

Corneal Perforation of Mooren's Ulcer In Melanoderm: About 13 Cases



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Abstract

Introduction: Corneal perforation is frequent in black people presenting Mooren's ulcer. We tried to evaluate in this study the characteristics of this pathology in our practice context.

Patients and method: We performed a prospective study of 4 years concerning black people suffering of Mooren's ulcer and presenting perforated cornea at presentation. They received corticosteroids and cyclophosphamid in eye drops as medical treatment. The surgical procedure was made of conjunctivectomy associated with corneal sutures or amniotic graft.

Results: Thirteen eyes of eleven patients were involved during this period. The mean age was 29.54 years with a sex ratio of 0.57. Blindness was found in 9 eyes whereas 4 eyes were in visual impairment. Conjunctivectomy associated with corneal sutures was performed in 76.92 % of cases and amniotic graft in 23.08%. We found recurrence in 30.77% of cases and in univariate analysis, the pejorative facts of recurrence were the female sex, a age under 30 and a time to go hospital more than 3 months.

Conclusion: Perforated cornea seen in Mooren's ulcer is a sight-threatening pathology in black people. Recurrence is frequent and must be known by the patient and the specialist.

Introduction

Mooren's ulcer (MU) is a peripheral or marginal corneal ulceration looking like a crescent whose anterior margin forms a very characteristic promontory over hanging the bottom of this ulceration [1]. Evoked for the first time by Bowman in 1849 through one case report, MU was described in 1867 as a single clinical entity by Mooren. It belongs to the group of peripheral ulcerative keratitis and occurs completely in absence of any diagnosable system in order that could be responsible for the progressive destruction of the cornea. It is a rare pathology but its frequency seems to be higher in Africa, China and India than in northern hemisphere. The exact pathophysiology remains uncertain, although there is evidence suggesting an autoimmune basis. Mooren's ulcer is a painful, relentless, chronic ulcerative keratitis which begins peripherally and progresses in three directions: circumferentially, centrally and in depth. The clinical presentation is a red eye with photophobia, watering and visual impairment and the diagnosis requires exclusion of the other disorders that could be responsible for the progressive destruction of the cornea. At last with any treatment, this pathology can progress and reaches descemetocoele and corneal perforation. The management of MU is very difficult with no codification and requires medical and surgical procedures at the stage of perforation. Recurrence is frequent and the prognosis could therefore be threatened. Epidemiological, clinical and

the epidemiological data of perforated Mooren's ulcer are scarce, especially in West Africa and mainly in Côte d'Ivoire. So it seems fairly reasonable to bring our contribution to the evaluation of this pathology through the description of epidemiological features, clinical aspects, our practice therapeutic approaches in the aim to appreciate evolution and prognosis of MU at the stage of perforation in African black people.

Patients and Method

We performed a prospective study from January 2010 to January 2016 in the university hospital of Treichville in Abidjan. This study obtained the approval of the local ethics committee. After a complete eye examination, African black people suffering of Mooren's ulcer and presenting perforated cornea at presentation were included in the study. The patients had to undergo medical and surgical treatment with a regular follow-up during the study period. They received topical corticosteroids before the surgical treatment. This surgical procedure was conjunctivectomy associated with either corneal sutures or amniotic graft. Post-operative treatment was topical corticosteroids and eye drops of cyclophosphamide at 1% obtained by magisterial preparation. Statistical analysis was performed with Epi Info™ 7 software in its 1.3.3 version (CDC, Atlanta). Descriptive analysis was carried out and $p < 0,05$ was considered statistically significant.

Results

Thirteen eyes of eleven patients with 2 bilateral cases were involved during this period of 6 years. The mean age was 29.54 years with extremes of 21-38 years and the sex ratio was 0.57. Any previous ocular health history was found among 69.23 % of patients while past history of pterygium surgery and ocular burn were respectively seen in 23.08% and 7.69% of cases. We observed a delay in reaching out the hospital center in 61.54 % of cases and blindness (visualacuitylowerthan 1/20) was found in 9 eyes where as 4 eyes were in visual impairment (visualacuity between 1/20 and 3/10). Corneal perforation was located in nasal sector in 61.54 % of cases, this perforation mean size was lower than the associated ulcer as seen in Table 1. Conjunctivectomy

combined with corneal sutures was performed in 76.92% of cases and amniotic graft in 23.08%. We noticed a improvement of visual acuity between one line and four lines after treatment in 53.85% of cases whereas the visual acuity was stationary in 38.46%of cases. After medical and surgical procedures 30.77% of patients presented legal blindness and the Table 2 shows final visual acuity. No complication was observed in 10 eyes while cataract and uveitis were respectively seen in two and one eye. 30.77% of cases experience drecurrence and the univariate analysis (Table 3) showed that the risks factors of recurrence were the female sex, a age under 30 and a delay of consultation more than 3 months. Amniotic graft (AG) was more reliable than corneal suture because when AG is not achieved, there is a great risk of recurrence.

Table 1 : Distribution of the eyes according to the circumferential extent of the ulceration and the perforation in degree.

Ulceration area (degree)	Perforation	Number	Percentage
(mean in degree)			
90	60	8	61.54
120	100	2	15.39
150	110	2	15.39
180	130	1	7.68
Total		13	100

Table 2 : Distribution of eyes according to final visual acuity.

Visual acuity	Number	Percentage
Blindness (<1/20)	4	30.77
Visual impairment ([1/20-3/10])	4	30.77
3/10	2	15.39
4/10	2	15.39
5/10	1	7.68
Total	13	100

Table 3 : Univariate analysis of risk factors for perforated ulcer recurrence.

	Recurrence		
	Yes	No	
Female gender (Age)	4	3	p=0,026
- <30 ans	4	2	p=0,046
- ≥30 ans	0	7	
Delay of consultation >3 mois	7	1	p=0,017
Bilaterality			
- Yes	1	1	p=0,514
- No	3	8	
Ulceration area ≥90%	3	5	p=0,94
Amnioticgraftachievement	3	6	p=0,016

Discussion

Mooren’sulcer is a rare affection with many disparities concerning epidemiology, clinical features, the rapeutic approaches and prognosis. These disparities refer mainly to the geographics areas of the world. In Northern Hemisphere

countries, this pathologyis not very frequent with a slow progression and barely reaches the perforation stage. On the contrary, it is most frequent in China, India and West Africa and it affects young people, is more aggressive with a rapid evolution towards perforation (Figure 1). The last disparity is about treatment, northern countries where MU is less severe

offer keratoplasty, the gold standard treatment not available yet in many countries in West Africa. The mean age of the study population was 29.54 years with extremes of 21 and 38 years. This is similar to Fasina's study mean age [4] in Nigeria (30.9 years), but is lower than Chen's study mean age [5] in China (48, 4 years). Thus, in light of these studies, the young age of people with Mooren's ulcer in Africa is «currently seen». In our study, some patients had a previous ocular health history of pterygium surgery and ocular burn. Many authors [4] report that trauma is a trigger for the onset of MU, no matter of the stage. Indeed, we believe that pterygium surgery could be a triggering factor in exposing the corneal antibodies during the excision of the pterygium head. These fragilized antibodies would be more vulnerable to aggression of the conjunctival antigens in the adjacent area to the excised pterygium. Thus the trauma by inducing an immunological conflict seems to be a promoting factor without being the cause of the ulcer since no antecedent was found in 69.23% of the patients.



Figure 1: Perforated Mooren's ulcer. Note the perforation of 3h to 5h in spite of a small ulcer.

In this study, we reported 69.23% cases of blindness secondary to MU at the perforation stage; this emphasized once again that severe stage of MU had a major impact on visual acuity. This high blindness rate could also be in relationship with the long delay of consultation mentioned above, which exceeds three months in the majority of cases (61.54% of the eyes) and the subsequent delay in treatment. The analysis of ulcer associated with corneal perforation shows that this perforation area is always smaller than the ulcer area. Perforation usually occurs in the central zone of the ulcer, which seems to be the first area where the ulcer would have begun before its perilimbal circumferential extension. This central area is early destroyed in depth at the origin of the perforation. This analysis of the extent reveals that this pathology in our tropics tends to go faster in depth than in periphery (Figure 1). Indeed no total peripheral involvement of 360 degrees was observed, unlike Srinivasan in India, which found 15% of total peripheral ulceration [6]. This rapid evolution in depth is very threatening for the prognosis because very often leading to evisceration. The treatment of MU is difficult and long. It sometimes requires the combination of several therapeutic approaches and must be adapted to the clinical response. At the stage of perforation, the treatment is

resolutely surgical associated with a medical therapy made of corticosteroids and / or immuno suppressive drugs.

The use of local and general corticosteroids is justified by the accessibility of these molecules in our regions and also by the recovery of some cases of less aggressive MU by means of cortico therapy which constitutes the first line treatment of the pathology. The broad use of immuno suppressants in the patient's management in our study is due to the aggressive nature of this disease in black people. Indeed this molecule has proved effectiveness in melano derms but its systemic use is at the origin of important side effects. Nevertheless, this immuno suppressive treatment in the context of a perforated ulcer must be systematically associated with a surgical treatment (Figure 2), the only one able to restore the anatomical integrity of the eye ball. In fact, the immuno suppressant or corticosteroids treatment brings healing of the ulcer but have no influence on the corneal perforation as well as hernia of iris. Conjunctival resection and its low cost are justifiable reasons which make this technique a therapeutic approach of choice in the treatment of MU [8]. Easy access of this surgical technique keeps promoting this procedure in the management of MU in Côte d'Ivoire. It has the advantage of being able to be carried out without an operating microscope. Moreover, this technique has a therapeutic advantage and allows a histological diagnosis of the resected conjunctiva.

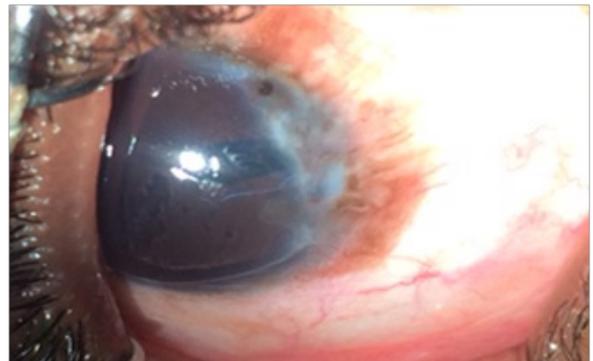


Figure 2 : Result of a perforated Mooren's ulcer treated by conjunctival desinsertion and corneal sutures. Note the sequential peripheral corneal opacity as well as a latero-deviated pupil on the perforation side.



Figure 3: Perforated Mooren's ulcer treated by amniotic membrane graft. Note the corneal perforation from 4h to 5h.

The corneal sutures after iris viable reintegration is often difficult to achieve in case of significant substance loss of the cornea. It is almost made under tension and is performed with separate simple corneo-limbal or corneo-scleral points. Only three cases (23.08%) in this study received human amniotic membrane graft (Figure 3). Amniotic membrane grafting is a recent therapeutic approach in the management of MU. This idea comes from the good results seen in treatment with amniotic membrane transplantation in corneal ulceration due to other causes. Its interest in the treatment of MU at the perforation stage is two fold: anatomical in order to compensate the loss of substance and by creating an immunosuppressive environment conducive to wound healing [9]. After medico-surgical treatment, a stationary acuity was noted in 38.46% of the cases. This stationary visual acuity could be judged un satisfactory after the treatment performed during this study. But the target of treating a perforated MU is the preservation of the anatomical integrity of the eyeball in order to avoid spontaneous evisceration. Complications may occur during the evolution of a perforated MU. We noted two cases of cataract and one case of uveitis in respectively 15.4% and 7.7% of cases. As for Chen [5] in China, he found these two complications at respective rates of 6.8% and 2.3%. Cataract is frequent during this pathology especially at the stage of perforation. The treatment of this cataract is difficult and must be done best by phacoemulsification or by the technique of «small incision cataract surgery». Indeed, Srinivasan [6] points out that the limbal incision in manual extracapsular cataract surgery would be a real risk factor for progression and even recurrence. The scleral incision would have the double advantage of moving the incision site away from the limbus, which is the inflammatory location of origin of this ulceration, and not to unmask the target corneal antigens.

Recurrences after healing are frequent during MU. We report 30.77% of recurrences in this study, this result is close to the results observed in Chen's study [5] in China which reported 25.6% of recurrences. This pathology is readily recurrent as described in the literature. This appears to be due to the fact that there are unknown etiologies of the MU and the most likely cause of this disease would be an immunological conflict. This recurrence is frequently encountered during these so-called «idiopathic» pathologies. The patient should be informed of the functional symptomatology of the recurrence in order to

consult as soon as possible. The analysis of the risks factors of recurrence of this pathology shows that any young melanoderm patient under 30 years of age, female and who would have a late consultation of more than 3 months would have more risk of recurrence of perforation secondary to MU. In these cases, amniotic graft would have better results in terms of visual function as well as a safety factor to avoid a possible recurrence.

Conclusion

MU at the stage of perforation is a rare but redoubtable pathology and seriously threatening for the organic and visual prognosis. It is more aggressive in melanoderm young people. The medical and surgical treatment has maintained the integrity of the eye ball. The functional prognosis due to the corneal opacities is far to be satisfactory. Keratoplasty could improve this functional prognosis in cases of perforated MU in black African people.

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