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Case Report

Henoch-Schönlein Purpura with Ocular Manifestations in Adult: Case Study Report and Review of the Literature

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Abstract

Henoch Schönlein purpura is a systemic vasculitis that affects small vessels on children mostly. Thrombotic events are a rare complication, and only a few cases have been reported. Among those, a case of bilateral central retinal arterial occlusion and a case of central retinal vein occlusion over a two-week-long period of hemodialysis, both of them previously diagnosed with Henoch Schönlein purpura. In this paper, we describe the case of an adult patient diagnosed with Henoch Schönlein purpura and arterial hypertension, chronic renal failure in hemodialysis for 20 days; who presents decreased visual acuity for 15 days in the right eye. The findings were disc edema, macular edema, vascular tortuosity, and hemorrhages in four quadrants. Central retinal vein occlusion was considered and antiangiogenic therapy and laser photocoagulation were performed with positive response.

Keywords: *Central Retinal Vein Occlusion; Henoch Schönlein Purpura; Hemodialysis; Central Retinal Vein Thrombosis*

Introduction

Henoch Schönlein purpura is a systemic vasculitis that affects small vessels on children mostly. Thrombotic events are a rare complication, and only a few cases have been reported.

Case Presentation

A 34-year-old male patient, diagnosed with Henoch Schönlein purpura (HSP) by clinical history, arterial hypertension for four years and chronic renal failure for one year and a half. The patient has been receiving dialysis for 20 days under pharmacological treatment with calcium, phosphorus, Clonidine, Losartan, B complex y Omeprazole., resorts to medical check-up due to a 15-day-long right-eye decrease in visual acuity.

Ophthalmology test: Visual acuity better corrected on the right eye (RE) hand movements and left eye (LE) 20/20. In the biomicroscopy it was evidenced that the anterior segment was healthy in both eyes. The intraocular pressure with Goldman´s tonometer showed RE 12 mmHg LE 13 mmHg. In the indirect ophthalmoscopy on the right eye the optical nerve presents edema, cupping 0.4, severe macular edema with tortuous dilation of the venous system, flame-shaped hemorrhage in four quadrants. LE: with changes of hypertensive re-

tinopathy. It is diagnosed with occlusion of the retina central vein with secondary macular edema on the RE and monthly intravitreous antiangiogenic therapy with Aflibercept was prescribed for a 3-month-long period. In fluorescein angiography multiple nonperfusion areas were evidenced, therefore, Argon laser photocoagulation was indicated. Through periodic monitoring intraocular pressure on the RE is found (27 mmHg), Rubeosis Iridis, and vitreous hemorrhage at fundoscopy. Antiangiogenic therapy is started, ocular hypotensor, panretinal photocoagulation with involution of the neovessels and adequate control of intraocular pressure under monitoring.

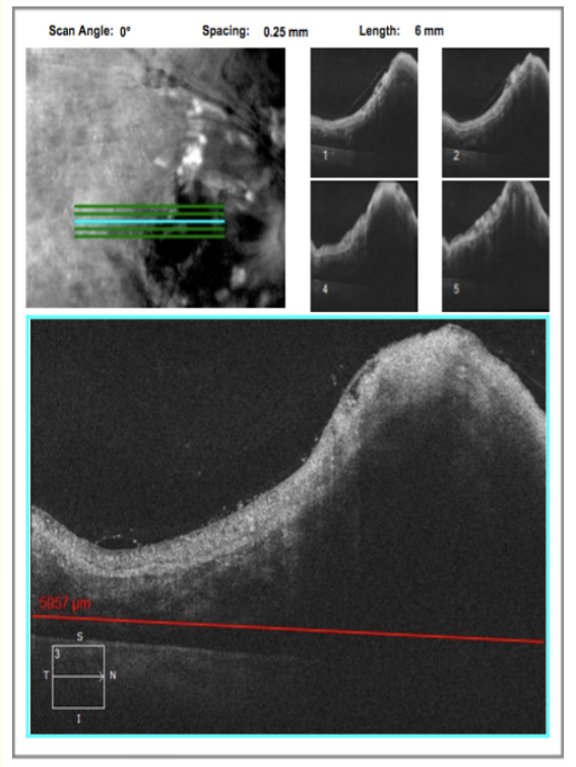

Figure 1: *Tomography of RE Macula optical coherence where secondary macular edema at CRVO is evidenced. Source: Bernardo Quijano, Oftalmocenter.*

Discussion

Henoch Schönlein purpura (HSP) is one of the most common types of vasculitis of small vessels during infancy [1]. It has an incidence of 3 to 26 out of 100.000 children, mainly among 4 - 7 years of age. In adults, it represents 0.1 to 0.8 out of 100.000 individuals [2,3]. It was described for the first time by William Haberden in 1801 who reported two cases of which he denominated "Purpura Maculae" due to the purpuric lesions on the skin found among his patients. It was not until 1837 that Johann Lukas Schönlein denominated "rheumatic peliosis" to the combination of arthralgia and arthritis with the macular rash setting the difference of what was known as hemorrhagic purpura until then, given that it did not present the hemorrhagic components of the mucosa and the skin. In 1874 Eduard Heinrich Henoch described 4 cases on children and asserted that not every case was of a self-limited nature and it was also associated with renal illness and could even cause death [4]. It constitutes a syndrome of autoimmune etiology that is characterized by deposits of immune complexes IgA and usually IgG and C3 in the small blood vessels that affect multiple organs and systems. The increase in the IgA synthesis relates

to the exposure to an antigen processed in the mucosae. It is not clear whether it is a bacteria, virus or parasite in a genetically susceptible individual. The role of genetic factors has been supported by the identification of genes of susceptibility in the human leukocytary antigen (HLA). HLA DRB1 [5-7]. Frequently, it is presented after an infection of the superior respiratory system; clinically, it is evidenced by non-thrombocytopenic palpable purpura that mainly affect inferior limbs, arthralgia or arthritis, abdominal pain, vomit, intestinal hemorrhages, and a third part of the patients presented renal manifestations that consist of macro/microscopic hematuria, proteinuria and nephrotic syndrome [7,8].

In most cases, the course of the illness is self-limited, nonetheless, the degree of renal manifestation modifies the prognosis from benign with reappearance of hematuria years after the initial case to chronic renal illness and death. The acute phase settles spontaneously in the 94% of children and 89% of adults and only requires symptomatic management. The use of therapies such as corticosteroids, immunomodulators and plasmapheresis is considered according to the degree of kidney and pulmonary compromise [8,9].

Associated ocular alterations

Within the ocular manifestations associated to Henoch Schönlein purpura, Cyril Barton in 1976 described a case of central retinal vein occlusion (CRVO) in a 12-year-old male patient in maintenance dialysis whose base illness was HSP, presenting the episode 36 hours after the session [10], Yamabe H described a case of HSP in 1988 associated with keratitis and anterior granulomatous uveitis [11]. M Muquit., *et al.* [12] reported a case in 2005 of a patient with keratitis with a superficial epithelial defect a month after the disappearance of the purpuric symptoms that was resolved adequately with conventional treatment, one month after that, the patient presented bilateral anterior granulomatous uveitis with trabeculitis responded to a treatment with corticoids; other differential diagnoses were discarded. In 2005, Chuan., *et al*. [13] reported a case of non-arteritic anterior ischemic optic neuropathy in a 54-year-old man, with diabetes mellitus type 2, diagnosed with HSP 18 months before, for which he received a treatment with steroids that was suspended due to bad glycemic control and manifested exacerbation of the purpura symptoms two weeks before the ocular symptoms. Wu T-T., *et al*. [14] published a case in 2002 in which a 6-year-old girl presented HSP along with a bilateral obstruction of the retina central artery; she was given a diagnosis of HSP with compatible kidney biopsy, systemic therapy was initiated and required hemodialysis due to acute kidney failure.

On the other hand, it is relevant to mention the risk of retina venous occlusion posed by Chang., *et al.* published in 2016 who found in their study that patients in the final stage of the chronic kidney illness in dialysis present a Hazard Ratio HR 3.05 with retina vein occlusion (RVO), possibly due to coexistence of various factors in the physiopathology of the two entities, such as: severe alteration in the microcirculation, atherosclerosis and coagulation disorders associated with platelet dysfunction; on the second hand, they share systemic risk factors such as arterial hypertension, mellitus diabetes, hyperlipidemia, coronary disease and congestive cardiac failure [15].

Our case deals with an adult patient with Henoch Schönlein purpura unlike most cases which are in children. This patient dealt with chronic kidney insufficiency for more than a year and initiated hemodialysis therapy and five days later presented right-eye decrease in visual acuity, diagnosed with intravitreous antiangiogenic therapy and panretinal photocoagulation that correlates with the treatment received in other cases reported in the literature.

Conclusion

The retina vascular occlusion is an ocular manifestation described in hemodialysis patients that causes ocular morbidity and alteration in their quality of life. Our case describes a patient with Henoch Schönlein Purpura who presents an occlusion of the central vein of the retina few weeks after initiating hemodialysis; a manifestation of the base illness is not discarded.

Ethical Responsibilities

- Human Protection: Authors to this paper declare that no human experiments were carried out for this research.
- Data Confidentiality: Authors to this paper declare that no personal information from patients appear in this report.
- Right to Privacy and Informed Consent: Authors to this paper declare that no personal information from patients appear in this report.

Conflicts of Interest

Authors to this paper declare having no interest conflicts.

Bibliography

1. Penny K., *et al*. "An epidemiological study of Henoch-Schönlein purpura: Kay Penny and colleagues suggest nurses need to be aware of the signs and symptoms, and treatments that are available to better understand this disease". *Pediatric Care* 22.10 (2010): 30-35.
2. Gardner-Medwin JM., *et al*. "Incidence of Henoch-Schonlein purpura, Kawasaki disease, and rare vasculitides in children of different ethnic origins". *The Lancet* 360.9341 (2002): 1197-202.
3. Watts RA., *et al*. "What is known about the epidemiology of the vasculitides?" *Best Practice and Research Clinical Rheumatology* 19.2 (2005): 191-207.
4. Matteson EL. "Notes on the history of eponymic idiopathic vasculitis: the diseases of Henoch and Schönlein, Wegener, Churg and Strauss, Horton, Takayasu, Behçet, and Kawasaki". *Arthritis Care and Research* 13.4 (2000): 237-245.
5. Weiss PF., *et al*. "Temporal Association of Streptococcus, Staphylococcus, and Parainfluenza Pediatric Hospitalizations and Hospitalized Cases of Henoch-Schonlein Purpura". *The Journal of Rheumatology* 37.12 (2010): 2587-2594.
6. Audemard-Verger A., *et al*. "IgA vasculitis (Henoch–Shönlein purpura) in adults: Diagnostic and therapeutic aspects". *Autoimmunity Reviews* 14.7 (2015): 579-585.
7. Helander SD., *et al*. "Henoch-Schönlein purpura: clinicopathologic correlation of cutaneous vascular IgA deposits and the relationship to leukocytoclastic vasculitis". *Acta Dermato-Venereologica* 75.2 (1995): 125-129.
8. Yang Y-H., *et al*. "The diagnosis and classification of Henoch–Schönlein purpura: An updated review". *Autoimmunity Reviews* 13.4-5 (2014): 355-358.
9. Roberts PF., *et al*. "Henoch-Schönlein Purpura: A Review Article". *Southern Medical Journal* 100.8 (2007): 821-824.
10. Barton CH and Vaziri ND. "Central retinal vein occlusion associated with hemodialysis". *The American Journal of the Medical Sciences* 277.1 (1979): 39-47.
11. Yamabe H., *et al*. "IgA nephropathy and Henoch-Schönlein purpura nephritis with anterior uveitis". *Nephron* 50.4 (1988): 368-370.
12. Muqit MMK. "Henoch-Schonlein purpura with keratitis and granulomatous anterior uveitis". *British Journal of Ophthalmology* 89.9 (2005): 1221-1222.
13. Chuah J and Meaney T. "Anterior ischaemic optic neuropathy secondary to Henoch–Schönlein Purpura". *Eye* 19.9 (2005): 1028-1028.
14. Wu T-T., *et al*. "Henoch-Schonlein purpura with bilateral central retinal artery occlusion". *British Journal of Ophthalmology* 86.3 (2002): 351-352.
15. Chang Y-S., *et al*. "Risk of Retinal Vein Occlusion Following End-Stage Renal Disease". *Medicine (Baltimore)* 95.16 (2016): e3474.

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