Purtscher's retinopathy – case report and literature review

Retinopatia Purtschera – opis przypadku i przegląd piśmiennictwa

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Słowa kluczowe: uraz, mikroembolizacja, retinopatia Purtschera.

Abstract

Purtscher's retinopathy is a vision-threatening state that most frequently occurs in young or middle-aged males after head or chest injury. The main symptom reported by patients is sudden, painless deterioration of vision, which can affect one or both eyes. In most patients, the changes observed in the fundus of the eye resolve within several months, and visual acuity returns to the state from before the injury. The occurrence of ischaemic changes concerning the macula or optic nerve disc may cause permanent deterioration of visual acuity. We present a case of a 43-year-old male with Purtscher's retinopathy after a traffic accident. The article discusses clinical image, aetiology, and the current state of knowledge concerning the possibilities in diagnostics and treatment of Purtscher's retinopathy.

Streszczenie

Retinopatia Purtschera jest stanem zagrażającym widzeniu, występującym najczęściej u mężczyzn w młodym lub średnim wieku w następstwie urazów głowy lub klatki piersiowej. Głównym objawem zgłaszanym przez pacjentów jest nagłe, bezbolesne pogorszenie widzenia, które może dotyczyć jednego lub obu oczu. U większości pacjentów zmiany obserwowane na dnie oka ustępują w ciągu kilku miesięcy i ostrość wzroku wraca do stanu sprzed urazu. Wystąpienie zmian niedokrwiennych dotyczących plamki lub tarczy nerwu wzrokowego może skutkować trwałym pogorszeniem ostrości wzroku. W pracy przedstawiono przypadek retinopatii Purtschera u 43-letniego pacjenta po wypadku komunikacyjnym. Opisano obraz kliniczny, etiopatogenezę oraz aktualny stan wiedzy dotyczącej możliwości diagnostyki i leczenia retinopatii Purtschera.

Introduction

In 1910, Dr. Othmar Purtscher (1852–1927) described the image of the fundus of the eye in a patient who had undergone a severe head injury. The description presented disseminated, surface, non-uniform, white foci with accompanying retinal haemorrhages [1]. Since then, analogous changes observed in the fundus of the eye have been termed as Purtscher retinopathy (PR). In cases when the changes equivalent to PR do not develop as a consequence of injury, but result from acute pancreatitis (AP), collagenosis, systemic lupus erythematosus, lymphoproliferative disorders, bone marrow transplantation, breast plasty, orthopaedic surgeries, or intravitreal injection, they are called Purtscher-like rethinopahy [2].

At present, microembolisation of the retinal and choroidal vessels with the development of ischaemic zones is considered the most probable cause of occurrence of PR, as well as Purtscher-like rethinopahy [2–4].

The main symptom of retinopathy is sudden, painless deterioration of vision, which may occur directly or within several days after injury. The prognosis in the course of retinopathy is uncertain [2–5].

We present a case of a 43-year-old male with unilateral PR following a traffic accident.

Case report

As a consequence of a traffic accident the patient had experienced L3 vertebral body fracture and fracture of the body of the sternum. The patient was hospitalised in the Orthopaedic Ward. Computed tomography scan of the head and mandible did not show any post-injury changes. On the first day after the accident, the patient reported sudden deterioration of vision in the right eye. In ophthalmologic examination, visual acuity in the right eye was on the level of counting fingers at 0.5 m, and visual acuity in the left eye was 1.0 accord-

ing to the Snellen scale. Intraocular pressure in both eyes remained within the normal range. Examination of the anterior segment in both eyes did not reveal any deviations from the normal state. Examination of the fundus of the right eye showed multiple confluent foci of cotton wool spots, massive retinal oedema, intraretinal haemorrhages, and Purtscher flecken (Figure 1). The image of the fundus of the left eye did not differ from the normal state (Figure 2).

Spectral-domain optical coherence tomography (SD-OCT Copernicus HR from Optopol) showed hyperreflectivity in the inner retinal layer, and cystoid macular oedema with serous retinal detachment of right eye. The central retinal thickness (CRT) in the right eye was 633 µm (Figure 3), and in the left eye it remained within the normal range.

In the fluorescein angiography (FA) examination (Visucam 500, Carl Zeiss) an area of hypofluorescence was found corresponding to the lack of perfusion between vascular arches, smaller foci of hypofluorescence corresponding to the intraretinal haemorrhages (Figure 4), and closed arterioles with leakage in the later stages (Figure 5).

In the case described, the diagnosis of PR was made based on the clinical image and results of the above-mentioned examinations.

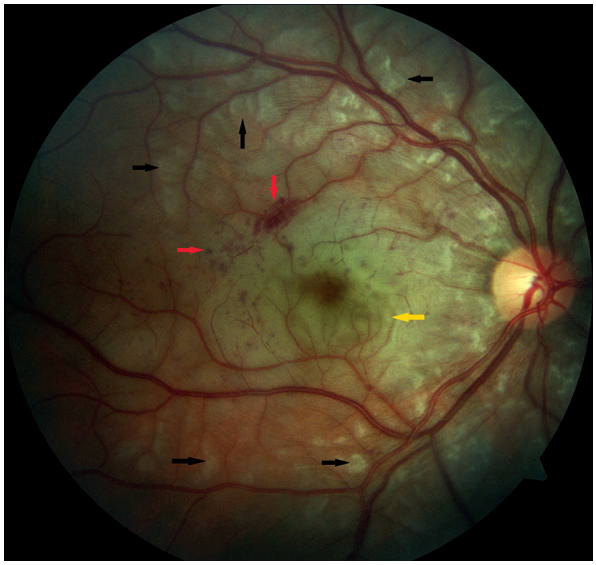

Figure 1. Image of the fundus right eye-visible swelling of the retina with merging puffs of cotton wool spots (yellow arrow), Purtscher flecken (black arrows), and retinal haemorrhages (red arrows)

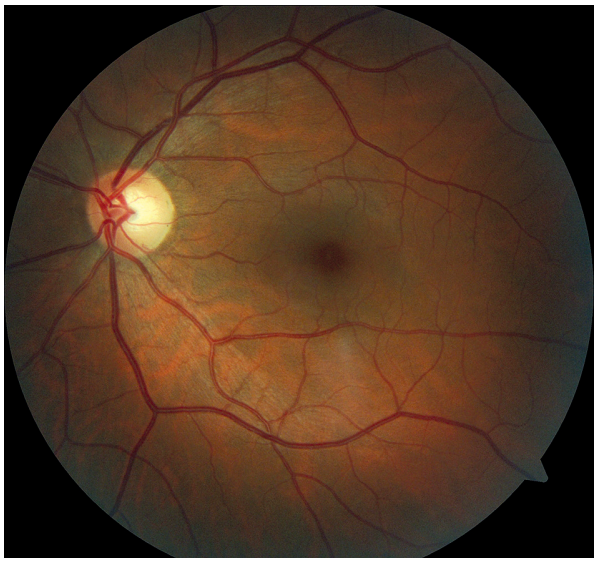

Figure 2. Image of the fundus left eye – normal image of the fundus

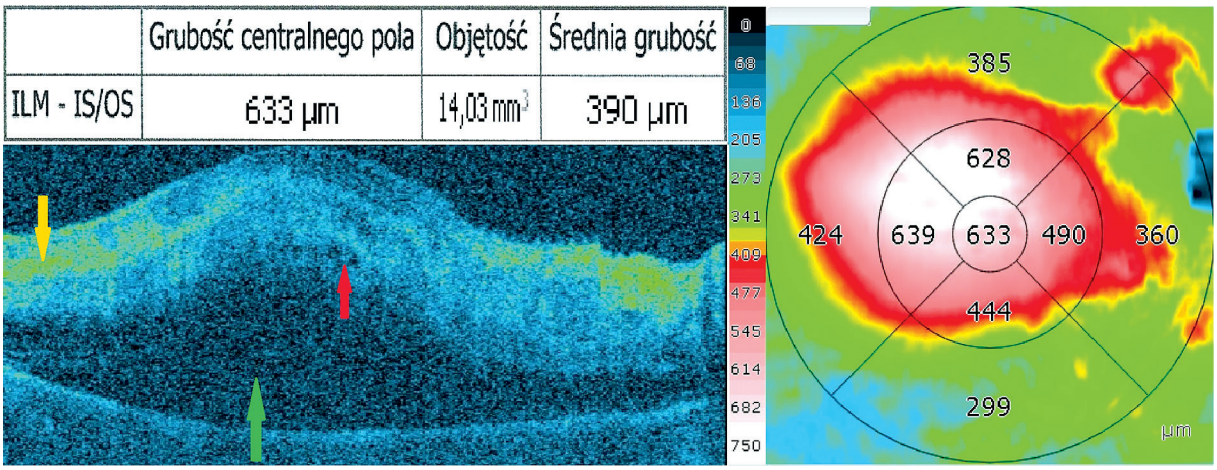

Figure 3. Optical coherence tomography image of the right eye with a map of the thickness of the retina. Optical coherence tomography shows hyper-reflectivity in the inner retinal layer (yellow arrow), cystoid macular oedema (red arrow), and serous retinal detachment (green arrow)

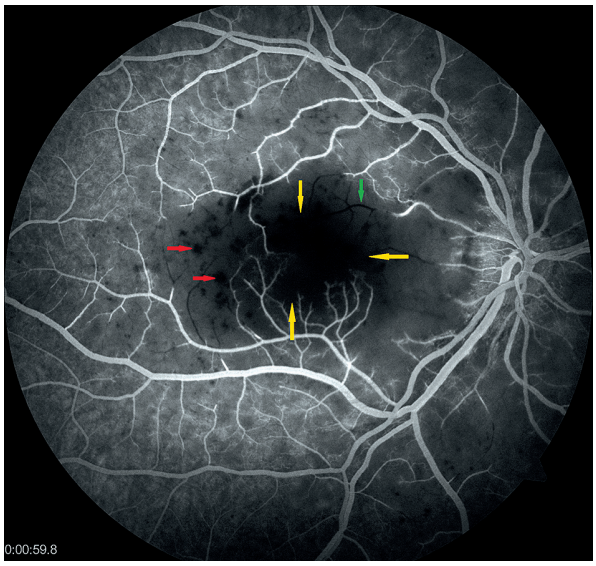

Figure 4. Fundus of the right eye in the first minute of the FA examination. Hypofluorescence area corresponds to the lack of perfusion (yellow arrows), retinal haemorrhages (red arrows), and visible closed blood vessels (green arrow)

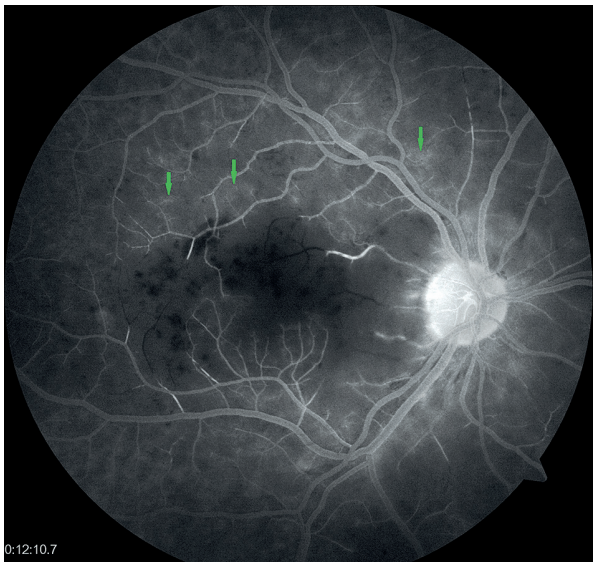

Figure 5. Image of the fundus of the right eye 12 min after FA. Changes visible in Figure 4 accompanied by the leakage of small precapillary vessels (green arrows)

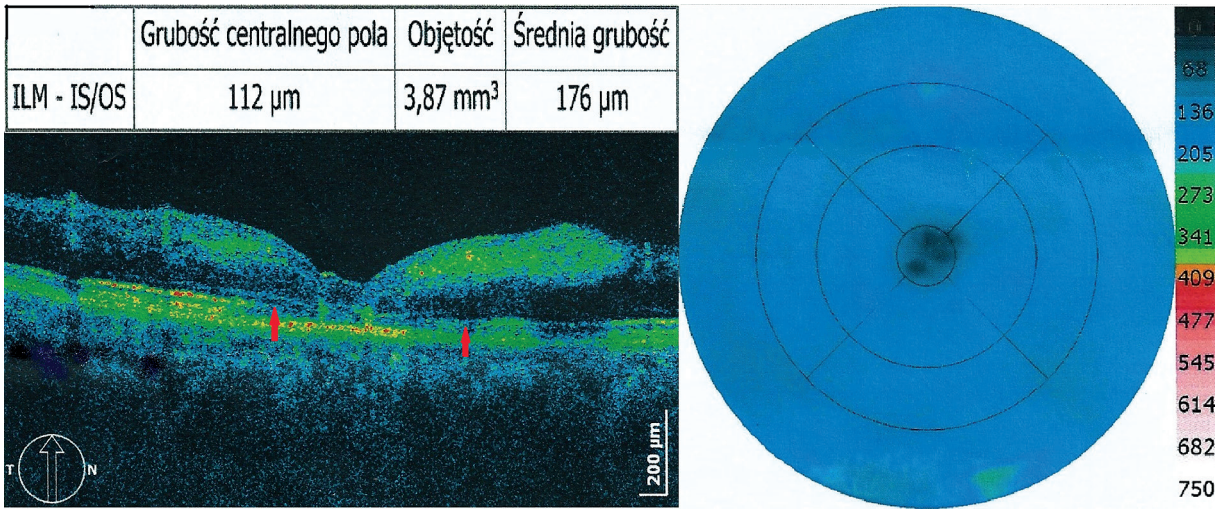

Figure 6. Optical coherence tomography image of the right eye with a map of the thickness of the retina. State two months after injury. Optical coherence tomography shows deterioration of the subretinal fluid, IS/OS defect (red arrows), and atrophy of the macular retinal layers

The treatment covered general administration of steroids and anti-oedema drugs (mannitol, carbonic anhydrase inhibitors), a local administration of steroid drops, and non-steroid anti-inflammatory drugs.

On day 10 after the accident, the patient was discharged home. Visual acuity in the right eye was limited to counting fingers at 3 m. In the image of the fundus of the eye, a partial regression of changes was observed in the form of reduced oedema. Steroid therapy was maintained in decreasing doses, both generally and locally.

In the second month after the accident, visual acuity in the right eye was – 0.3, and in the left eye – 1.0, according to the Snellen scale. In the SD-OCT image the subretinal fluid deteriorated completely but we observed considerable decrease in central retinal thickness to 112 µm, corresponding to its atrophy and photoreceptor inner/outer segment (IS/OS) defect (Figure 6). Examination of the fundus of the eye showed persisting intraretinal haemorrhages, as well as whitening of the optic nerve disc. The FA exami-

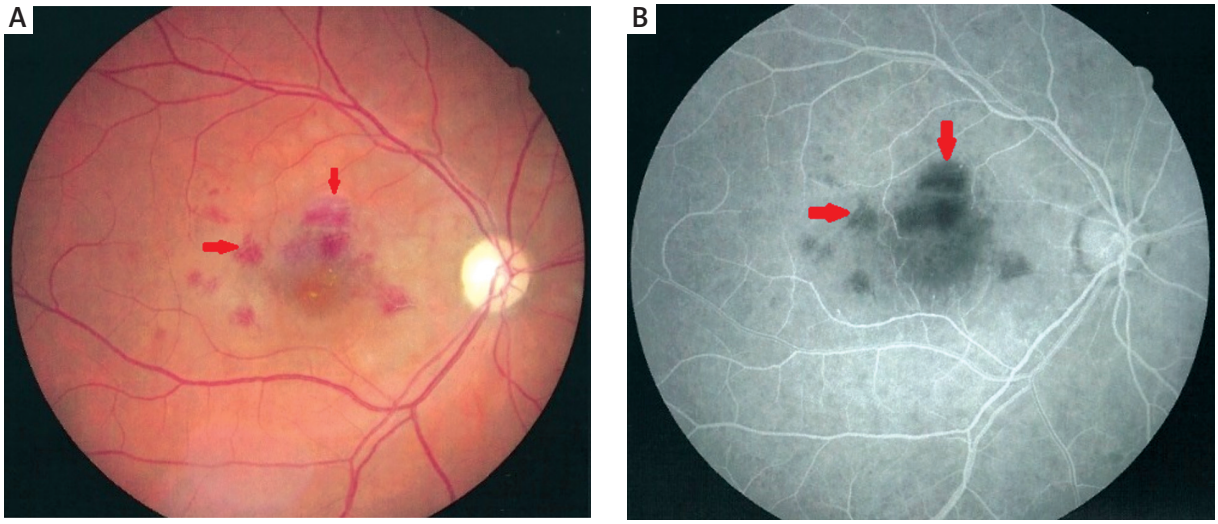

Figure 7. A – image of the fundus right eye-visible pale optic nerve disc and retinal haemorrhages (red arrows). **B** – image of right eye fundus during FA examination – shows reduction of ischaemic areas and improved capillary perfusion; hypofluorescence area corresponds to the retinal haemorrhages (red arrows). State 2 months after injury

nation revealed reduction of ischaemic areas and improved capillary perfusion (Figure 7).

Discussion

Purtscher retinopathy is a vision-threatening condition most frequently observed in young or middle-aged males as a consequence of head or chest injuries [3]. A considerable number of PR cases refer to retinopathies that develop after traffic accidents [6–13].

Data concerning the incidence of PR are varied. Agrawal *et al.* estimated the frequency of occurrence of PR and Purtscher-like retinopathy at 0.24 patients per million annually [4]. These data pertain to patients from the UK and Ireland. However, it is considered that the incidence of PR may be higher due to its asymptomatic course in some cases, as well as diagnostic difficulties encountered in patients in a severe general condition [14].

The diagnosis of PR is based on the clinical image and evaluation of the fundus of the eye. The main symptom reported by patients is sudden, painless deterioration of vision, which may affect one or both eyes. Although the binocular presentation of symptoms occurs in 60% of cases, cases of uniocular retinopathy are also known [7–12, 15].

The deterioration of vision may occur directly after the injury or within 24–48 h after the injury [3]. In the presented case, the deterioration was observed on the first day after injury.

Examination of the fundus of the eye is indispensable to confirm the diagnosis of PR. Typical changes in the image of the fundus of the eye include Purtscher flecken, cotton wool spots, macular oedema, swelling of the optic disc, and retinal haemorrhages [3]. In the presented case, fundus examinations showed retinal haemorrhages, cotton wool spots, massive retinal oedema, and many Purtscher flecken on the posterior pole of the right eye (Figure 1). The pathognomonic Purtscher flecken in this pathological entity are areas of retinal whitening clearly demarcated by the area within 50 µm from the normal retina. It is considered that the occlusion of precapillary arterioles is responsible for the development of Purtscher flecken [3].

The SD-OCT may be a supplementary test in the diagnostics of PR. On the SD-OCT image, cotton wool spots appear as hyperreflective areas in the inner layers of the retina [16]. Purtscher flecken on an SD-OCT scan correspond to increased reflectivity of the intranuclear layer (INL) [17]. In the presented case, the SD-OCT also revealed hyperreflective inner retinal layers, including the INL (Figure 3). It is presumed that increased reflectivity of the INL may be a marker of the extent of the ischaemia [18].

The SD-OCT imaging allows visualisation of the varying extent of macular oedema, which, when considerably advanced, may lead to the disintegration of the layers of the retina, detachment of the neurosensory retina and retinal pigment epithelium, and, consequently, diminished visual acuity. Seung *et al.* report a case of Purtscher's retinopathy with diffuse serous macular detachment [19]. In our case, SD-OCT showed similar changes with neurosensory retinal detachment as a consequence of subretinal fluid accumulation (Figure 3).

Analysis of the retinal thickness allows observation of the evolution of changes. The thickness of the retina directly affects its function; therefore, the results of retinal thickness measurement may be used as a prognostic factor. In our case, the CRT in the right eye was 633 µm (Figure 3). It was confirmed that

a quick remission of oedema is associated with a more favourable prognosis concerning visual acuity [5].

Additional useful examination in PR is obtained with an FA examination, which can visualise the circulation of the retina and choroid. Changes visible in FA include blockade of choroidal fluorescence, occluded arterioles, areas of non-perfusion, as well as late leakage from the retinal vessels in areas of ischaemia, and optic disc oedema [20]. In the presented case, in FA examination, severe capillary non-perfusion was observed in the early phase (Figure 4), and late phase showed leakage of fluorescein dye in the non-perfusion areas (Figure 5). It was found that the area of choroidal hypofluorescence may persist up to 5 months after the diagnosis [21]. In our case, in the second month after the accident, the FA examination revealed reduction of ischaemic areas and improved capillary perfusion (Figure 7).

There are many theories concerning the pathogenesis of PR. Purtscher connected the described changes with extravasation of lymph from the retinal vessels as a result of increased intracranial pressure after injury [1]. Another proposed mechanism of the development of changes corresponding to the PR image was the widening of venous blood vessels as a result of increased pressure in the chest secondary to its compression. It is considered that compression of the chest, resulting from chest compression by safety belts during traffic accidents, may led to the development of retinopathy [9, 22]. Holak *et al.* suggested that a varied haemodynamic situation occurring at the moment of injury might explain the presence of unilateral changes [7].

At present, microembolisation of the retinal and choroidal vessels is considered as the most probable mechanism in the development of both PR and Purtscher-like retinopathy [3, 4, 7]. According to the cause of retinopathy, the occlusive material may be: the air, lipids, aggregates of leukocytes, thrombocytes, fibrin, or immunoglobulins. Both the trauma and inflammatory state, e.g. acute pancreatitis, lead to activation of the complement system, the role of which in the pathogenesis of retinopathy has been confirmed by laboratory tests [23, 24]. Analyses of computer fluid dynamics simulations suggest that in the course of retinopathy there occur rheological disorders, which cause lesions of the endothelium. Probably these mechanisms do not exclude each other [25].

Currently, there is not one mechanism which would combine all the possible causes of the development of retinopathy, nor a uniform standard of treatment of patients with PR. The majority of researchers recommend the administration of steroids during treatment. There are reports concerning improvement of visual acuity after the use of steroid therapy; however, there are no randomised studies confirming the effectiveness of such a management [18, 26–28]. In our case, after steroids treatment visual acuity in the right eye improved from the level of counting fingers at 0.5 m to 0.3 m according to the Snellen scale.

Holak *et al.*, as the main goal of administration of steroids, consider their beneficial effect in the reduction of oedema. In their opinion, quick remission of swelling of the retina is the factor which is favourable from the aspect of prognosis [5, 7].

The prognosis in the course of retinopathy is uncertain. In the majority of patients, changes observed in the fundus of the eye disappear within 2 months, and visual acuity returns to the state from before the trauma [3]. In severe cases, changes in the retinal pigment epithelium may be observed, as well as macular and optic nerve disc atrophy or neovascularisation, which lead to irreversible deterioration of vision [3, 29]. Functional changes, such as non-acute central vision and scotoma in the field of vision, persist in 50% of cases.

According to Holak *et al.*, the prognosis in PR depends mainly on the duration and the extent of severity of the changes observed during the early period of the disease. Quick remission of oedema is related with favourable prognosis with respect to visual acuity, irrespective of other factors, and the duration of acute retinal changes is the decisive factor for late prognosis [5].

In the presented case, despite the remission of subretinal fluid and oedema accompanied by an improvement of visual acuity, on the SD-OCT scan there was an IS/OS defect and atrophy of the macular retinal layers, which, as well as whitening of the optic nerve disc, are a consequence of the ischaemic changes and are related with the lack of possibilities to regain visual acuity to the pre-trauma state.

Conclusions

Patients who experienced injuries and traffic accidents should be provided ophthalmological consultations for the occurrence of PR. Both the patients who report vision disorders and those in a severe general condition, without contact, who are unable to report such disorders, should be examined. In the case of severe changes that persist for a long time, atrophy of the macula, optical nerve disc, or neovascularization may occur, leading to irreversible deterioration of vision. Despite the lack of uniform standards of treatment, it seems justified to attempt to apply steroid therapy in patients with PR. In each case, such a decision should be made individually, considering also the undesirable effects and limitations of such a treatment.

Conflict of interest

The authors declare no conflict of interest.

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