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# High-altitude retinopathy – case report

## *Retinopatia wysokogórska – opis przypadku*

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**Summary:** High-altitude retinopathy is one of altitude-related illnesses. Its signs include high-altitude retinal hemorrhages, dilated vessels and peripapillary hyperemia. Increased intracranial pressure seems to be the main cause of all high-altitude diseases including high-altitude retinopathy, cerebral oedema and high-altitude pulmonary oedema. We present the case of high-altitude retinopathy in a 35-year-old woman who reported decreased vision in her right eye, scotomas and high-altitude retinopathy after ascending to more than 7000 meters above sea level. The associated optical coherence tomography findings, fundus photography and literature review are presented.

High-altitude retinopathy is an important multifactorial condition of unknown mechanism and etiology, which significantly impacts human vision. Climbing high mountains can cause retinopathy in otherwise healthy people and may lead to permanent sequelae such as retinal nerve fiber layer and optic nerve defects. These symptoms, however, may resolve without causing any permanent damage to the retina. Conservative treatment may help to relieve them.

With increasing popularity of mountaineering, ophthalmologists should be prepared to diagnose and treat high-altitude retinopathy.

**Key words:** high-altitude retinopathy, mountain sickness, retinal hemorrhages.

**Streszczenie:** Retinopatia wysokogórska należy do chorób związanych z przebywaniem na dużych wysokościach. Do objawów retinopatii wysokogórskiej należą: krwotoki siatkówkowe powstające podczas przebywania na znacznych wysokościach, rozszerzone naczynia i przekrwienie okołotarczowe. Wydaje się, że główną przyczyną wszystkich zmian w przebiegu choroby wysokościowej, takich jak np. retinopatia wysokogórska, obrzęk mózgu i obrzęk płuc, jest wzmożone ciśnienie śródczaszkowe.

Prezentujemy przypadek retinopatii wysokogórskiej u 35-letniej kobiety, u której doszło do pogorszenia widzenia w prawym oku, powstania mroczków i retinopatii wysokogórskiej po wejściu na wysokość ponad 7000 metrów n.p.m. Przedstawiamy wyniki badania optycznej koherentnej tomografii, fotografię dna oka oraz przegląd literatury medycznej.

Retinopatia wysokogórska jest ważną jednostką chorobową, ponieważ znacząco wpływa na pogorzenie wzroku. Przyczyna choroby jest wieloczynnikowa, dokładny mechanizm i etiologia zaś pozostają nieznane. Wspinaczka wysokogórska może spowodować retinopatię u zdrowych osób, w konsekwencji może skutkować trwałymi następstwami (np. ubytkami włókien nerwowych siatkówki i ubytkami włókien nerwowych nerwu wzrokowego). Objawy mogą też ustąpić, nie pozostawiając żadnych trwałych uszkodzeń siatkówki. Leczenie zachowawcze może być pomocne w zmniejszeniu nasilenia objawów.

Ze względu na wzrastającą popularność wspinaczek wysokogórskich lekarze okuliści często mogą mieć do czynienia z przypadkami retinopatii wysokogórskiej, które nie powinny im przysparzać trudności ani w procesie diagnozowania tej jednostki chorobowej, ani w procesie jej leczenia.

**Słowa kluczowe:** retinopatia wysokogórska, choroba wysokościowa, krwotoki siatkówkowe.

### Introduction

High-altitude retinopathy (HAR) is part of a syndrome of clinical entities that define high-altitude illnesses, namely: acute mountain sickness (AMS), high-altitude pulmonary oedema (HAPE), and high-altitude cerebral oedema (HACE) (1–3).

The signs of HAR include high-altitude retinal hemorrhages, dilated vessels and peripapillary hyperemia. Increased intracranial pressure (ICP) seems to be the main cause of high-altitude diseases including HAR, HACE and HAPE (1).

The development of high-altitude illnesses depends not only on the altitude, but also the rate of ascent, the individual susceptibility, the baseline cardiopulmonary status and other possible genetic and environmental factors (2). There are few reports describing ocular changes related to AMS.

### Case report

We present a case of bilateral high-altitude retinopathy in a 35-year-old woman during exposure to high altitude. The patient complained about decreased vision in her right eye, first noted while she was trekking to a maximum altitude of 7134 meters on the border of Tajikistan and Kyrgyzstan 1 week earlier. She was not using supplemental oxygen while mountaineering, she did not have any frostbite injuries, either. The patient did not use any medication during the trip and was not taking any medication when she presented at our department. She was not hypertensive and had no previous history of any ocular or systemic disease and ocular trauma. She had already been to high mountains before, however, she had never experienced similar symptoms.


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The patient attributed her decreased vision to the fact that she frequently took her sunglasses off in order to see the camera display better.

A complete ophthalmic examination was performed, including best corrected visual acuity using standard Snellen charts, tonometry, slit-lamp biomicroscopy and funduscopic examination.

On admission, the best corrected visual acuity (BCVA) was 0.1 RE and 1.0 LE. Intraocular pressure (IOP) was 10 mmHg in both eyes. Anterior segment appeared normal in both eyes. Dilated fundus examination revealed bilaterally clear, quiet vitreous and flat, intact optic nerve discs. There were retinal hemorrhages located in the right macula, as well as superficial retinal hemorrhages along the vascular arcades in both eyes (Fig. 1). Color vision was normal, with no afferent pupillary defect in both eyes, and no signs of papilloedema.

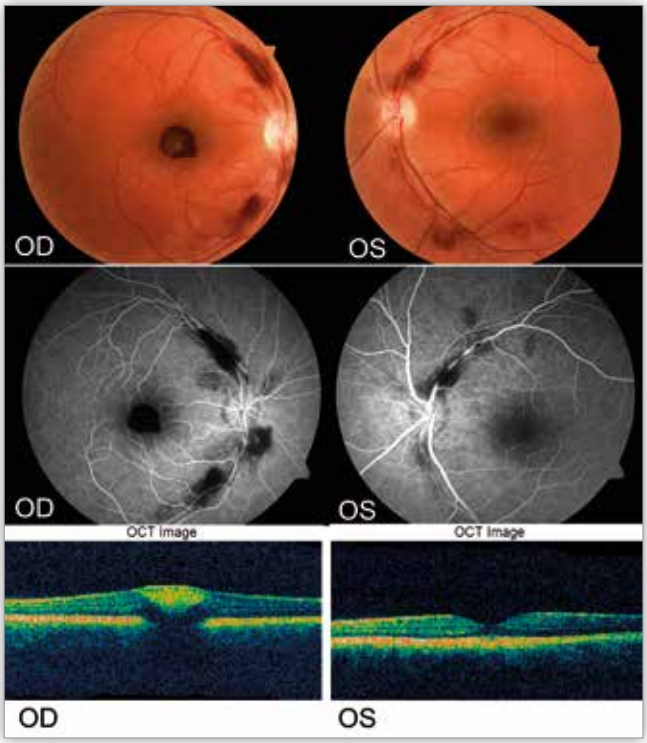


**Fig. 1.** Examinations performed on admission: color fundus photographs, fluorescein angiography and macular OCT, showing multiple intraretinal hemorrhages, including a single macular hemorrhage in the right eye.

**Ryc. 1.** Badania wykonane w dniu przyjęcia pacjentki do szpitala: kolorowa fotografia dna oka, angiografia fluoresceinowa i OCT plamki – rozsiane wybroczyny śródsiatkówkowe, w tym pojedynczy wylew krwi do plamki w prawym oku.

Color fundus photographs were taken and additional examinations were performed, i.e. fluorescein angiography, macular optical coherence tomography (OCT) with the Stratus OCT (Carl Zeiss Meditec) after mydriasis with 1% Tropicamide.

OCT imaging showed superficial intraretinal hyperreflectivity at the fovea in the right eye, corresponding to the retinal hemorrhages. OCT scans of the fovea in the left eye were normal.

Fluorescein angiography revealed areas of hypofluorescence, corresponding to fluorescein blockage, due to the retinal hemorrhages.

Head CT scan was performed in order to exclude intracranial bleeding, however it was unrevealing, just like the chest x-ray. Standard blood tests were performed – full blood count, haemoglobin, hematocrit, coagulation panel (APTT, fibrinogen, D-dimers), liver function tests, creatinine, urea, lipids, glucose, electrolytes, von Willebrand factor, CRP. Blood tests were normal, von Willebrand disease was not confirmed.

The patient's pregnancy test was negative. Consultation of internal medicine specialist and a hematologist did not reveal any pathologies.

The patient's retinal hemorrhages were treated with oral Methylprednisolone 32 mg for 10 days, with tapering doses afterwards. Oral Calcium with vitamin C, as well as Aspargin (magnesium and potassium supplementation) and Cyclonamine (etamsylate) were also added.

On discharge, one week later, BCVA amounted to 0.2 RE and 1.0 LE. Intraocular pressure (IOP) was 10 mmHg in both eyes.

On follow-up, 3 months after the initial visit, the patient's BCVA improved to 1.0 in the right eye. Ophthalmoscopy and macular OCT scan showed almost complete resolution of the retinal hemorrhages (Fig. 2). Visual fields were normal.

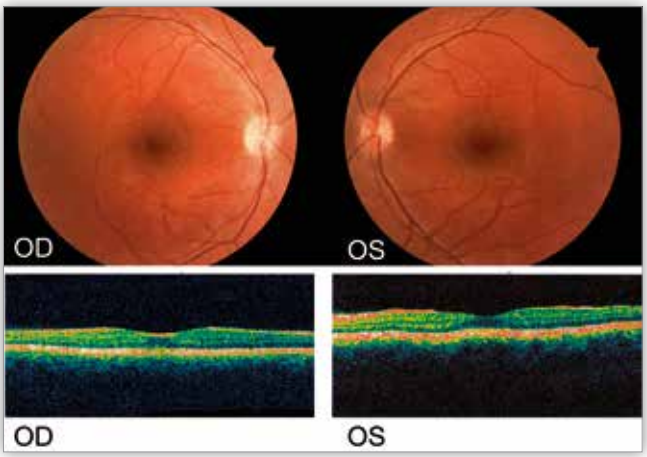


**Fig. 2.** Color fundus photograph and macular OCT at 3 months – the central macular haemorrhage has resolved.

**Ryc. 2.** Kolorowa fotografia dna oka i wynik badania OCT plamki po 3 miesiącach leczenia – wylew krwi do plamki uległ wchłonięciu.

## Discussion

It is known that people who stay at altitudes higher than 3600 m are at risk of developing retinal hemorrhages. High-altitude retinopathy (HAR) is a common finding among travelers at altitudes exceeding 5000 m and may precede more serious manifestations of high-altitude sickness, including cerebral or pulmonary oedema. High-altitude pulmonary oedema is a noncardiogenic pulmonary oedema occurring in otherwise healthy subjects at high altitudes, which can be life-threatening (4). An increasing number of unacclimatized people is exposed to high-altitudes, which in turn increases the frequency of AMS (3). It is thought that AMS and HACE may be part of the same clinical entity and that the frequency of HAR in AMS has a multifactorial cause: the altitude, the rate of ascent, the individual susceptibility, the baseline cardiopulmonary status as well as other genetic and environmental factors (3).

Compared to its weight, the retina shows the highest metabolic activity of all human tissues. Moreover, vascular blood supply

| | Grade I/ Stopień I | Grade II/ Stopień II | Grade III/ Stopień III | Grade IV/ Stopień IV |
|---|---|---|---|---|
| Dilated retinal veins/ Poszerzenie naczyń żylnych siatkówki | + | + | + | + |
| Venule-arteriole ratio (v:a)/ Stosunek szerokości n. żylnych do tętniczych (wsp. A/V) | 3: 2 | 3.5: 2 | 4: 2 | 4.5: 2 |
| Retinal and/or pre-retinal hemorrhages/ Krwotoki siatkówkowe lub/oraz przedsiatkówkowe | < 1 disc area/ < 1 pole tarczy n. II | < 2 disc areas/ < 2 pola tarczy n. II | < 3 disc areas/ < 3 pola tarczy n. II | more than 3 disc areas/ powyżej 3 pól tarczy n. II |
| Other signs/ Inne objawy przedmiotowe | - | - | paramacular hemorrhages or vitreous hemorrhages/ krwotoki okołoplamkowe lub wylewy do ciała szklistego | macular hemorrhages, vitreous hemorrhages, or papilloedema/ krwotoki w plamce, wylewy do ciała szklistego lub obrzęk tarczy n. II |

**Tab. I.** A classification system for HAR, as described by Wiedman et al. (10).
**Tab. I.** Klasyfikacja HAR wg Wiedmana I wsp. (10).

to the brain and the retina have a close anatomic correlation and similar vascular regulatory mechanisms (3). The exact pathogenesis of HAR is not well known. The proposed mechanism is vasodilatation resulting from hypoxia and connected with an increased arterial pressure (1). As the retinal blood flow is regulated by tissue oxygen tension, a decrease in arterial oxygen partial pressure quickly induces an increase in retinal blood flow (2, 3).

It is thought that dysfunctional autoregulation causes cerebral blood flow to increase, which may lead to capillary overperfusion and cause vasogenic cerebral oedema (3, 4). It has also been postulated that the increased intracranial pressure may be the principal cause of high-altitude diseases, namely HAR, HACE and HAPE (4).

Retinal hemorrhages may go unnoticed, unless they are large enough to impair vision or occur near the macula, which happened to our patient. In the right eye, the macular hemorrhage caused decreased vision, whereas in the left eye hemorrhages were located near large retinal vessels, the macula was uninvolved, so the patient retained full visual acuity.

Seth et al. (2) demonstrated by OCT that the hemorrhages are located in the superficial retina. In our case, macular OCT confirmed that the macular hemorrhage in the right eye was located in superficial retinal layers. McFadden et al. (5) described engorgement and tortuosity of the retinal vessels, optic disk hyperemia, as well as occurrence of retinal hemorrhages with a white center (a fibrin-platelet thrombus, so called Roth spots), which are due to capillary fragility and rupture. This phenomenon supports the proposed explanation of HAR etiology, according to which retinal bleeding results from hypoxic damage to vascular endothelial cells at high-altitudes, which causes a vascular wall rupture after some time and delayed blood leakage. Other possible explanations include: increased blood viscosity, increased blood clotting, decreased oxygen transport capacity and increased blood flow, impaired autoregulation, increased sympathetic activity, increased capillary permeability, ischemic-reperfusion injury causing endothelial cell damage, neutrophil activation and formation of reactive oxygen species (2–4, 6).

The initial symptoms of AMS usually start a few days after the arrival at high-altitudes to lessen a few days later. The mild persisting symptoms include headache, dizziness, nausea, vomiting, fatigue, weakness and insomnia (3, 4).

The signs of HAR include decreased visual acuity, scotomas, visual field changes, high-altitude retinal hemorrhages, dilated and tortuous retinal vessels, peripapillary hyperemia, optic disc swelling, as well as nerve fiber layer infarction and vitreous hemorrhage (3–5). More severe symptoms may develop, such as: high-altitude pulmonary oedema (HAPE) and/or high-altitude cerebral oedema (HACE) with severe hypoxemia being the potential cause of a quick progression from AMS to HACE (3).

The retina is the only part of the central nervous system where the non-invasive visualization and measurement of nerve fibers is feasible. There have been several studies to demonstrate optic disc oedema due to increased retinal capillary blood flow and RNFL thickness increase using optical coherence tomography (OCT) (3). The mean peripapillary RNFL thickness is said to increase after high mountain expedition, even if it is not accompanied by any subjective clinical symptoms (3). Wiedman et al. (7) proposed a four-grade HAR classification system (Table I). As the total area of preretinal hemorrhages in our patient was less than 3 disc areas and macular hemorrhage was present, without concomitant vitreous hemorrhage or papilloedema, she met the criteria of grade 3 HAR.

HAR may be mild and transient. However, it may also lead to permanent sequelae. Ho et al. (4) found that the best-corrected visual acuity almost returned to baseline values three weeks after a high mountain expedition, however, vascular engorgement and tortuosity of the retinal vessels were still present in all studied eyes. They also found retinal nerve fiber layer defects in some climbers, some time after the expedition, as well as optic nerve defects on GDx examination. Arora et al. (6) found that 10 out of 50 subjects who were in high mountains had HAR and 4 climbers had branch retinal vein occlusion with macular oedema. They also found that the majority of patients recovered fully, however, patients with severe lesions such as premacular hemorrhage, showed partial recovery and in 2 individuals no visual recovery was confirmed even after 24 weeks.

Most papers, published to date, focus on HAR etiology and symptoms, without the detailed therapeutic recommendations. There have been no guidelines for HAR treatment yet. It is therefore important to discuss various possible treatment modalities in order to establish an optimal treatment method.


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Published reports emphasize the crucial role of prevention. It is important that climbers ascend slowly, taking time to get used to the altitude. Supplemental oxygen as well as descent from high altitude are recommended in subjects who already developed symptoms of high-altitude illness.

Various therapies for AMS, HAR and HACE have been described, e.g.: hyperbaric oxygen therapy (HbOT), supplemental oxygen, acetazolamide, dexamethasone, Ginkgo biloba, and non-steroidal anti-inflammatory drugs (NSAIDs) (2). In our patient, we chose conservative treatment. In order to decrease capillary leakage, blood cell extravasation and possible optic nerve fiber oedema, oral Methylprednisolone was started. Steroids show strong anti-inflammatory effect, they constrict blood vessels and decrease vascular permeability, which minimizes leakage of fluid, proteins and blood cells.

In order to protect vascular endothelium, decrease capillary permeability and improve hemorrhage resorption, Calcium, vitamin C and etamsylate were introduced. Three months later, the patient's BCVA returned to normal and she did not report any vision complaints. This improvement was confirmed by fundus photographs and macular OCT, which were normal.

## Conclusion

HAR is an important condition due to its potential impact on vision. Moreover, it is associated with AMS and life-threatening HACE (1, 2). HAR has a multifactorial etiology and its exact mechanism and causes are still unknown.

Climbing high mountains can cause retinopathy in otherwise healthy individuals leading to permanent sequelae (e.g. retinal nerve fiber layer defects and optic nerve defects). Nevertheless, symptoms may resolve without causing any permanent damage to the retina.

With increasing popularity of mountaineering, ophthalmologists should be prepared to diagnose and treat HAR. Conservative treatment may help to relieve symptoms.

## Financial Disclosure

Authors confirm that they do not have any commercial or proprietary interest in any product or company mentioned.

## References:

1. Vinger PF: *Sports Injuries* (in:) Albert D. M., Jakobiec F. A. (eds.) Principles and Practice of Ophthalmology, W. B. Saunders Company, 2000, CD-ROM.
2. Seth RK, Adelman RA: *High-altitude retinopathy and optical coherence tomography findings*. Semin Ophthalmol. 2010; 25(1–2): 13–15.
3. Ascaso FJ, Nerin MA, Villen L, Morandeira JR, Cristobal JA: *Acute mountain sickness and retinal evaluation by optical coherence tomography*. Eur J Ophthalmol 2012; 22: 580–589.
4. Tsung-Yu Ho, Wei-Fong Kao, Shui-Mei Lee, Po-Kang Lin, Jin-Jong Chen, Jorn-Hon Liu: *High-altitude Retinopathy after climbing mount aconcagua in a group of experienced climbers*. Retina 2011; 31: 1650–1655.
5. McFadden DM, Houston CS, Sutton JR, Powles AC, Gray GW, Roberts RS.: *High-altitude retinopathy*. JAMA 1981; 245: 581–586.
6. Arora R, Jha KN, Sathian B: *Retinal changes in various altitude illnesses*. Singapore Med J 2011; 52(9): 685–688.
7. Wiedman M, Tabin GC: *High-altitude retinopathy and altitude illness*. Ophthalmology 1999; 106: 1924–1927.

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