

Atypical Presentation of Acanthamoeba Keratitis- Case Report and Review of the Literature

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2. Key words

Acanthamoeba keratitis; Cornea;
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1. Abstract

The purpose is to present painless Acanthamoeba keratitis and to summarize actual literature data on this ocular infection. A 48-year-old women with a history of occasionally soft contact lens wear was referred to our hospital for management of left eye visual acuity impairment. Upon examination, the best-corrected visual acuity was 0,7 for her left eye. There was epitheliopathy resembling dendritic-like ulcer in the lower corneal hemisphere. Recurrent herpetic keratitis was diagnosed and treated, but with no improvement. Further observations revealed a dense, mid-peripheral ring-like stromal infiltrate. The patient did not report any pain. Corneal scraping was performed, and it was positive for Acanthamoeba trophozoites. She was treated with amoebicidal therapy consisting of propamidine isethionate 0.1% and polyhexamethylene biguanide 0.02%, neomycin. After three months, despite of amoebicidal therapy a stromal infiltrate failed to improve and the patient required a therapeutic penetrating keratoplasty. At the last follow-up visit, our patients' visual acuity was hand movement. The patient is waiting now for re-transplant combined with cataract surgery. She did not complain of any ocular pain in the entire course of her disease.

1.1. Conclusions: Acanthamoeba keratitis should be considered in differential diagnosis even in a lack of typical symptom such as pain, especially in a soft contact lens wearer.

3. Introduction

Acanthamoeba Keratitis (AK) is a severe and potentially vision-threatening corneal infection. The pathogen, Acanthamoeba, is a free-living protozoan that is distributed in various environments including air, soil, dust, and water. There are two stages in the life cycle of amoeba, a mobile trophozoite and a dormant cyst. The hallmark symptom of Acanthamoeba keratitis is exquisite pain, disproportionate to the clinical pictures. Other manifestations are often non-specific and may include: unilateral photophobia, tearing, redness. AK is often misdiagnosed as herpetic, bacterial, mycotic keratitis or corneal epithelial erosion (especially in the initial stage of the disease). It all makes that not only therapy but also diagnostic is a challenge. Although a couple of cases reports of AK exist in the literature, this is the first reported case of painless AK in Poland.

4. Case report

A 48-year-old myopic female presented unilateral keratitis with the deterioration of visual acuity in the left eye for a few weeks. According to the medical history, she was occasionally wearing contact lenses, five months ago treated with aciclovir due to herpetic keratitis of the left eye in the other medical center. Upon the exam-

ination, the Best Corrected Visual Acuity (BCVA) was 0.7 in the left eye and in the right eye it was 1.0. An ocular examination revealed benign conjunctival hyperaemia and minor epitheliopathy resembling dendritic-like ulcer in the lower corneal hemisphere of the left eye (Figure 1). Corneal sensation in the left eye was decreased. Recurrent herpetic keratitis was diagnosed, local and oral acyclovir treatment was initiated. Despite a slight improvement - no epitheliopathy in the lower corneal hemisphere, the stromal edema was observed, moreover the visual acuity of the left eye deteriorated to 0.3 (Figure 2). There was no abnormal finding in the left eye. The patient reported no ocular pain. During further observations, a dense, mid-peripheral ring-like stromal infiltrate developed (Figure 3). Visual acuity of the affected eye deteriorated to counting fingers. A deep corneal scrapings were harvested and inoculated into a dish of E. coli plated over non-nutrient agar. The culture was positive for Acanthamoeba spp. PCR for Acanthamoeba and confocal microscopy were performed. Intensive therapy with 0.1% propamidine isethionate, 0.2% polyhexamethylene biguanide, neomycin with fluconazole/ketoconazole (systemic) was initiated. After three months, despite of amoebicidal therapy a stromal infiltrate failed to improve and the patient required a therapeutic penetrating keratoplasty (Figure 4). The material was transferred to

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the eye pathology laboratory. The macroscopic histological examination of the host corneal tissue showed the specimen consisted of a haze corneal button measuring 8mm in diameter. In the microscopic examination: corneal epithelium was markedly attenuated and focally detached. A marked stromal edema was present together with focal infiltrations of neutrophils. There was a wide distribution of amoebic cysts and trophozoites within the stroma. Descemet membrane appears normal, but it was massively covered with inflammatory cells (mainly neutrophils). Only few endothelial cells were identified (Figure 5). To the last moment before the surgery, the patient did not report any pain. During the first follow up visit ocular examination revealed transparent corneal transplant secured with 16 Nylon 10/0 sutures (Figure 6). After three months, the transplant lost transparency, few sutures were loosen (Figure 7), moreover a cataract was diagnosed in the left eye. At the last follow-up visit, our patients' visual acuity was hand movement. The patient is waiting now for re-transplant combined with cataract surgery.

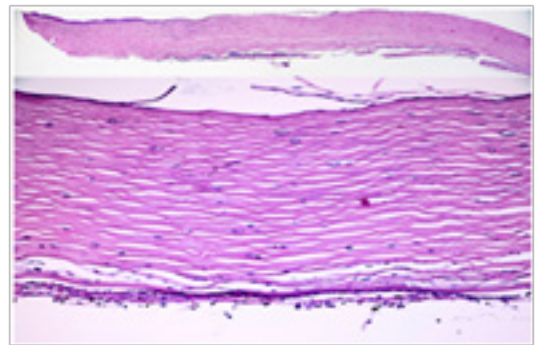


Figure 5: Ocular pathology examination of the patient's cornea demonstrating multiple Acanthamoeba cysts (arrows)

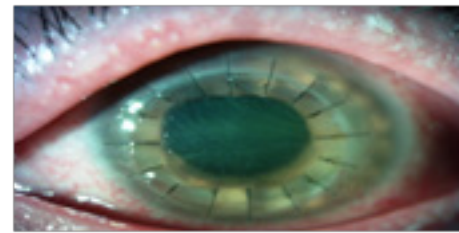


Figure 6: Clear corneal transplant



Figure 7: The transplant lost the transparency, cataract developed

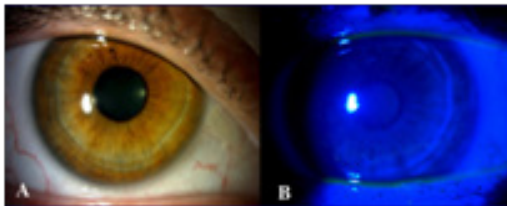


Figure 1: Slit lamp examination A conjunctival hyperaemia B with cobalt filter - minor epitheliopathy resembling dendritic-like ulcer in the lower corneal hemisphere

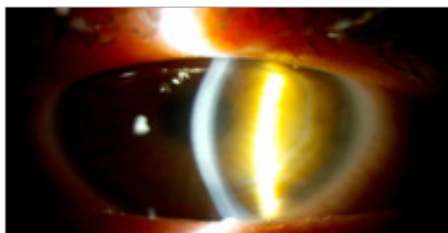


Figure 2: Slit lamp examination revealed stromal edema

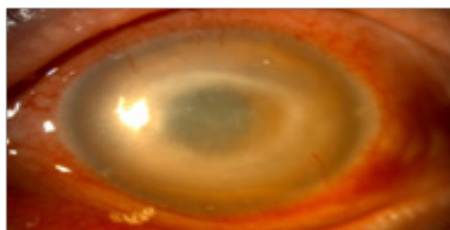


Figure 3: Slit lamp examination showed a dense, mid-peripheral ring-like stromal infiltrate

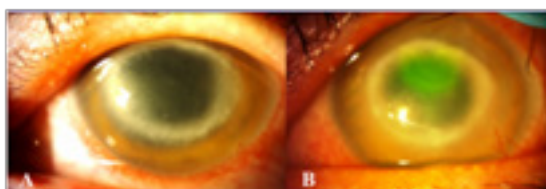


Figure 4: Slit lamp examination showed ring infiltration and extensive stromal infiltrate and epithelial defect

Table 1: Cases of painless AK existing in the literature

Reference	No of patients	Contact lens use	Previous infections or coinfections
Shukla ¹¹	1	+	Herpes simplex
	2	+	Non-specific ocular infection
	3	+	Bacterial
	4	+	previous history of herpes keratitis
	5	+	-
Kwok ⁵	6	+	-
Georgakopoulos ¹⁸	8	+	Herpes simplex
Tabin ⁷	8	-	Adenoviral conjunctivitis
	9	+	Herpes Simplex
Roters ¹⁹	10	+	Staphylococci, Enterococci
Elabjer ²⁰	11	+	Microbial keratitis
Stemberger ²¹	12	+	Herpes simplex
Claerhout ²²	13	No information	Viral/bacterial/ fungal
	14		
	15		
	16		
Perry ²³	17	+	Herpes simplex

5. Discussion

Acanthamoeba keratitis is a potentially vision-threatening infection of the cornea. The difficulty in diagnosis arise from the non-specific signs and symptoms in early stages, moreover this pathology may mimic other infections [1]. The German Acanthamoeba Keratitis Registry have revealed that in 47,6% herpetic, in 25,2 % mycotic,

and in 3,9% bacterial keratitis was misdiagnosed in acanthamoeba keratitis patients [2]. Moreover, AK may be present as a secondary or opportunistic infection in patients. We should keep in mind, that in about 23% of the cases, a mixed infection with virus, bacteria or fungi is present [3]. According to the other source, Acanthamoeba infections are polymicrobial, with 12,5% being coinfections with bacteria, 40% with fungi, and 5% triple infections. Moreover, coinfections established 55% of all Acanthamoeba infections [4]. Table 1 summarized cases of painless AK existing in the literature. Some studies have revealed the propensity for Acanthamoeba to coexist with other microbes, which increases their pathogenicity as well as survival, likewise hinders their treatment.

In the literature, the pain in Acanthamoeba keratitis is described as severe and disproportionate to the clinical findings during the slit lamp examination [5, 6]. Moreover, perineural annular infiltrates are also pathognomonic, but they are not always present and might have a late onset [7]. Other initial presenting symptoms are frequently nonspecific and may include: unilateral photophobia, tearing, redness [8]. Therefore, Acanthamoeba keratitis can be often misdiagnosed as herpes keratitis [9] or epithelial erosions during the early course of the disease resulting from the presence of an epithelial defect. However, a high clinical suspicion must remain for those patients who present with risk factors of Acanthamoeba keratitis in the absence of pain [6].

Our case demonstrates an atypical and unusual presentation of Acanthamoeba keratitis. The patient did not complain of any ocular pain in the entire course of her disease. She reported minor discomfort, but not pain. The mechanism for painless Acanthamoeba keratitis is not completely understood [10], but it is suggested to be a result of either perineuritis [11], pre-existing neurotrophic cornea (such as herpes keratitis), or pretreatment with topical steroids, which mask clinical signs of this keratitis [12, 13]. The differences in the immune response and virulence might affect variable clinical presentation. Our patient was diagnosed as having herpes simplex keratitis five months before presentation, and this situation may have resulted in decreased corneal sensation. These circumstances can explain a painless course of the disease that caused considerable delay in the proper diagnosis and treatment in our patient. Also, it led to avoidable loss of visual acuity before transplant and also finally transplant failure.

The corneal graft was performed in our patient due to the lack of the treatment's response and worsening of the local condition. According to the literature, therapeutic penetrating keratoplasty might be considered when the infection escalates to the paracentral corneal stroma despite maximum anti-amoebic therapy [14]. The rate of corneal transplantation for controlling the acanthamoeba keratitis ranged from 5 to 68% [15]. The surgical treatment should be delayed until the eye is uninflamed and after completion of anti-amoebic treatment due to the poor prognosis and high risk of

graft failure [16]. Post-operative complications after keratoplasty, except rejection of the graft, include: recurrence of Acanthamoeba infection, other infection, glaucoma, cataract, wound leak, and irregular astigmatism. Roozbahani observed 75% graft failures, 50% cataract, 17% uncontrolled glaucoma and 8% reactivation of acanthamoeba in his study group of therapeutic penetrating keratoplasty for acanthamoeba keratitis [17]. Unfortunately, the graft failure and cataract development were observed in our patient, therefore she is waiting for combine surgery.

In conclusion, Acanthamoeba keratitis must be considered in the differential diagnosis of keratitis even without the classic signs of severe pain, especially in contact lenses wearer and patients who have had herpetic keratitis. Incorrect diagnosis and treatment may further complicate the clinical picture and delay the proper treatment. Moreover, it significantly reduces the quality of patient's life due to poor visual acuity.

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