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Purtscher and Purtscher-Like Retinopathies: What Do We Know?

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Abstract

Purtscher and Purtscher-like retinopathies are rare but important to recognize. Its diagnosis is clinical, in which at least 3 of the following diagnostic criteria should be present: 1) Purtscher-flecken (visible in the fundoscopy, they are intra-retinal whitening areas with a clear zone on either side of the vessels), 2) intra-retinal hemorrhages, 3) cotton wool spots (these are located superficially to the vessels and have ill-defined edges), 4) plausible etiology, 5) compatible complementary examination (such as angiography and Optical Coherence Tomography). Purtscher's retinopathy is traumatic (usually head trauma, chest compression or long bone fracture) and the term Purtscher-like retinopathy is reserved for non-traumatic causes (acute pancreatitis, HELLP syndrome, carcinoma, among many others).

Supportive treatment should be performed as well as treatment of the underlying etiology. Other treatments, such as corticosteroids, intra-vitreal injection of anti-VEGT or intra-vitreal injection of tissue plasminogen activator, are theoretically logical but it is unknown whether they are beneficial compared to observation. The prognosis is variable and depends on the severity of retinal alterations at presentation, as well as the underlying cause. Further evidence regarding treatment and prognosis is necessary, but difficult to conduct considering this is an uncommon disease.

Keywords: Purtscher; Purtscher-like; Retinopathy; Medical retina; Review; Corticosteroids

Introduction

Purtscher and Purtscher-like retinopathies are rare [1] but important to recognize clinically. Purtscher's retinopathy is due to trauma [2] and its fundoscopic signs may include [3]: Purtscher-flecken (which are considered to be pathognomonic) [4], intra-retinal hemorrhages, cotton wool spots, among others.

Purtscher-like retinopathies are believed to have a similar pathology but their etiologies are non-traumatic, namely: acute pancreatitis [5], valsalva maneuver [6], thrombotic thrombocytopenic purpura [7], lupus [8].

Angiography, Optical Coherence Tomography (OCT), Visual field and other complementary exams can be performed and are considered useful to support the diagnosis [3].

Regarding treatment, there are controversies, with many doctors administering corticosteroids while others performing a supportive treatment or no treatment (solely observation) [9].

It is important to recognize its clinical signs, treatment and prognostic factors; therefore we performed a systematic review regarding Purtscher and Purtscher-like retinopathies [3], in order to further characterize this retinopathy, to identify prognostic factors and to assess if the treatment with corticosteroids was useful, in comparison with a conservative treatment.

Commentary

This commentary article summarizes the main findings of your systematic review [3], as well as a recent bibliographic review and further describes Purtscher and Purtscher-like retinopathies:

Epidemiology, terminology and etiology

As previously stated, these retinopathies are rare; there are a few studies that tried to assess its frequency [1,10] and it is generally believed that its incidence is of 0.24 persons per million per year [1] or even higher (since they can be asymptomatic [10]).

Purtscher retinopathies are always traumatic whereas Purtscher-like retinopathies are always non traumatic.

Etiologies of Purtscher's retinopathy include mainly: head trauma [2], chest compression [11], long bone fracture [12]. In our systematic review [3], from 670 articles initially found, we included 68 cases of Purtscher or Purtscher-like retinopathies, in which 23 were caused by trauma (33.8%).

Regarding Purtscher-like retinopathy, our study [3] allowed the identification of 13 cases associated with acute pancreatitis (19% of all cases) [5], 6 cases after Valsalva maneuvers [6], 5 cases associated with thrombotic thrombocytopenic purpura [7], 3 cases with hemolytic uremic syndrome [13], 3 cases with cryoglobulinemia in hepatitis C [14], 3 pregnancy-related [15] cases, among others. There are many other etiologies involved in Purtscher-like retinopathies, namely (this list is non-exhaustive and new etiologies continue to be identified and published) pancreatic cancer [16], lupus [8], renal sclerodermia

[4], multiple myeloma [17], nephrotic syndrome [18], retrobulbar anesthesia [19], coil embolization of carotid aneurism [20], thrombotic microangiopathy [21], acute allograft rejection [22], shaken baby syndrome [23] (in which the Purtscher flecken are decisive for differential diagnosis), iron-deficiency anemia [24], prostate surgery [25], dacryocystorhynostomie [26] and even PMMA injection into buttocks (which was called the "Brazilian booty retinopathy" in a report published in 2016) [27].

Pathophysiology

There are several theories regarding the pathology of Purtscher and Purtscher-like retinopathies. It is believed that microembolization is responsible for the occlusion of the pre-capillary arterioles and microvascular infarct of the retinal nerve fiber layer, consequently forming cotton wool spots and Purtscher flecken [11,27]. Microembolization is believed to be caused either by: leukocytary aggregation (with consequent leukoembolization) and complement activation [7,8] or by fat emboli (described in long bone fractures [12]) or by pancreatic proteases in systemic circulation (described in acute pancreatitis [28]).

Other postulated mechanisms include: capillary endothelial damage [15], hyperviscosity [17], sudden increase of intracranial pressure causing pre-capillary occlusion in lamina crivosa [29] or even vascular endothelial dysregulation caused by a rheological event [30].

Histologically, there is edema of retina internal layers and cystoid spaces with an abrupt transition to normal retina [31].

Clinical findings

The main symptom is uni- or bi-lateral sudden loss of vision.

The diagnosis is clinical, although it can be supported by complementary examination. To aid in the diagnosis, the utilization of the following diagnostic criteria is recommended [1,3]:

Diagnostic criteria of Purtscher and Purtscher-like retinopathies

At least 3 of 5 of these criteria should be met:

1. Purtscher flecken
2. Retinal hemorrhages, low-to-moderate number (1-10)
3. Cotton-wool spots (typically restricted to posterior pole)
4. Probable or plausible explanatory etiology
5. Complementary investigation compatible with diagnosis

Purtscher-flecken are very typical but are unfortunately underreported (we identified Purtscher-flecken in 63% of all cases but they were reported in only 23% [3]; other studies show a prevalence of 50% [4]). They are pathognomonic of this pathology; therefore they should be better recognized. They are intra-retinal whitening areas with a clear zone on either side of the vessels (within 50 mm) [3,4]. On the other hand, cotton-wool spots have ill-defined edges, and do not respect the clear zone in the proximity of vessels, being located superficially over vessels **(Figure 1)**.

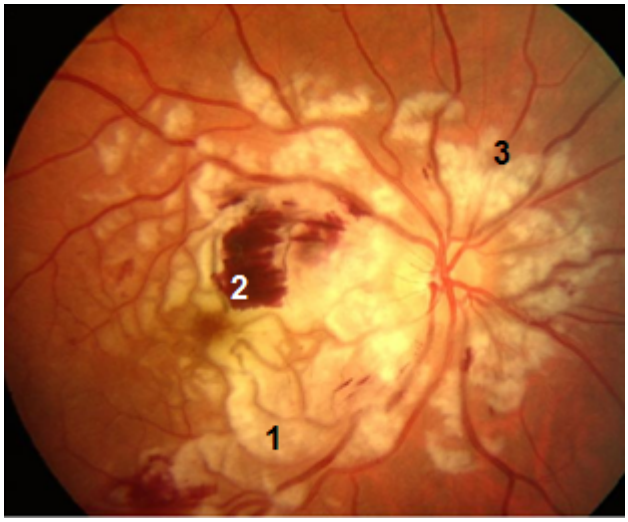

Figure 1 Purtscher's retinopathy. All of the 3 criteria of Purtscher retinopathy that are visible in a fundoscopic examination are present: 1) Purtscher flecken (with a clear space between vessels and with well-defined edges), 2) Retinal hemorrhages, 3) Cotton wool spots (that are located superficially to vessels and with poorly defined edges).

Complementary examination

Fluorescein angiography [1,3]

This exam usually shows areas of non-perfusion and retinal ischemia (present in 70% of the angiographies that were performed [3]). Other alterations identified were: early hypo-fluorescence, a delay in filling of vessels, late leakage and peripapillary staining.

Ocular coherence tomography (OCT) [1,3,26,27,32]

Retinal edema is frequently identified. Within 1-6 months, there is either normalization or macular atrophy (the latter is more probable when severe alterations of the macula are visible at presentation).

Visual field tests

They are not frequently performed; alterations include: central scotoma, nasal step and arcuate scotoma. When there is a visual field defect at presentation, approximately half (55%) become a persistent [3].

Visual evoked potentials

Might show increased latency and decreased amplitude of responses [3].

Electroretinogram

It may demonstrate depression of the a and b waves, with partial amelioration throughout time in the majority of cases [3].

Treatment

First, conservative treatment should be performed according with the etiology.

Second, many doctors add intra-venous corticosteroids in high doses but there is no clear evidence regarding whether they are beneficial of not [1-3,33]. Corticosteroids might be useful in accelerating visual acuity recovery such as previously reported [2,34] due to their ability to stabilize damaged neuronal membrane and microvascular channels, and to inhibit granulocyte aggregation related to complement activation [7,9]. However, the risk of adverse drug effects should be taken into account [3].

More recently, it has been postulated that since many Purtscher-associated etiologies were recognized precipitants of thrombotic microangiopathy [35], research should be aimed at the identification of molecules in physiological and pathological cascades of thrombotic microangiopathy to identify new therapeutic targets for patients with Purtscher retinopathies [36].

There are also recent clinical cases in which anti-vascular endothelial growth factor (anti-VEGF) agents were administered [37], and patients improved. Anti-VEGF agents may be useful theoretically since they may counteract endothelial dysregulation and pathological retinal microvascular permeability [38]. However, since spontaneous improvement is frequent, we cannot yet conclude of its utility and further evidence is needed.

The same caution applies to a recent case report of a Purtscher-like retinopathy following dacryocystorhinostomy [26], which was treated with intra-vitreal tissue plasminogen activator.

Prognostic and visual acuity improvement

It has been reported that presence of a Purtscher-like retinopathy in patients with severe acute necrotizing pancreatitis is a poor prognostic factor for death10. In our study, all of the 5 deaths occurred in patients with Purtscher-like retinopathies (thrombotic thrombocytopenic purpura, cryoglobulinemia with hepatitis C, lupus, pancreatic carcinoma and necrotizing vasculitis in lung carcinoma) [3].

As for visual acuity improvement, poor prognostic factors [1,39] are optic disk swelling, leakage seen on angiography, choroidal hypoperfusion, involvement of the outer retina and retinal capillary non-perfusion. We also identified macular edema and pseudo-cherry red spot as statistically significant poor prognostic factors [3,39]. Additionally, Holak et al. [28] stated that, although those prognostic factors are important, the duration of retinal changes is decisive for late prognosis. Male gender might be a good prognostic factor [3] for visual acuity improvement, as well as trauma and acute pancreatitis (in comparison with other etiologies for Purtscher like retinopathies). Male gender is associated with traumatic etiology, thus it is unclear whether it has a good prognostic value itself.

Conclusion

In conclusion, Purtscher and Purtscher-like retinopathies easy to diagnose once the diagnostic criteria are respected (Purtscher-flecken, intra-retinal hemorrhages, cotton wool spots, plausible etiology, compatible complementary examination).

Good prognosis factors are male gender and traumatic etiology. Poor prognostic factors are related to the severity and duration of retinal changes.

Supportive treatment should be performed as well as treatment of the underlying etiology. Other treatments, such as corticosteroids, intra-vitreal injection of anti-VEGT and intra-vitreal injection of tissue plasminogen activator, are theoretically logical but further studies are necessary to verify whether they are beneficial in comparison to conservative treatment.

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