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Keratoconus: The Masquerade

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1. Abstract

We describe the case of an 11-year-old girl with a background of anisometropia and corneal scarring who had a misleading appearance of keratoconus. The patient initially presented at age 6 with a unilateral reduction of visual acuity (VA) in her left eye (6/15). There was also evidence of left corneal scarring which appeared chronic; possibly secondary to previous herpes simplex keratitis (HSK). She was treated with glasses and though her vision initially stabilized (VA 6/6 in both eyes), she was lost to follow-up due to multiple missed appointments. Her vision remained stable for 5 years before she presented with another reduction of VA in her left eye (6/48).

The patient was investigated using Pentacam which showed ectatic changes suggestive of keratoconus. There was also progression of the corneal scar in her left eye. With an impression of active stromal herpetic keratitis, the patient was commenced on 200 mg Acyclovir TDS and Dexamethasone minims TDS. 2 months later, her VA improved to 6/7.5 in the left eye. Pentacam was then repeated and the astigmatism, as well as anterior steepening values had reduced. Considering this, clearly HSK had led to a misleading appearance of keratoconus upon initial imaging.

This case highlights the potential false positive diagnosis of keratoconus that can occur in younger patients with HSK. Although pentacam results showed advanced ectasia, one should remain mindful of differential diagnoses; particularly seeking out any signs of herpetic eye disease and keeping a low threshold for appropriate treatment.

2. Keywords: Keratoconus; Pentacam; Herpes Simplex Keratitis; Paediatric; Topography factors

3. Introduction

First described in 1854, keratoconus is a progressive, ectatic corneal disorder which is defined by apical corneal thinning, corneal steepening and central corneal scarring [1-3]. The corneal thinning causes conical protrusion, myopia and irregular astigmatism leading to an impairment in quality of vision; often having a significant impact on the patient's quality of life [1]. It is a relatively common condition with largely unknown aetiology [4]; though associations have been made with corneal trauma mechanisms [5,6].

Corneal topography is one of the most widely used tools for diagnosing keratoconus and monitoring disease progression. In particular, recent advances from slit-scanning topography (Orbscan) to Schleimplug camera-based (Oculus Pentacam) can more accurately distinguish normal corneas from clinically keratoconic corneas [7,8]. We describe the rare case of a paediatric patient who had a misleading appearance of keratoconus on Pentacam imaging.

4. Case Presentation

A 6-year-old girl with a background of anisometropia and

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central corneal scarring presented to a tertiary care centre with unilateral reduction of visual acuity (VA) in her left eye (6/15). On examination, there was evidence of left corneal scarring which appeared chronic with no acute changes. A diagnosis of potential herpes simplex keratitis (HSK) was made based on the clinical appearances. She was treated with glasses and though her vision initially stabilised (VA 6/6 in both eyes), she was lost to follow-up due to multiple missed appointments.

Her vision remained stable for 5 years before she presented with another reduction of VA in her left eye (6/48). On examination, she had left central corneal stromal scarring with deep stromal vessels. There was no evidence of anterior chamber inflammation or corneal ulcers; however, the cornea did appear to have some stromal inflammation. Based on this information, a diagnosis of stromal herpetic keratitis with associated corneal thinning was made.

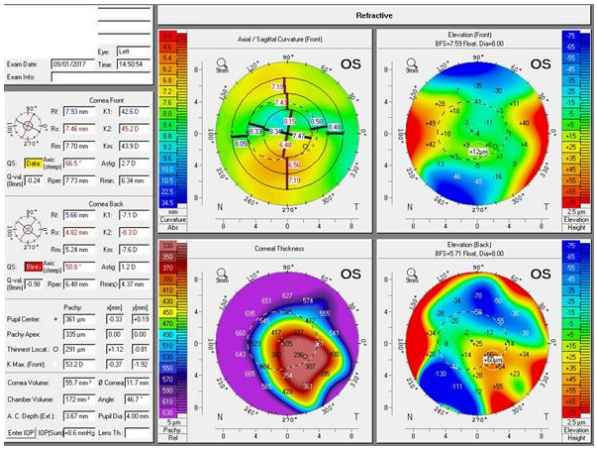

Figure 1: Central thickness maps show the position of a 'cone' in the inferotemporal areas giving a misleading appearance of keratoconus.

5. Investigations

The patient was investigated using Pentacam HR imaging which showed ectatic changes suggestive of keratoconus as well as a positive Belin ABCD score, highlighted in figure 3. Progression of the corneal scar in her left eye was also noted. It was unclear from the Pentacam images whether this was due to underlying keratoconus or whether the herpetic keratitis had induced such changes.

Furthermore, the right eye showed no evidence of keratoconic changes which supported our suspicion of herpetic keratitis with associated scarring and thinning in the left eye.

6. Outcome and Follow-up

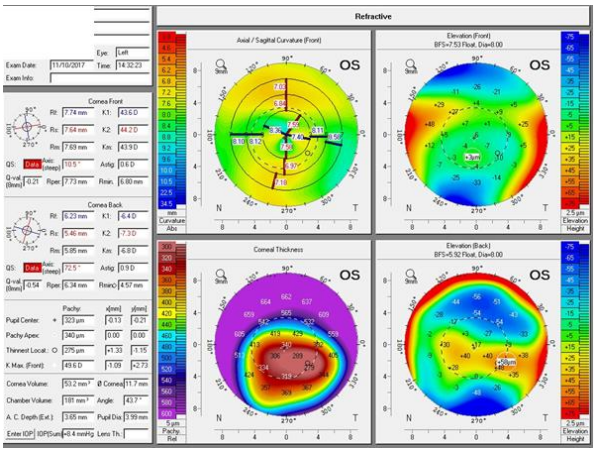

Figure 2: Repeat pentacam images 2 months after the first follow-up clinic.

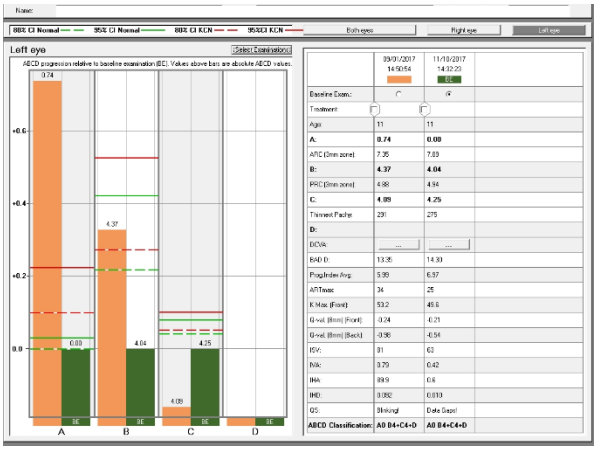

Figure 3: A comparison of Belin Ambrosio ABCD scores between the 2 pentacam images. **A:** anterior surface radius of curvature in 3mm zone; **B:** posterior surface radius of curvature in 3mm zone; **C:** corneal thickness at the thinnest point; **D:** distance corrected visual acuity.

With an impression of left active stromal herpetic keratitis, the patient was commenced on 200 mg acyclovir TDS and Dexamethasone minims TDS with a view to closely monitor the cornea via imaging. In the follow-up clinic 2 months later, the patient was comfortable and her visual acuity had stabilized to 6/7.5 in the left eye. Pentacam was then repeated and the appearances now correlated more closely with clinical findings. There were still residual changes showing where the scar and thinning was, but with lower values for both astigmatism and anterior steepening. These findings, as well as clinical improvement, made the diagnosis of HSK flare-up more likely as opposed to keratoconus. The Belin-Ambrosio scores had also

decreased, but still suggested the possibility of keratoconus (or some form of ectasia) on the readings. The only evidence still supportive of keratoconus was the posterior elevation around the scar site and the inferotemporal displacement evident on the pachymetry map.

7. Discussion

This case highlights the potential false positive diagnosis of keratoconus that can occur in younger patients with HSK. The most likely explanation for this patient's initial Pentacam changes was the HSK scar causing the cornea to form an ectatic appearance similar to that of keratoconus. Two months later, the improved pentacam changes correlated with the HSK flare-up resolving with medical treatment. It is also possible for the scar to have had a flattening effect on the cornea, thereby causing a change in the ectatic appearance predominantly on the posterior surface as natural healing occurs in the absence of an underlying progressive ectasia. However, this explanation is far less likely in keratoconus and has only been described once before in literature [9]. As keratoconus is a progressive ectasia, there would be an expectation of progression rather than improvement in the values especially at such an age. In paediatric keratoconus it has been well established that when it is diagnosed earlier, there is a far greater risk of rapid progression. The improvement in vision with HSK treatment in just 2 months without any cross-linking process further supports this diagnosis [10].

With the introduction of the Belin-Ambrosio Enhanced Ectasia Display, we are now able to compare corneal elevation data with a commonly used 'enhanced reference surface' [11,12]. The Belin ABCD system is used in conjunction with this in order to grade the stages of keratoconus. Our patient had a positive Belin ABCD score which contributed to the masquerade of keratoconus. One must bear in mind that keratoconus is a form of ectasia and any type of ectasia can create the changes suggested by the Belin-Ambrosio score. In this case, the ectatic changes flattened anteriorly once the infective process became quiescent.

Although Pentacam results alone were strongly suggestive for keratoconus in this patient, it is also important to keep patient demographics and clinical findings in mind. A 2018 study by Roshdy et al. states that there is an increased risk of elevation index fallices in patients that are below 21 years of age [13]. This supports our hypothesis that younger patients are at a higher risk of misdiagnosis. To prevent this, Roshdy et al. suggests that elevation indices with altered normative data or pachymetry based indices should be used in patient's outside of the age range of 21-40 [13].

In such a case, if cross linking had been requested urgently based on the Pentacam changes alone, it could have been potentially devastating for the patient causing reactivation of the herpetic infection, melting and perforation leading to visual loss [14]. It is therefore imperative that patients reviewed in keratoconus clinics are not seen solely on a virtual basis. There must be a full clinical assessment at least on the first visit by the consultant team. In the case of paediatric or complex cases (such as in Down's Syndrome), follow-up in consultant clinic rather than virtual clinic is recommended. Indeed. there are examples of HSK developing or becoming activated after corneal cross linking suggesting a subclinical quiescent form which may become activated by UVA light [15]. Kymionis [16] has similarly stated concerns about the use of CXL in HSK and the risk of HSK reactivation when no clear history was evident.

It is tempting to begin prompt corneal cross linking (CXL) in such patients to provide better outcomes but one must be clear that there is progressive ectasia before starting treatment. In children, defining when is a suitable time is unclear as there is a risk of rapid progression which may lead to the opportunity for corneal cross linking to be missed.

Unfortunately, the definition of ectasia progression remains unclear according to the 2015 Global Consensus on Keratoconus and Ectatic Diseases [17]. The panel defined progression by a consistent change in 2 or more of the following parameters: thinning and/or thinning or changes in the pachymetric rate of change, elevation of

the anterior corneal surface and/or elevation of the posterior corneal surface. The panel also agreed that further quantitative data to define progression is needed [17]. In children, this change can occur quicker, so from the basis of this case report, there must be close follow-up where diagnosis is unclear, as rapidly progressive keratoconus can also result in apical scarring and not all cases will show clear evidence of herpetic disease as this case did. This patient may also be at risk of developing progressive ectasia from the previous episodes of herpetic infection, therefore long-term follow-up with corneal topography is also recommended in such cases analysing both eyes.

8. Learning Points

- HSK scarring can cause a misleading appearance of keratoconus on Pentacam imaging
- Although Pentacam is a useful aid, it must be used in conjunction with clinical findings in order to make an accurate diagnosis.
- Clinicians must be cautious when diagnosing keratoconus in a patient with a history of HSK; particularly if the patient is young.
- One should establish a definite progression of ectasia before CXL takes place.

HSK is a contraindication for corneal cross linking.

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