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Case report
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Paediatric nevus comedonicus syndrome with band keratopathy and ipsilateral cataract

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Abstract

Background: Band keratopathy is characterized by a horizontal band of grey opacification on the surface of the cornea. Nevus comedonicus is a rare epidermal nevus that has been reported with ipsilateral cataract and corneal changes.
Case report: A 14-year-old boy presented with bilateral blurred vision with recurrent red eye. He had a history of skin dryness and scarring on his face. On examination, greyish-white opacifications were observed in both corneas. We initially diagnosed the patient with juvenile idiopathic arthritis, but since the systemic condition was unrelated to the condition, we consulted a dermatologist. It turns out that the eye condition was related to nevus comedonicus syndrome. The patient then underwent corneal chelation using ethylenediaminetetraacetic acid (EDTA). Following the procedure, visual acuity in the left eye improved from 0.05 to 0.125, while visual acuity in the right eye, which had lens opacification, remained hand movement.
Conclusion: Chemical chelation using EDTA in band keratopathy patients with nevus comedonicus syndrome is useful for enhancing corneal clarity.

Keywords: band keratopathy, corneal chelation, ethylenediaminetetraacetic acid, nevus comedonicus syndrome

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Sindrom nevus comedonicus pediatrik dengan keratopati jalur dan katarak ipsilateral

Abstrak

Latar belakang: Keratopati jalur dicirikan sebagai jalur mendatar opasifikasi kelabu pada permukaan kornea. Nevus comedonicus adalah nevus epidermal jarang berlaku yang telah dilaporkan bersama katarak ipsilateral dan perubahan pada kornea.
Laporan kes: Seorang budak lelaki berusia 14 tahun hadir dengan penglihatan kabur bilateral dan mata merah berulang. Beliau mempunyai sejarah kekeringan kulit dan parut pada wajahnya. Pada pemeriksaan, opasifikasi putih-kelabu dilihat pada kedua-dua kornea. Pada mulanya kami mendiagnosis pesakit dengan artritis idiopatik juvenil, tetapi oleh kerana keadaan sistemik tiada kaitan dengan keadaan ini, kami merujuk pesakit kepada pakar dermatologi. Ternyata keadaan mata ini adalah berkaitan dengan sindrom nevus comedonicus. Pesakit kemudiannya menjalani pengkelatan kornea menggunakan asid *ethylenediaminetetraaceti* (EDTA). Selepas prosedur tersebut, akuiti penglihatan pada mata kiri bertambah baik dari 0.05 ke 0.125, manakala akuiti penglihatan pada mata kanan, yang mengalami opasifikasi pada kanta, tidak berubah pada gerakan tangan.
Kesimpulan: Pengkelatan kimia menggunakan EDTA dalam kalangan pesakit keratopati jalur dengan sindrom nevus comedonicus adalah berguna untuk mempertingkatkan kejelasan kornea.

Kata kunci: keratopati jalur, pengkelatan kornea, asid ethylenediaminetetraacetic, sindrom nevus comedonicus

Introduction

The development of grey opacification of the superficial cornea and Bowman's sign are 2 symptoms of band keratopathy, a chronic, degenerative corneal disorder. In the early stages of the disease, this condition is asymptomatic; however, as the condition worsens, the patient experiences discomfort and a decline in vision as a result of disruption of the corneal epithelium.[1,2] Numerous therapy strategies, including mechanical debridement, ethylenediaminetetraacetic acid (EDTA) chelation, and excimer laser phototherapeutic keratectomy, have been used to control band keratopathy.[3,4] Nevus comedonicus is a rare kind of epidermal nevus that manifests clinically as clusters of dilated follicular apertures with keratotic plugs, which resemble comedones.[5] This case report aims to provide a band keratopathy case and treatment for a child with nevus comedonicus syndrome.

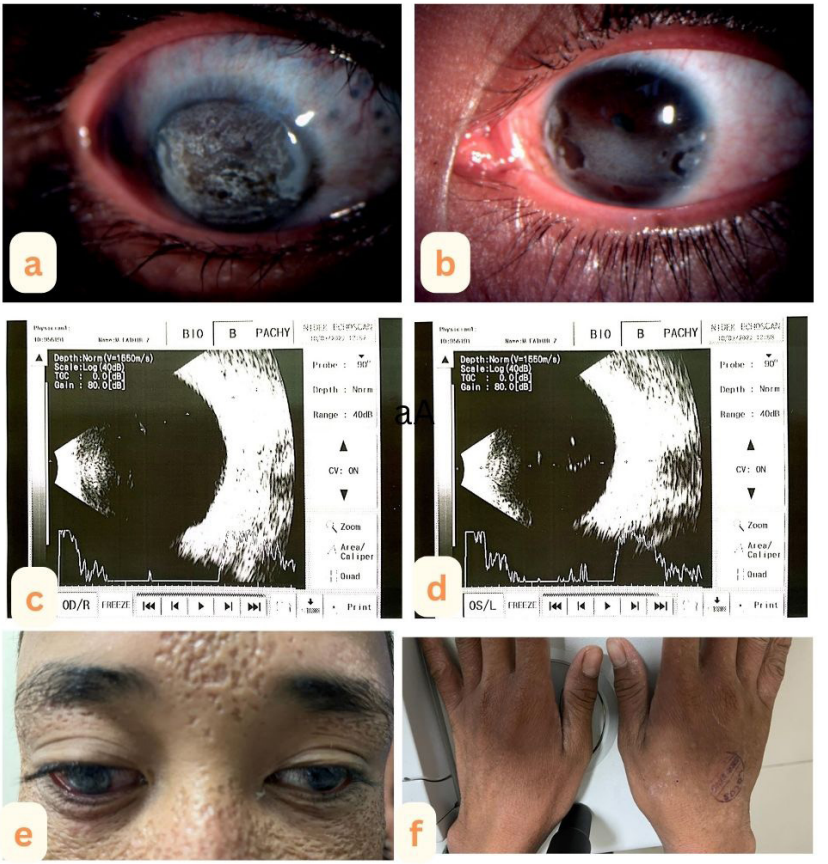

Fig. 1. Corneal surface of the patient with *(a)* total keratopathy in the right eye and *(b)* band keratopathy in the left eye. Ultrasound examination of the *(c)* right eye and *(d)* left eye showing mild opacity. The scar on the patient's *(e)* skin and *(f)* hands do not show any bone deformity.

Case report

A 14-year-old boy presented with bilateral ocular redness. During the examination, the patient stated that he had experienced blurred vision for the last 5 years, which was worsening. The complaint was not associated with ocular pain, watering, photophobia, or foreign body sensation. The patient had a history of dry skin and facial scars. There was no history of joint pain, bone deformity, or joint swelling. A family history with similar signs and symptoms was also denied. Physical

examination showed vital signs were within normal limits and cicatrix was found in the glabellar, malar, zygomatic, and nasal areas. No deformity was seen in the patient's extremities. Other general examinations were within normal limits.

An ophthalmologic examination revealed normal eye position and full eye movement in all directions. Visual acuity in the right eye was hand movement and 0.05 in the left eye. Intraocular pressure was normal by palpation in both eyes. Slit-lamp examination of the right eye showed cicatricial upper eyelid and corneal keratopathy. Other structures, such as the anterior chamber, pupil, iris, and lens, were difficult to assess due to the presence of corneal keratopathy (Fig. 1a). Slit lamp examination of the left eye revealed cicatricial effusion of the upper eyelid, Efron grade 2 conjunctival hyperaemia, low tear level, corneal band keratopathy, moderate anterior chamber depth with flare and cells difficult to assess, irregular pupil, iris synechiae, and lens opacity (Fig. 1b).

Ultrasound showed inflammatory cells and vitreous fibrosis bilaterally (Figure 1c-d). The patient was diagnosed with bilateral band keratopathy and complicated cataract in the left eye. He was prescribed artificial tear therapy and vitamin A palmitate bilaterally. The patient was then referred to the Rheumatology department to rule out juvenile idiopathic arthritis (JIA) and Dermato-Venereology to determine if there were any skin abnormalities related to his ocular condition (Fig. 1e-f).

Examinations in the Rheumatology section did not show any abnormalities related to JIA or other autoimmune disorders. The consultation at the Dermato-Venereology section revealed a relationship between the condition of the patient's cornea and lens and his skin disorder, which was diagnosed as nevus comedonicus.

The patient was advised to undergo optical penetrating keratoplasty with lens extraction and implantation of an intraocular lens for both eyes, but because there was no corneal donor available, the patient was scheduled to undergo bilateral chemical chelation with EDTA. In the procedure, opacification was seen due to grade 3 keratopathy in the right eye (the pupillary edge was difficult to see) in all quadrants, while the left eye had grade 3 keratopathy in the interpalpebral area (Fig. 2a-b). After a keratectomy with the use of EDTA, the right and left anterior chambers showed no active inflammation, but there was posterior attachment of the iris and cloudiness in the anterior capsule of the lens bilaterally.

The patient was treated with levofloxacin 6 times a day bilaterally, artificial tear drops 6 times a day bilaterally, sodium hyaluronate eye drops 4 times a day bilaterally, and oral mefenamic acid 500 mg 3 times a day. A bandage contact lens was used in both eyes. One day after surgery, the patient complained of blurred vision with minimal pain. Visual acuity was hand movement in the right eye and 0.125 in the left eye. Slit-lamp examination of both anterior segments revealed Efron grade 3 conjunctival injection, bandage contact lens attached to the cornea, moderate anterior chamber without flare and cells, irregular pupils with posterior iris synechiae, and cloudy lens. Figure 2c displays documentation of the left eye one day following surgery. Whereas there is no documentation for the right eye

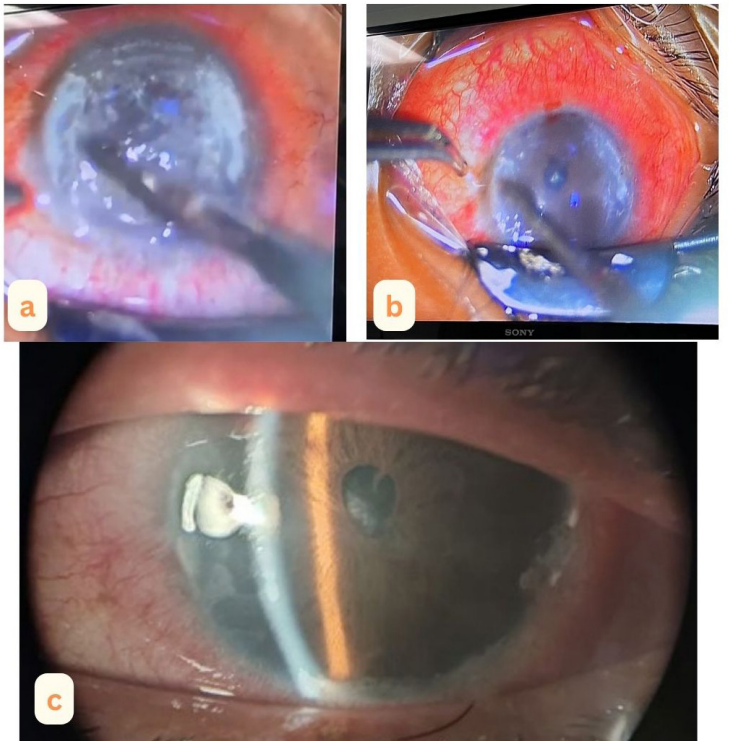

Fig. 2. EDTA procedure performed on the patient's *(a)* right and *(b)* left eye, as well as the condition of the left eye 1 day after the procedure.

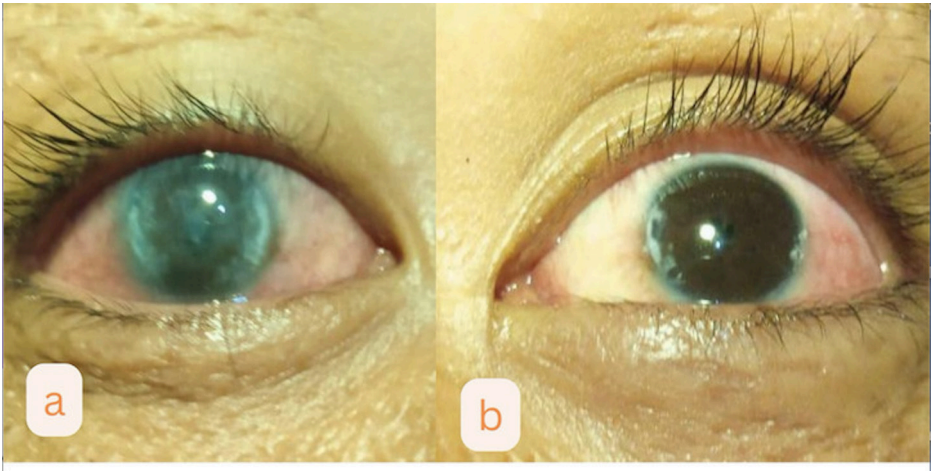

Fig. 3. The patient's ocular surface in the *(a)* right and *(b)* left eye 4 months after surgery. The central cornea appears clearer.

The patient returned for follow-up 1 week after surgery. There was a decrease in conjunctival injection. We advised the patient to follow up for an evaluation after 2 weeks. The management plan for the patient involved lens extraction in the left eye, followed by the right eye. Due to the right eye's level 2 clarity, we recommended a penetrating keratoplasty. Four months later, the patient reported no pain, no redness, and improved vision (Fig. 3).

Discussion

Band keratopathy is a chronic, degenerative corneal condition characterised by the appearance of grey opacity on the superficial cornea. Calcium deposits in the subepithelium, Bowman's layer, and anterior stroma usually occur in the interpalpebral region and give a "Swiss cheese" appearance due to the scattered holes in the cornea.[1] Bands of keratopathy begin at the peripheral centre along the horizontal meridian and slowly progress centripetally to involve the visual axis. Progression of band keratopathy is usually slow (months to years), but there are cases that progress rapidly (weeks to months).[6]

Band keratopathy can occur independently of inflammatory or systemic disease, but it is more commonly secondary to chronic inflammation, systemic or hereditary disease associated with calcium metabolism abnormalities, or topical or intraocular drugs that cause local disturbances in calcium metabolism in ocular tissues. The mechanism of calcium deposition in the cornea is not known with certainty, but it is associated with exposure to the cornea. This can be the result of precipitation remaining as the tears evaporate or because of the lower pH in this area.[1,2,7]

The most common cause of band keratopathy in children is JIA uveitis, with clinical manifestations of joint pain, swelling, warmth, effusion, and limited joint movement. A retrospective study of 125 patients with JIA-associated uveitis in Milan, Italy, showed that the most common ocular complications were posterior synechiae, cataracts, band keratopathy, glaucoma, and macular oedema.[8]

The patient was a 14-year-old boy with clinical features of band keratopathy, so we initially suspected JIA. However, signs and symptoms suggestive of JIA were not found in the patient, so JIA was ruled out. The diagnosis of band keratopathy is based on clinical history and slit-lamp examination. Given the broad aetiology of band keratopathy, it is important to perform a thorough history and serological examination to identify the underlying aetiology. Signs and symptoms of band keratopathy include decreased visual acuity, foreign body sensation, watering, photophobia, and sometimes ocular redness.

Slit-lamp examination will reveal white-grey plaques with fine deposits in a horizontal band distribution from 9 to 3 hours with lucent holes that do not involve the periphery of the cornea. The first-line treatment option for band keratopathy

is medical management of the underlying disease, especially in asymptomatic or mildly symptomatic patients.[1,3]

Medical therapy includes artificial tear drops, gels, and eye ointments that can help with irritation, watery eyes, foreign body sensation, and photophobia. Various surgical options are available to remove keratopathic bands, including mechanical debridement with a blade, chemical chelation, and phototherapeutic keratectomy. The main goals of treatment are to remove the opacity caused by calcium deposits and restore a flat ocular surface. In most cases, vision can be improved. However, in eyes with poor visual potential, this procedure is performed primarily to improve ocular comfort. Manual debridement with blade scraping is effective in removing keratopathic bands but may cause the corneal surface to become irregular. Chelation with EDTA works by absorbing calcium and softening deposits. The current most popular method for treating band keratopathy is chemical chelation using EDTA.[1,2,4,7]

Nevus comedonicus lesions are generally present at birth in approximately 50% of patients, and most patients develop lesions before the age of 10 years, without racial or sexual preference. Lesions are most commonly found on the face, neck, upper arms, chest, and abdomen. The diagnosis of nevus comedonicus is based on clinical examination and histopathology.[5,9]

Conclusion

Band keratopathy is a common degenerative corneal disease that can be either idiopathic or associated with underlying systemic and ocular conditions. The most common treatment for band keratopathy is chemical chelation using EDTA. Chelation has proven to be a simple, effective, and safe treatment. Band keratopathy in this case was part of the nevus comedonicus syndrome, which is due to a rare skin disorder.

Declarations

Informed consent for publication
The patient and the patient's parent provided informed consent for the publication of this case report.

Competing interests
None to declare

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None to declare

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References

1. Das AV, Pillutla LN, Chaurasia S. Clinical profile and demographic distribution of band shaped keratopathy in India: A study of 8801 patients. Indian Journal of Ophthalmology. 2022 May;70(5):1582-5. http://doi.org/10.4103/ijo.ijo_2541_21
2. Mannis MJ, Holland EJ. Cornea. 5th Edition. Section 3. Corneal Dystrophies, Ectatic Disorders, and Degenerations. Amsterdam: Elsevier; 2016; p.865-8.
3. Al-Hity A, Ramaesh K, Lockington D. EDTA chelation for symptomatic band keratopathy: results and recurrence. Eye. 2018 Jan;32(1):26-31. https://doi.org/10.1038/eye.2017.264
4. Jhanji V, Rapuano CJ, Vajpayee RB. Corneal calcific band keratopathy. Current Opinion in Ophthalmology. 2011 Jul;22(4):283-9. https://doi.org/10.1097/ICU.0b013e3283477d36
5. Chauhan P, Meena D, Sharma S. Dermoscopy of nevus comedonicus: A case series. Indian J Paediatr Dermatol. 2020;21(3):232. https://doi.org/10.4103/ijpd.IJPD_29_20
6. Najjar DM, Cohen EJ, Rapuano CJ, Laibson PR. EDTA chelation for calcific band keratopathy: results and long-term follow-up. Am J Ophthalmol. 2004 Jun;137(6):1056-64. https://doi.org/10.1016/j.ajo.2004.01.036
7. Singh P, Tripathy K. Keratopathy. 2023 Aug 25. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2025 Jan. Available from: https://pubmed.ncbi.nlm.nih.gov/32965824/
8. Marelli L, Romano M, Pontikaki I, Gattinara MV, Nucci P, Cimaz R, Miserocchi E. Long Term Experience in Patients With JIA-Associated Uveitis in a Large Referral Center. Front Pediatr. 2021 Jun 28;9:682327. https://doi.org/10.3389/fped.2021.682327
9. Timbó RPF, Gomes MK, Falcão EMM, Ramos-E-Silva M. Nevus Comedonicus: Case Report. Case Rep Dermatol. 2017 Oct 4;9(3):184-189. https://doi.org/10.1159/000480722