Comments on: Relapse of acute lymphoblastic leukemia presenting as masquerade uveitis with hypopyon in a child

Dear Editor,

We read the interesting paper by Rehman *et al*.[1] on "relapse of acute lymphoblastic leukemia presenting as masquerade uveitis with hypopyon in a child" published in the Indian Journal of Ophthalmology. The authors presented a 5-year-old girl with acute lymphoblastic leukemia (ALL) who had decreased visual acuity (20/200) in the affected left eye for the past 5 days. Intraocular pressure was increased and the young girl had anterior chamber precipitation, which the authors called "hypopyon." The article of the authors is most timely for the differential diagnosis of "hypopyon" and "pseudohypopyon," particularly for a prompt diagnosis of neoplastic masquerading syndromes. However, we have a few important observations on the ocular findings of the child and suggestions to make it an interesting read for the benefit of all pediatricians and ophthalmologists interested in ocular complications of ALL because [Fig. 1] the presented pediatric case gives objective initial clues for performing a true definition of the anterior chamber sediment and differential diagnosis.

First, although diffuse superficial conjunctival hyperemia is clearly seen when we evaluate the presented [Fig. 1a] of the article, we do not agree with the authors that the child had ciliary congestion. Etiologically speaking, hypopyon is a layered meniscus formation in the anterior chamber of the eye resulting from acute intraocular inflammation that classically presents with severe ciliary hyperemic injection or congestion, a violaceous hue, conjunctival, episcleral, and deep scleral vasodilatations.[2] The term "warm hypopyon" is used for such a "hot disease." If we carefully look at [Fig. 1a] again, the perilimbal area of the involved left eye of the child is not dusky red. In contrast, the circumcorneal area is rather white. In addition, according to the presented case, the child did not complain of ocular pain, lacrimation, or photophobia, all of which are cardinal symptoms of acute uveitis.

Second, the shape of the non-infectious uveitic collections is characteristic, which results in sediment formation typically heaped up centrally rather than at its edges. However, the anterior chamber meniscus of [Fig. 1a] is heaped up at its edges, especially on its temporal border. Such macroscopic signs to the naked eye (macro-) should have forced the authors to make the diagnosis of "pseudohypopyon" from tumoral etiologies. Therefore, the term "hypopyon," which was stated throughout the text, does not seem to be correct. Strictly speaking, the appropriate terminology "tumoral pseudohypopyon" should be preferred for such a precipitate that indicates "masquerade syndromes of malignancy." Indeed, these anterior chamber collections contain neoplastic cells, such as the authors also find in their unique case upon an aqueous tab, which revealed the presence of atypical lymphoblast cells.[1] However, it should be kept in mind that repetitive head shaking or chronicity of the disease may consequently result in 1+ to 3+ secondary inflammatory (uveitic) reactions due to neoplastic cell irritation to the uveal tissues.

Third, the differentiation of "pseudohypopyon" from "hypopyon" is of utmost importance for pediatricians and ophthalmologists because their managements and prognoses are completely different.[3] Indeed, the sediment's behavior can be an indicator of some specific etiologies. As a result, my next question to the authors is that did they evaluate the mobility of the precipitation? In other words, we wonder whether the anterior chamber meniscus was shifting or not upon posture change? It has been demonstrated that the etiology of anterior chamber collection affects the mobility of the precipitation at various speeds.[3] Actually, a postural change of a patient may cause the dispersion of the sediment if it contains a lower concentration of fibrinous exudate (such as in tumoral pseudohypopyon), which totally dislocates into another part in the anterior chamber *within seconds* upon head-tilting to one side. Such mobile sediment in the direction of leaning is called "shifting pseudohypopyon," which is a pathognomonic feature of "tumoral pseudohypopyon." Thus, if the authors had changed the position of the child to one side, they would possibly have encountered the shifting feature of the collection. On the other hand, uveitic sediments have "non-shifting" properties, which is completely immobile that can be called as "plastic hypopyon" as a result of fibrinous exudates. However, uveitis in ocular Behçet disease also causes shifting hypopyon; but, the collection dislocates slowly *within 5–10 min* upon head-tilting, not *within seconds* as seen in "neoplastic pseudohypopyon."

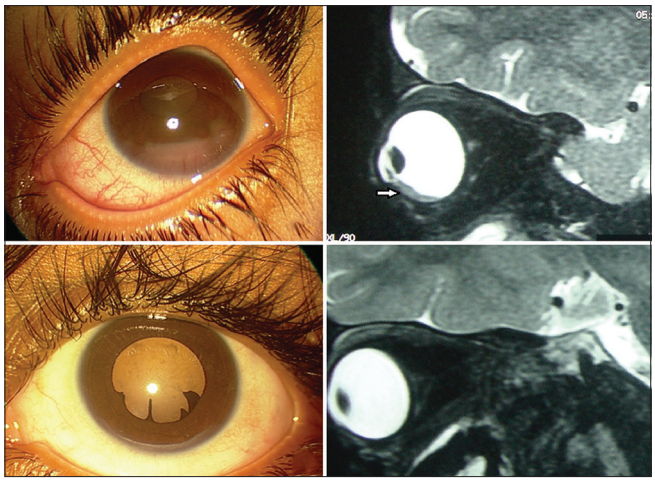

Figure 1: Slit lamp photograph shows blood-tinged, pinkish, 2-mm cold "macro-pseudohypopyon" in an uninjected white eye without ciliary congestion

Finally, the color of the sediment and the appearance of the circumcorneal area may direct clinicians to a particular etiology.[4] Rehman *et al*.[1] stated in their case report that the anterior segment showed a 2 mm whitecolored hypopyon. If we carefully look at the [Fig. 1] again, the sediment is not white. On the contrary, the "blood-tinged" pinkish nature of the collection can clearly be realized, which again indicates spontaneous hyphemia due to ALL or lymphoma.[5] Such anterior chamber precipitation should be defined as "blood-streaked pinkish-colored pseudohypopyon," not white-colored hypopyon. Indeed, an "uninjected white perilimbal area" should not be expected in a "hot disease," namely uveitis. As stated above, however, 1+ to 3+ anterior chamber reactions may be encountered in such cases as a result of uveal tissue irritation from neoplastic cells that are layered in the anterior chamber of the eye. Although the color of both "non-infectious uveitic hypopyon" and "non-tumoral pseudohypopyon" are generally "white or grayish-white,"[2] ocular lymphoma or leukemia causes spontaneous, "blood-streaked," tumoral pseudohypopyon consisting of hemorrhagic neoplastic cells. Indeed, magnetic resonance imaging (MRI) scan showed leukemic deposits in the anterior chamber (tumoral pseudohypopyon), iris, ciliary body, and pars plana, which responded well to reinduction therapy and external beam radiotherapy. It should be remembered that secondary inflammatory reactions in the anterior chamber of the eye after pseudohypopyon irritation require steroidal and/or non-steroidal anti-inflammatory drop application along with a cycloplegic agent. Therefore, if the young girl has received such management, posterior synechiae formation may not have been encountered in this pediatric patient post-radiotherapy.

Taken together, the type of meniscus in the present young girl may be named as descriptive as possible, *e.g.*, *"a 5-year-old child with unilateral, blood-tinged, pinkish, (possibly) shifting, 2-mm cold macro-pseudohypopyon,"* which may direct clinicians away from hypopyon formation with an inflammatory etiology, and rather to a potential list of "tumoral pseudohypopyon." Therefore, if the anterior chamber meniscus is examined with its complete features (shape, mobility, and color), pediatricians and ophthalmologists can make (or exclude) an ocular or systemic diagnosis on time and direct such children to the proper department or unit for prompt medication.

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Conflict of interest

There are no conflicts of interest.

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