Case report

ANTERIOR NODULAR SCLERITIS ASSOCIATED WITH VOGT-KOYANAGI-HARADA DISEASE: A CASE REPORT

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Abstract

Purpose To report a young patient who initially manifested with bilateral anterior nodular scleritis and subsequently developed ocular features typical of Vogt-Koyanagi-Harada (VKH) disease.

Case description A 9-year-old girl was diagnosed with bilateral anterior nodular scleritis, which improved with topical administration of corticosteroids and systemic nonsteroidal anti-inflammatory drugs. Four months later, after the treatment was withdrawn, she developed a recurrence of scleritis together with sunset glow fundus and Dalen-Fuchs nodules in both eyes.


Keywords: anterior nodular scleritis, Vogt-Koyanagi-Harada

Vogt-Koyanagi-Harada (VKH) syndrome is a systemic disease, probably of autoimmune origin, and is characterized by various ocular, cutaneous and neurologic abnormalities. The most common ocular features include chronic bilateral granulomatous panuveitis in association with serous retinal detachment, development of sunset glow phenomenon and Dalen-Fuchs nodules. Systemic manifestations include vitiligo, poliosis, alopecia, signs of meningeal irritation, and auditory disturbances. The disease usually affects heavily pigmented Asian people in their third to fourth decades of life, with a slight predominance in women.(1) Scleritis is typically a painful inflammatory process, regularly associated with diverse systemic diseases such as rheumatoid...
arthritis, systemic lupus erythematosus and Wegener’s granulomatosis. Both disorders, scleritis and VKH, occur rarely in childhood. We describe a 9-year-old girl who presented with anterior scleritis and later developed features typical for VKH.

**Case Report**

A 9-year-old girl complained of decreased vision, ocular pain and redness of her left eye. The clinical diagnosis was anterior nodular scleritis. At that time, retinal examination was without abnormalities. Treatment with steroid and antibiotic drops (1% prednisolone acetate 4 times a day and ciprofloxacin 4 times a day) had insufficient effect and the patient was referred. Her visual acuity was 20/20 OD and 20/30 OS and bilateral anterior nodular scleritis was observed (Figure 1). The anterior chamber showed cells, and retinal examination was normal. She had no previous history of systemic diseases or use of medications. No systemic complaints or signs were present. Laboratory investigations, including complete blood count, sedimentation rate, kidney and liver function test, rheumatoid factor, anti-nuclear antibody and radiological chest x-ray, were all within normal limits. HLA B-27 was negative. She underwent a computerized topography (CT) scan of the orbits, which revealed focal thickening, and enhancing sclera at the superomedial portion of the left eye, without adjacent organ invasion (Figure 2). The treatment with topical corticosteroids (1% prednisolone acetate 4 times a day) was continued, with added nonsteroidal anti-inflammatory drugs (Mefenamic acid; 500 mg per day) giving good effect on the scleritis.

Three months later, the medication was withdrawn and 4 weeks thereafter, the patient experienced a recurrence of ocular pain. Visual acuities were 20/20 in both eyes. Mild recurrence of bilateral nodular scleritis was noted as well as aqueous and vitreous cells. At that time, fundus examination revealed bilateral “sunset glow fundus” with Dalen-Fuchs nodules in the peripheral retina, and macular hyperpigmentations (Figure 3). A CT of the chest was normal; specifically, no signs of sarcoidosis or tuberculosis were noted. The audiometry was normal in both eyes.

![Figure 1](image1.png)

**Figure 1.** 1A: Sectorial dilatation of episcleral vessels in the right eye. 1B: Nodular shape with dilation of the episcleral vessels in the superomedial portion of the left eye.
Anterior nodular scleritis associated with VKH

Figure 2. CT orbit showing focal thickening and enhancing sclera at the superomedial portion of the left eye without adjacent organ invasion

Figure 3. Retinal findings in both eyes 4 months after the onset of scleritis revealing bilateral “sunset glow appearances”, with Dalen-Fuchs nodules in the peripheral retina, and granular macular hyperpigmentations.

Discussion

Vogt-Koyanagi-Harada syndrome has been rarely reported in children. To the best of our knowledge the youngest reported patient in the literature with VKH syndrome was a 3-year-old child with coexisting type-1 diabetes mellitus and celiac disease. Our 9-year-old female patient exhibited retinal findings of “sunset glow appearance” together with Dalen-Fuchs nodules, but systemic features of VKH were absent, according to revised diagnostic criteria for probable VKH disease.
Scleritis can be associated with systemic connective tissue, including rheumatoid arthritis, Wegener granulomatosis, systemic lupus erythematosus and sarcoidosis. Anterior scleritis has been rarely described in children. Sporadic cases of scleritis in children were previously reported, e.g. necrotizing scleritis due to varicella zoster infection in a six-year-old boy(6) and posterior scleritis in a 7-month-old infant.(7) Scleritis in VKH is rare. There were occasional cases of posterior scleritis with exudative retinal detachment, which has been proposed as an initial sign of VKH disease.(8) However, to the best of our knowledge so far an association between VKH disease and anterior scleritis has not been reported. Our patient had two unusual clinical features: anterior nodular scleritis and sunset glow fundus associated with small atrophic retinal lesions suggestive of Dalen-Fuchs nodules at a young age.

The differential diagnosis of our patient included sarcoidosis, which is a disease of unknown etiology and characterized by multisystemic granulomatous inflammation. The most common findings in posterior segment disease were vitritis (69%) and choroidal “punched-out” lesions (56%).(9) Although sarcoidosis rarely causes nodular scleritis, there are reported cases of sarcoidosis diagnosed by a biopsy of sclera nodules.(10) We did not perform a sclera biopsy in our patient. Sarcoidosis is relatively rare in childhood.(11) A child with sarcoidosis usually suffers from extrapulmonary involvement and/or parenchymal lung disease.(11) However, CT chest scans, and laboratory and clinical evaluations show no evidence of pulmonary involvement or other signs compatible with the diagnosis of sarcoidosis.

In conclusion, we report an otherwise healthy child with unusual clinical presentation, in which bilateral anterior nodular scleritis preceded the development of retinal findings typical of VKH disease.

References

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รายงานผู้ป่วย: VOGT-KOYANAGI-HARADA ที่มีการอักเสบของเปลือกลูกตาส่วนหน้า

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บทคัดย่อ

วัตถุประสงค์ เพื่อรายงานผู้ป่วยเด็กหญิงที่มีอาการแสดงเริ่มแรกเป็นการอักเสบของเปลือกลูกตาส่วนหน้าของตาทั้งสองข้าง โดยในเวลาต่อมามีอาการแสดงทางตาของโรค Vogt-Koyanagi-Harada รายงานผู้ป่วย เด็กหญิงอายุ 9 ปีมีอาการและการแสดงของการอักเสบของเปลือกลูกตาส่วนหน้าในตาทั้งสองข้างที่มีการสีสันสีน้ำตาลแดงที่มีลักษณะของ sunset glow และ Dalen-Fuchs nodules ที่ตาทั้งสองข้าง เขียนในวารสาร 2552;48(1),24-29.

สรุป โรค Vogt-Koyanagi-Harada อาจมีความสัมพันธ์กับการอักเสบของเปลือกลูกตาส่วนหน้า

คำสำคัญ: anterior nodular scleritis, Vogt-Koyanagi-Harada