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Case Report

# Spontaneous hyphema in juvenile idiopathic arthritis uveitis

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## ABSTRACT

**Background.** Juvenile idiopathic arthritis (JIA) is a rheumatic disease that may be associated with ocular involvement in childhood. Classical findings of JIA uveitis are cells and flare; hyphema, bleeding in the anterior chamber of the eye, is a rare finding.

**Case.** An 8-year-old girl presented with 3+ cells and a flare in the anterior chamber. Topical corticosteroids were started. A follow-up examination 2 days later revealed hyphema in the affected eye. There was no history of trauma or drug use, and the laboratory test results did not suggest any hematological disease. Systemic evaluation resulted in the diagnosis of JIA by the rheumatology department. The findings regressed with systemic and topical treatment.

**Conclusions.** The most common cause of hyphema in childhood is trauma, but it can rarely be seen with anterior uveitis. This case highlights the importance of recognizing JIA-related uveitis in the differential diagnosis of hyphema in childhood.

**Key words:** hyphema, Juvenile idiopathic arthritis, uveitis.

Juvenile idiopathic arthritis (JIA) is the most common rheumatic disease in childhood. Among the extra-articular complaints of JIA, uveitis is a frequent finding. JIA-related uveitis often presents as silent chronic anterior uveitis in oligoarticular or rheumatoid factor (RF) (-) polyarticular forms.[1] This type of uveitis is generally asymptomatic and can be detected on slit-lamp examination but can lead to complications such as cataracts, glaucoma, band keratopathy and cystoid macular edema which can cause visual impairment and blindness.[2] Hyphema, which is bleeding in the anterior chamber of the eye, is often caused by trauma in childhood. Iris anomalies, inflammatory/infectious factors, post-surgical conditions and some systemic diseases should also be considered in the etiology.[3,4] There are very few reports of hyphema related to uveitis and it is an even less common condition in JIA uveitis.[3]

This paper presents a case of unilateral spontaneous hyphema with JIA, highlighting the need to consider JIA-related uveitis in the differential diagnosis of spontaneous hyphema in children.

## Case Report

An eight-year-old girl presented with decreased vision in her left eye for the last two days. On ophthalmic examination, her best corrected visual acuity was 20/20 in the right eye and counting fingers from 3 meters in the left eye. Anterior segment examination of the right eye was within normal limits, but in the left eye the conjunctiva was slightly hyperemic and 3+ cells and flare were observed. Intraocular pressure

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was measured as 12 mmHg in the right eye and 14 mmHg in the left eye. A dilated examination revealed normal bilateral fundi. The history taken from the parents revealed that she had experienced swelling and pain attacks in her left ankle, left heel and left wrist lasting 4-5 days in the previous year and uveitis attacks twice but did not receive any systemic treatment. Topical dexamethasone 0.1% hourly and cyclopentolate 1% drops 3 times per day were prescribed for anterior uveitis. At the follow-up examination two days later, there was no regression in inflammatory activity and approximately 1 mm hyphema was observed in the left anterior chamber (Fig. 1). Neither rubeosis iridis nor any iris anomalies were detected. Trauma history and drug use such as aspirin were excluded.

A consultation to the Department of Pediatric Rheumatology confirmed that she had arthritis at the age of seven and had been treated for arthritis. She was diagnosed with oligoarticular JIA. Further evaluation of complete blood count, peripheral blood smear and coagulation parameters were within normal limits. Chest X-ray and PPD tests were reported to be normal. Brucella, Salmonella typhi and paratyphi and HLA B5/ B51/ B27 antigen tests were negative, RF value, anti-nuclear antibody and anti- ds DNA antibody test were within normal limits.

The Pediatric Rheumatology department prescribed subcutaneous methotrexate (MTX) 10 mg weekly with oral prednisolone 0.5 mg/kg per day. Two weeks later, visual acuity increased to 20/40 in the left eye, reaction decreased to 1+ cells, hyphema disappeared and iris pigments became evident on the lens. By the end of the first month, visual acuity improved to 20/20 in the left eye. Oral corticosteroids were tapered over a period of two months. Two years later, MTX had to be discontinued due to abnormal liver function tests. As the patient experienced recurrent uveitis and arthritis, adalimumab was initiated. Despite systemic treatment, the patient had 1-2 anterior uveitis attacks per year during the four-year follow-up period, there was no recurrence of hyphema in any of the other attacks.

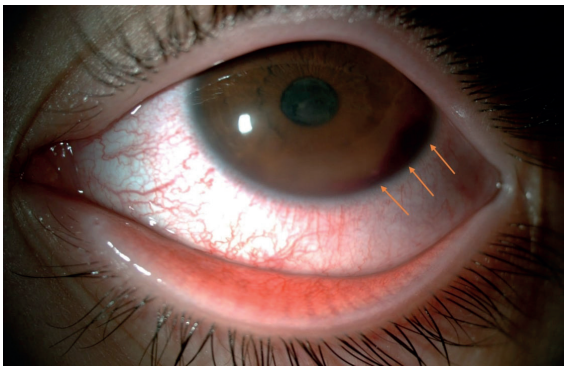

**Fig. 1.** Hyphema in the anterior chamber

The family was informed about the case presentation and written consent was obtained.

## Discussion

In this report, we present a female patient with JIA uveitis and unilateral spontaneous hyphema. Trauma is the most important cause of hyphema in childhood.[5] Post-traumatic hyphema pathophysiology can be classified as rupture of vessels in the iris and/or ciliary body after stress caused by antero-posterior equatorial expansion of the globe in blunt trauma, deterioration of fragile vascular structures in the pupillary sphincter or angle due to a sudden increase in intraocular pressure, and direct damage to vessels and hypotonia in lacerating traumas.[6] Spontaneous hyphema may be seen in hematological diseases with impaired blood parameters and a tendency to bleed, such as sickle cell anemia, leukemia, haemophilia and immune thrombocytopenia in childhood.[7,8] Furthermore, the use of anti-coagulants[9] or non-steroidal anti-inflammatory drugs (NSAID) (anti-platelet activity)[3,10], may contribute to hyphema formation. In addition, infiltrating iris anomalies such as histiocytosis, retinoblastoma or juvenile xanthogranulomatosis involving the iris and ciliary body may cause hyphema.[7]

Hyphema due to non-traumatic reasons such as uveitis is a rare condition in children. Duke- Elder and Perkins[11] described petechial

hemorrhage and hemorrhagic iritis in severe inflammation of uveitis and explained hyphema in uveitis with 3 mechanisms; vessel damage secondary to vasculitis, leakage from sensitive rubeotic vessels, and increased diapedesis of inflammatory cells.[11] Rubeosis iridis is usually a sign of proliferative diabetic retinopathy, retinal vein occlusion, or anterior segment ischemia and appears as small, fine, disorganized vessels on the anterior surface of the iris. During anterior segment inflammation, prominent dilated iris vessels can be observed and can be distinguished from rubeosis iridis with stromal and radial location. It is thought that hyphema may occur in severe inflammation from prominent dilated and damaged vascular structure, increased capillary permeability due to the breakdown in the blood-aqueous barrier then subsequent diapedesis and leakage from vessels.[12] Fong and Raizman[3] reported five different cases with hyphema accompanying anterior uveitis. The diagnoses were Reiter's syndrome, juvenile chronic arthritis, ankylosing spondylitis (AS), idiopathic anterior uveitis, and herpes simplex. Rubeotic vessel formation was detected in 3 of 5 cases, one case was taking NSAID, and no suspicious condition was reported in the remaining case. In the current patient, no neovascularization or a prominent vascular structure were detected on the iris, and there was no history of systemic drug use.

There is only one previous case report of spontaneous hyphema related to JIA in the literature, reported by Shimada et al.[4] This was a 5-year-old girl with bilateral hyphema as a result of anterior uveitis. Her arthritis, ocular findings and her age suggested this disease. If rheumatological signs and uveitis are seen together in childhood, JIA, Behçet's disease and juvenile sarcoidosis/Blau syndrome should be considered first.[1] In uveitis due to JIA, an inflammatory reaction is seen in the anterior chamber, but the development of hyphema is rare. Increased permeability and diapedesis after inflammation may be the cause of hyphema. In the current case, the reason for spontaneous hyphema was not clearly identified. Spontaneous hyphema has also been reported in one case of syphilitic uveitis.[13]

In childhood, after excluding trauma and bleeding diatheses, it should be kept in mind that hyphema may occur in other conditions including JIA-related uveitis without a prominent iris vascular structure.

## Ethical approval

The family was informed about the case presentation and consent form was obtained.

## Author contribution

The authors confirm contribution to the paper as follows: study conception and design: FBA, SK; data collection: FBA; analysis and interpretation of results: FBA, SK; draft manuscript preparation: FBA, SK, SÖ. All authors reviewed the results and approved the final version of the manuscript.

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## Conflict of interest

The authors declare that there is no conflict of interest.

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