and this study showed that 15.4% of the cases reviewed had a history of trauma. This is close to the 10.7% history of trauma found in China¹ and the 17% found in India¹⁴.

Mooren's ulcer is difficult to treat and can be relentless in spite of various attempts at managing it⁵. Treatment is often aimed at promotion of healing and corneal re-epithelialization, as well as stopping the process of destruction¹⁰. This has led to the generalized belief that a step-wise approach is best in management of the ulcer, starting with steroid therapy, then surgical therapy, and immunosuppression. This study revealed however that a fraction of the patients received some form of steroid therapy. 55.8% of the patients received only medical therapy while the rest had surgical intervention including conjunctival resections, conjunctival flaps, cryotherapy, and corneal grafts. The 32.7% of the patients who lived more than 100 km from REC (town centre) were likely from low economic backgrounds. This affects how many of them are willing to pay admission and surgery fees for

effective management. With the mean age being 29.1 years and most of the patients being male, few would be willing to spend more time than they would think necessary in a hospital while their families depend on them in the village areas.

CONCLUSIONS

- (i) This primary study provided information on the clinical characteristics, management and treatment outcomes of patients with Mooren's ulcer at REC.
- (ii) The disease affects a young population even though we observed a more aggressive form in older patients.
- (iii) There is no clinical trial basis for the treatment decisions however and a prospective study to describe the pattern of disease in South-western Uganda as well as the specific risks and outcomes of the different treatments would be of great value¹.

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