Vernal Keratoconjunctivitis and Severe Keratoconus Associated with Autoimmune Polyglandular Syndrome Type II (APS-2); A Case Report

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Abstract

Vernal keratoconjunctivitis (VKC) is a bilateral, usually seasonally recurrent inflammation of the conjunctiva. Even though it is an uncommon type of allergic eye disease but in some cases it can cause severe damage to the ocular surface, leading to keratoconus (KC) and vision loss. We report a rare case of vernal keratoconjunctivitis and keratoconus diagnosed since childhood and then was diagnosed years later with Addison's disease and subclinical hypothyroidism. Even though the association between VKC and autoimmune diseases is still not clear but this case highlights the importance of having a low threshold to screen patients with VKC for autoimmune diseases based on their clinical presentation.

Keywords: Addison's disease; hypothyroidism; keratoconus; autoimmune polyglandular syndrome, type II; vernal keratoconjuctivitis.

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Case report:

A 19-year-old female patient with history of VKCand bilateral keratoconussince childhood presented toendocrinology clinic at age of 18 with complaints of generalized fatigue, unintentional weight loss and increased skin pigmentation at buccal area and palmer skin creases for about 4 years duration. Physical examination showed the hyperpigmentation at the buccal mucosa and palmer skin creases, with Beau's Grooves in her finger nails. Prior to this presentation, she was seen per many primary care physicians for the similar above complaints who failed to screen her for adrenal insufficiency. Laboratory work confirmed the presence of very low serum cortisol and high ACTH levels consistent with primary adrenal insufficiency, and she was started on hydrocortisone replacement. In addition, her laboratory tests showed mildly high TSH, normal T4 and low 25-OH-vitamin Dconsistent with subclinical hypothyroidism and vitamin D deficiency respectively and she was started on low dose levothyroxine and vitamin D replacement. Her serum screening for celiac disease was negative. She reports feeling much better since

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was started on the above hormone replacement therapy. She had palpebral and bulbar VKC with secondary mild bilateral mechanical ptosis. In addition to that, she had severe keratoconusin her left eye and advanced keratoconus in her right eye with corneal scarring. Recently, she underwent a successful right corneal transplant surgery. In regards to family history, her grandmother (paternal side) has history of primary hypothyroidism; otherwise her family history is not significant for VKC, KC, or Addison's disease.

Discussion

Ocular allergy is a common disease in daily practice. VKC could be idiopathic or associated with underlying etiologies including but not limited to immune disease, endocrine, genetic, neurogenic, and environmental[1,2]. Autoimmune Polyglandular Syndrome type II (APS-2) is the most frequent Autoimmune Polyglandular Syndrome.

The defining component of APS-2 is Addison’s disease, which is found in conjunction with either autoimmune thyroid disease or type1A diabetes mellitus. A group of non-endocrine autoimmune disorders, such as vitiligo, myasthenia gravis, thrombocytopenic purpura, Sjögren's syndrome, rheumatoid arthritis, and primary antiphospholipid syndrome can occur occasionally as part of APS-2[3]. Keratoconus is a corneal ectasia characterized by a central and paracentral thinning leading to visual impairment[4], it is not uncommon complication of VKC. The association between keratoconus and autoimmune diseases has been suggested per previous retrospective studies and few case reports[5,6,7]. Nemet AY and his colleagues have done a large retrospective observational case-control study to evaluate the association between keratoconus and immune disorders[5]. The study included 426 patients with variable immuned is orders except for Addison's disease; It showed a strong association between keratoconus and autoimmune conditions that may point to the role of the immune system in the pathogenesis of KC. Ocular manifestations in Addison's disease are rare and may include ptosis, blepharitis, blepharospasm, keratoconjunctivitis, corneal ulcers, episcleritis, cataract, and papilledema[8]. In 1962, GASS JD, known as father of macular diseases[9], had done a review of literature of 12 cases of idiopathic hypoparathyroidism with keratoconjuctivitis, in which seven of these cases had a definite adrenal insufficiency, the age range of these seven patients were in between 6 years to 30 years old[10]. Here, we present a rare case of VKC and severe KC diagnosed since childhood and was later diagnosed with APS-2 in her late teens. Up to our knowledge, this is the first case report of a case of APS-II associated with VKC.

The questions to be raised based on this case and the above literature review are: Can we add VKC as a part of the non-endocrine disorders that can be associated with APS-2? Do we need to routinely screen patients with VKC for autoimmune disorders including thyroid dysfunction and Addison's disease?

Conclusion

It is not at all clear whether the APS-2 was in any way related to the ocular disease. The association of VKC and KC is very well known. Their relation to APS-2 is tenuous.

Awaiting further data about the possible association between VKC and autoimmune

conditions; we suggest having a lower threshold to screen patients with VKC for autoimmune diseases based on their clinical presentation.

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حالة نادرة لمرض الرمد الربيعي والقرنية المخروطية الشديدة مع متلازمة أمراض الغدد المناعية المتعددة النوع الثاني

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الملخص

يعرف الرمد الربيعي أنه إلتهاب تحسسي يصيب القرنية والملتحمة في كلتا العينين بشكل موسمي متكرر. ويعد هذا المرض نوعاً نادراً من أمراض حساسية العين إلا أنه من الممكن في بعض الحالات الشديدة أن يتسبب بتلف سطح العين بالإضافة إلى حدوث القرنية المخروطية والعمى.

وقد قمنا بتسجيل ونشر حالة نادرة لمريض مصاب بالرمد الربيعي منذ الطفولة ومن ثم تم تشخيصه بعد عدة سنوات بمرض أديسون (قصور الغدة الكظرية) وقصور وظائف الغدة الدرقية.

وتشير هذه الحالة إلى ضرورة الانتباه لمرضى الرمد الربيعي وفحصهم مبكرا كلا حسب الأعراض والمؤشرات السريرية للكشف عن احتمالية وجود أمراض مناعية مثل قور الغدة الكظرية والغدة الدرقية.

الكلمات الدالة: مرض أديسون، الرمد الربيعي، القرنية المخروطية، الغدد المناعية المتعددة النوع الثاني.