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■ Case Report

Subconjunctival hemorrhage in Henoch-Schonlein purpura: a case report and literature review

Henoch-Schonlein purpurada subkonjonktival hemoraji: olgu sunumu ve literatür taraması

Yaprak Akbulut ^{1*} , Nesrin Büyüktortop ¹ , Cihat Şanlı ² , Erhan Yumuşak ¹ , Tevfik Oğurel ¹ 

¹ Kırıkkale University, School of Medicine, Department of Ophthalmology, Kırıkkale, Turkey

² Kırıkkale University, School of Medicine, Department of Pediatrics, Kırıkkale, Turkey

* Corresponding author: Yaprak Akbulut E-mail: yprk_akblt@hotmail.com ORCID: 0000-0002-7774-2681

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ABSTRACT

We report a 5-year-old boy who presented with diffuse abdominal pain, non-thrombocytopenic purpuric skin rash, and bilateral multiple foci of subconjunctival hemorrhage in the inferior bulbar conjunctiva. The patient was diagnosed clinically with Henoch-Schonlein purpura (HSP), according to EULAR/PRINTO/PRES criteria. HSP is a leukocytoclastic small vessel vasculitis. Ocular involvement is very rare. Episcleritis, keratitis, uveitis, cystoid macular edema and cotton wool spots, central retinal artery occlusion, anterior ischemic optic neuropathy and subperiosteal hematoma were reported previously. Here, we present a case of bilateral subconjunctival hemorrhage in a child with HSP for the first time in the English literature.

Keywords: Henoch-Schonlein purpura, ocular, subconjunctival hemorrhage, vasculitis

ÖZ

Bu yazıda diffüz abdominal ağrı, trombositopenik olmayan purpurik deri döküntüsü ve inferior bulbar konjunktivada bilateral multipl subkonjonktival hemoraji ile başvuran 5 yaşında bir erkek hasta sunuldu. Hastaya EULAR / PRINTO / PRES kriterlerine göre klinik olarak Henoch-Schonlein purpurası (HSP) ile tanısı konuldu. HSP bir lökositoklastik küçük damar vaskülitidir. Oküler tutulum çok nadirdir. Episklerit, keratit, üveit, kistoid maküla ödemi ve atılmış pamuk görünümü, santral retinal arter tıkanıklığı, ön iskemik optik nöropati ve subperiosteal hematoma daha önce bildirilmiştir. Burada, İngilizce literatürde ilk kez HSP'li bir çocukta bilateral subkonjonktival hemoraji olgusunu sunuyoruz.

Anahtar kelimeler: Henoch-Schonlein purpura, oküler, subkonjonktival hemoraji, vaskülit

INTRODUCTION

Henoch-Schonlein purpura (HSP) is an acute immunoglobulin (Ig)-A-mediated systemic leukocytoclastic vasculitis characterized by the involvement of small vessels of the skin, gastrointestinal system, kidneys, joints, and rarely the lungs and central nervous system. It is the most common type of vasculitis of childhood.¹ Diagnosis is based on the presence of non-thrombocytopenic palpable purpura (mandatory criteria), and one or more of the following: diffuse abdominal pain, histopathologic proof of leukocytoclastic vasculitis or proliferative glomerulonephritis with predominant IgA deposits, arthritis or arthralgia, and findings of renal involvement.² Genetic, environmental, and antigenic factors play a role in the etiology of HSP. Most patients report a preceding respiratory tract infection. It is thought that infectious agents may trigger a dysregulated immune response that results in inflammation and vasculitis [2]. Prognosis is generally good

Table 1. Literature review of patients with Henoch-Schonlein purpura and ocular involvement

Reference	Age, year/ sex	Ocular involvement
Ryder, 1976	6/M	Transient conjugate eye deviation and cortical blindness
Lorentz, 1980	14/F	Episcleritis
Yamabe, 1988	63/M	Anterior uveitis, keratitis
Ma'luf, 2002	5/M	Bilateral subperiosteal orbital hematoma
Wu, 2002	6/F	Bilateral central retinal artery occlusion
Chuah, 2005	54/M	Anterior ischemic optic neuropathy
Muqit, 2005	42/M	Keratitis and bilateral granulomatous anterior uveitis
Erer, 2010	39/M	Uveitis
Ergul, 2010 *	12/F	Unilateral generalized subconjunctival hemorrhage
Kaur, 2012	11/M	Bilateral uveitis
Nicholson 2013	24/M	Cystoid macular edema, retinal hemorrhages and cotton wool spots
Patheja, 2015	21/M	Cystoid macular edema, retinal hemorrhages and cotton wool spots
Present case	5/M	Bilateral multiple foci of subconjunctival hemorrhage

F: female, M: male, *: Not indexed in pubmed or embase



Figure 1. Purpuric skin rash mainly on the lower extremities

and symptomatic treatment is usually sufficient. Corticosteroids may be given to decrease the severity of abdominal pain and arthralgia. Immunomodulatory agents are used with severe nephritis [1].

Ocular involvement in HSP is very rare (**Table 1**). In this report, we present an unusual case of HSP associated with bilateral multiple foci of subconjunctival hemorrhage (SCH).

CASE REPORT

A 5-year-old boy was admitted to the Department of Pediatric Emergency, with diffuse abdominal pain and was suspected of having acute abdomen. The parents reported a history of recent pharyngeal infection. On physical examination, there was a purpuric skin rash mainly on the gluteal region and the lower extremities (**Figure 1**). Intestinal sounds were normoactive. There was no rebound or guarding on the abdominal examination or nausea or vomiting. Extremities and joints were normal with no swelling or tenderness. There was subconjunctival hemorrhage on both eyes. Other physical examination findings were normal. Blood pressure was 110/70 mmHg (%92 percentile). Complete blood count, urine analysis, erythrocyte sedimentation rate, and the bleeding profile were within normal limits. Ultrasonography revealed that the appendix was normal in shape and contour. The fecal occult test was positive. Serum complement C3, C4, perinuclear anti-neutrophil cytoplasmic antibodies (P-ANCA), cytoplasmic antineutrophil cytoplasmic antibodies (C-ANCA), and Ig A levels were within normal limits. Our patient was clinically diagnosed as having HSP according to the European League against Rheumatism/Paediatric



Figure 2. Multiple foci of SCH in the inferior bulbar conjunctiva of left eye



Figure 3. Multiple foci of SCH in the inferior bulbar conjunctiva of right eye



Figure 4. Multiple foci of SCH cumulating around the end of visible conjunctival vessels

Rheumatology International Trials Organisation/Paediatric Rheumatology European Society (EULAR/PRINTO/PRES) criteria due to the presence of the typical non-thrombocytopenic purpuric skin rash and gastrointestinal involvement. The patient was referred to ophthalmology because of the unusual SCH. Visual acuity was 20/20 in both eyes. Intraocular pressure was 11/11 mmHg. Periocular area and eyelids were normal in color and shape. There was no discharge, edema, papillary or follicular reaction. There were multiple foci of SCH in the inferior bulbar conjunctiva of both eyes (**Figure 2** and **3**). There were no other abnormalities in the anterior or posterior segment examinations. Intravenous 1mg/kg/day prednisone was administered due to severe abdominal pain, which was relieved on the third day of treatment. The skin rash resolved within a month and the SCHs resolved within 2 weeks. A year later, the patient had a recurrence of purple skin rash on the dorsal side of the feet and the distal part of the legs, again with a history of pharyngeal infection. However, there was no abdominal pain or arthralgia or ocular involvement. Laboratory tests were normal. There was no proteinuria or occult blood in the feces. The patient was followed up for 3 years with no further recurrence. The informed consent was received from the family.

DISCUSSION

HSP is a form of leukocytoclastic vasculitis in which small superficial vessels of the skin are primarily affected. Immunofluorescence studies showed that IgA, C3, and fibrin were deposited within the walls of involved skin vessels, and in the endothelial and mesangial cells of the kidney [1].

HSP rarely involves the eye. A literature review of ocular manifestations in HSP is summarized in **Table 1**. The most encountered ocular involvement was uveitis (n=4) [3-6], keratitis accompanied in two of these cases [3,4]. There are 2 cases of bilateral cystoid macular edema and cotton wool spots associated with decompensated HSP-related nephropathy [7,8]. The other possible vasculitis-related ocular manifestations in HSP were central retinal artery occlusion, subperiosteal hematoma, episcleritis and anterior ischemic optic neuropathy [9-13].

In HSP, the vessel walls are infiltrated by neutrophil granulocytes, and eventually become necrotic. The extravasation of erythrocytes through necrotic vessel walls to the surrounding dermis causes the purpuric skin rash [1]. In our case, we suggest that the conjunctival vascular endothelial cells may also have been affected by the immune complex-mediated immune response. Similar to the skin involvement, the extravasation of erythrocytes through damaged conjunctival vessels might have caused the appearance of SCH. SCH is an acute and non-painful hemorrhage between the conjunctiva and episclera in the absence of discharge and inflammation. It may vary from dot-blot hemorrhages to extensive areas of bleeding. Trauma, systemic vascular disease, sudden severe venous congestion, blood dyscrasias, anticoagulants, acute hemorrhagic conjunctivitis, and conjunctival tumours among others may cause SCH [15]. In our case, there was no history of trauma, hypertension, diabetes, thrombocytopenia, or any sign of ocular inflammation. Also, the presentation of SCH was atypical because it was not in a

single circumscribed localization, instead there were multiple foci of SCH cumulating around the end of visible conjunctival vessels (**Figure 4**).

The lack of risk factors, the bilateral and atypical presentation of hemorrhage, and the concurrent HSP symptoms suggest that SCH might be associated with HSP. However, a conjunctiva biopsy could not be obtained to prove vasculitic involvement. Systemic vasculitides have certain ocular manifestations. SCH is a known manifestation of Kawasaki disease, but it is not common among other pediatric vasculitides [15]. To our knowledge, there is no previous report of SCH associated with HSP in the English literature. HSP may cause severe ocular problems, especially in the presence of nephropathy. It would be better to consider an ophthalmologic consultation in the evaluation of patients with HSP. One of the accompanying ocular findings may be SCH. Accordingly, it would be better to keep HSP in mind in the differential diagnosis of SCH in childhood.

LITERATURE SEARCH

PubMed and EMBASE were searched without date restriction in August 2018 for English-language results using the following terms: Henoch-Schonlein purpura, Henoch-Schonlein's vasculitis, IgA vasculitis, ocular, ophthalmological, eye.

DECLARATION OF CONFLICT OF INTEREST

The authors declare no conflict of interest. There is no financial assistance.

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