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CASE REPORT

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Bilateral vision loss as first clinical manifestation of IgA nephropathy

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Abstract

Background Immunoglobulin A nephropathy (IgAN) is the most common primary glomerular disease, characterized by IgA deposition in the renal glomeruli. Its clinical manifestations range from asymptomatic hematuria to progressive chronic kidney disease (CKD), with rare cases presenting as nephrotic syndrome or rapidly progressive glomerulonephritis. The eye involvement could be present with a various spectrum of clinical presentation, unusually as first presentation and isolated.

Case presentation A 30-year-old male presented to the emergency department with rapidly progressive bilateral visual loss over two days. He had a history of mild upper respiratory symptoms treated with ibuprofen and a known history of asthma. Ophthalmologic evaluation revealed significant retinal findings, including hemorrhages, cotton wool spots, macular edema, and neurosensory detachment. His blood pressure was markedly elevated at 200/140 mmHg, prompting systemic evaluation. Laboratory tests revealed acute kidney injury with elevated creatinine and proteinuria. Renal ultrasound indicated cortical hyperechogenicity, and a renal biopsy confirmed IgAN with mesangial IgA deposition. After antihypertensive treatment and stabilization of renal function, the patient's vision and retinal findings improved significantly over three weeks.

Discussion This case illustrates an uncommon presentation of IgAN with acute hypertensive crisis manifesting initially only as bilateral visual impairment. The retinal findings, including Roth spots and choroidal involvement, highlight the importance of ophthalmic evaluation in systemic diseases. Although hypertensive retinopathy is well documented, its association with IgAN is rarely reported in literature, especially as first manifestation.

Conclusions IgAN can present atypically with isolated ocular symptoms, underscoring the need for a comprehensive multidisciplinary approach. Clinicians should consider systemic evaluation in patients with hypertensive retinopathy and acute visual loss to facilitate early diagnosis and management of underlying renal pathology.

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Background

Immunoglobulin A nephropathy (IgAN), also known as Berger's disease, is the most common primary glomerular disease globally, characterized by the deposition of immunoglobulin A (IgA) in the glomeruli of the kidneys [1]. This condition has different clinical manifestations. The most frequent clinical presentation is asymptomatic hematuria, followed by progressive chronic kidney disease (CKD). Rare presentations include IgAN associated with nephrotic syndrome and IgAN with rapidly progressive course. In 10% of cases IgAN is associated with a synpharyngitic macroscopic hematuria [2–4].

The epidemiology of IgAN reveals a notable geographic variation in its prevalence. It is more frequently diagnosed in Asian populations compared to Caucasians, which can be attributed to differences in healthcare practices, such as the routine screening of urine in some Asian countries and varying thresholds for performing kidney biopsies [2]. The pathogenesis of IgAN is complex and multifactorial, involving genetic, immunological, and environmental factors. The "four-hit hypothesis" is widely accepted to explain the disease mechanism, which includes the production of galactose-deficient IgA1 (Gd-IgA1), the formation of autoantibodies against Gd-IgA1, the creation of immune complexes, and their deposition in the glomerular mesangium, leading to inflammation and renal injury [3, 5, 6].

Recent advances in molecular and clinical research have provided deeper insights into the immunopathogenesis of IgAN. Key molecules such as Gd-IgA1, IgG anti-Gd-IgA1 antibodies, and soluble CD89 have been identified as critical players in the formation of pathogenic immune complexes [5]. Additionally, the role of the complement system, particularly the alternative and lectin pathways, has been highlighted in exacerbating the disease [7]. Genetic studies have also pointed to the involvement of specific genetic factors that promote the overproduction of aberrant IgA1, further contributing to disease development [1, 8, 9].

Despite these advancements, there is still no disease-specific treatment for IgAN. Current therapeutic strategies focus on managing symptoms and slowing disease progression. However, emerging treatments, including monoclonal antibodies targeting the immunopathogenic pathways of IgAN, offer hope for more effective and targeted therapies in the future [10, 11].

The purpose of this case report is to present an unusual clinical manifestation characterized exclusively by visual symptoms in a patient with no previous medical history, who was subsequently diagnosed with IgA nephropathy. Moreover, we show a complete multimodal imaging of the retinal clinical features.

The case

A 30-year-old male who presented to the emergency department with a progressive bilateral visual loss that had worsened over a period of two days. In the week preceding this event, the patient reported experiencing a persistent cough, sore throat, and mild fever, treated with ibuprofen 200 mg/day for 5 days with relief. Past medical history was positive for asthma, treated chronically with salbutamol 1spray/day. Ophthalmological history was negative and no familiarity for ophthalmological diseases was reported.

No other symptoms except for the visual loss were reported by the patient at the admission. The patient was addressed directly to the ophthalmology department for assessment of the visual loss through the fast-track service.

The ophthalmic evaluation showed: a best corrected visual acuity (BCVA) of 20/50 in the right eye and of 20/200 in the left eye, no sign of pupillary defects, no restriction in eye movements or pain, and the anterior segment exam was unremarkable.

After dilatation with tropicamide 1% eye drops fundoscopy was performed and showed: in the right eye several retinal hemorrhages at the posterior pole, cotton wool spots, pale/yellow spots of the choroid, two Roth spots, macular edema, and multiple neurosensory detachments in middle periphery; in the left eye peripapillary hemorrhage, partial disc swelling on the nasal side, cotton wool spots, pale/yellow spots of the choroid, macular edema, and multiple neurosensory detachments in middle periphery.

An ultra-wide field retinography using Clarus 500 (Carl Zeiss Meditec, Germany) was performed for a comprehensive evaluation of the retina (Figs. 1 and 2).

An ultra-wide field OCT using OCT-S1Xephilo (Canon, Japan) was performed to assess the macular edema (Figs. 3 and 4), the detachments of the neurosensory retina and retinal pigmented epithelium (RPE) alterations (Fig. 5).

Due to the retinal findings, patient's blood pressure was measured and found to be 200/140 mmHg, indicating a hypertensive crisis. Given the severity of the hypertension and the negative anamnesis, a comprehensive systemic workup was carried out.

Routine blood tests revealed significant abnormalities: most notably an elevated serum creatinine level of 2.97 mg/dL (reference range: 0.6–1.2 mg/dL), suggestive of impaired renal function, elevated serum urea level of 77 (reference range: 12–48 mg/dL), reduced hemoglobin level of 12.6 (reference range: 13–17.5 g/dL) and reduced hematocrit (reference range: 42.0–54.0).

A therapy with labetalol in bolus was started and due to poor response switched to continue infusion of urapidil

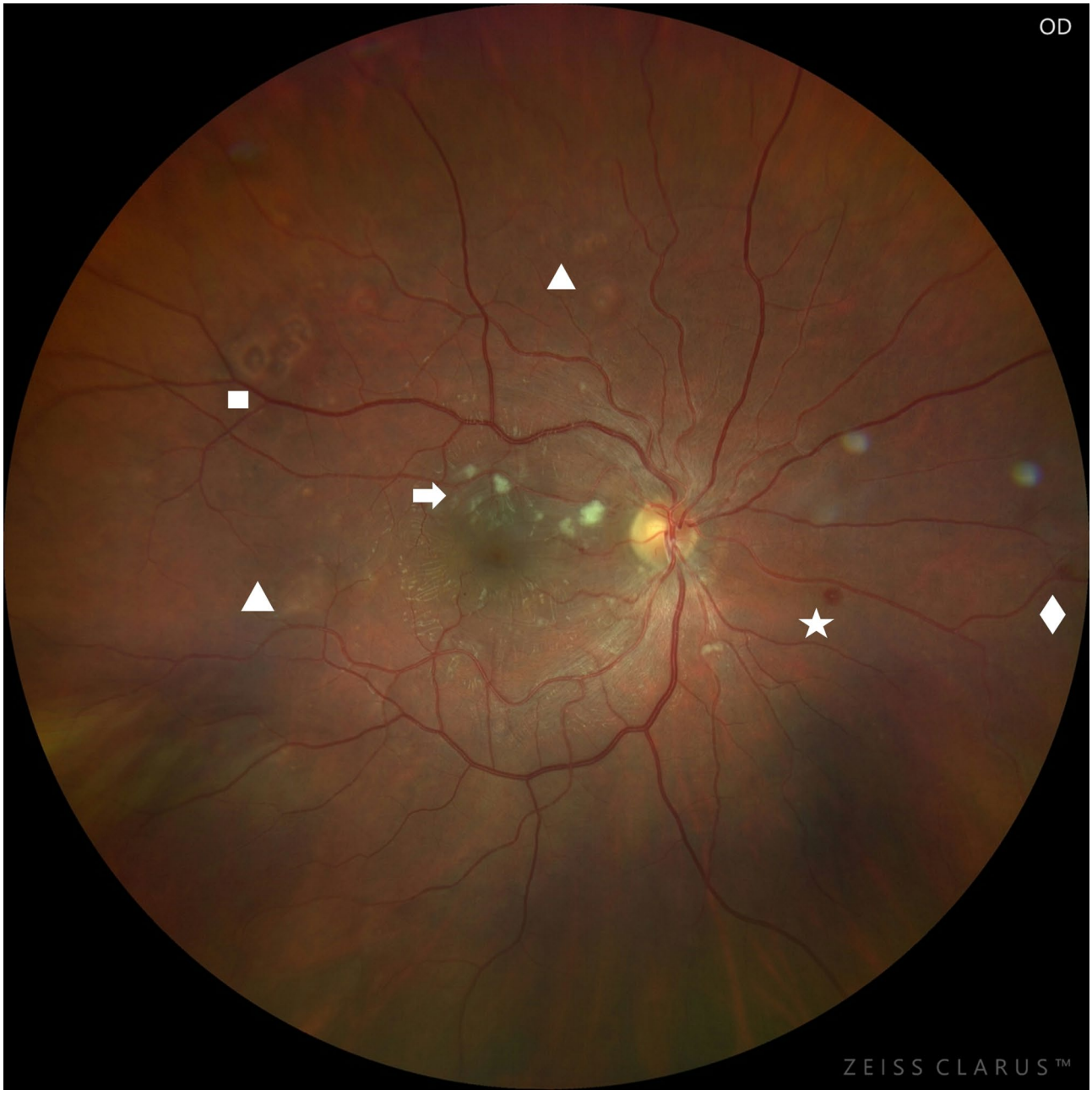

Fig. 1 Retinography of the right eye showing hemorrhage (rhombus), Roth spot (star), cotton wool spots (arrow), macular edema, pale/yellow spot of the choroid (triangle) and neurosensory detachments (square)

5 ml/h and oral therapy of doxazosin 2 mg. After 8 h the blood pressure was reduced to 150/100 mmHg.

The patient was then referred to Nephrology to proceed with further diagnostic investigations, hospitalization and therapy. Renal ultrasound and additional biochemical assessments were recommended to determine the etiology of the acute renal dysfunction.

Renal ultrasound showed hyper-echogenicity of the cortex and reduced corticomedullary differentiation.

Serologic tests were performed and tested negative for infectious diseases. Serum testing for immunoglobulins showed elevated IgA level of 456 (reference range: 70–400 mg/dL). Urine analysis showed high levels protein of 300 (reference range: 0 mg/dL) and presence of microhematuria.

During the hospitalization the patient underwent a renal biopsy to determine the cause of renal failure. Microscopic exam showed a high number of glomeruli with segmental focal glomerulosclerosis, focal

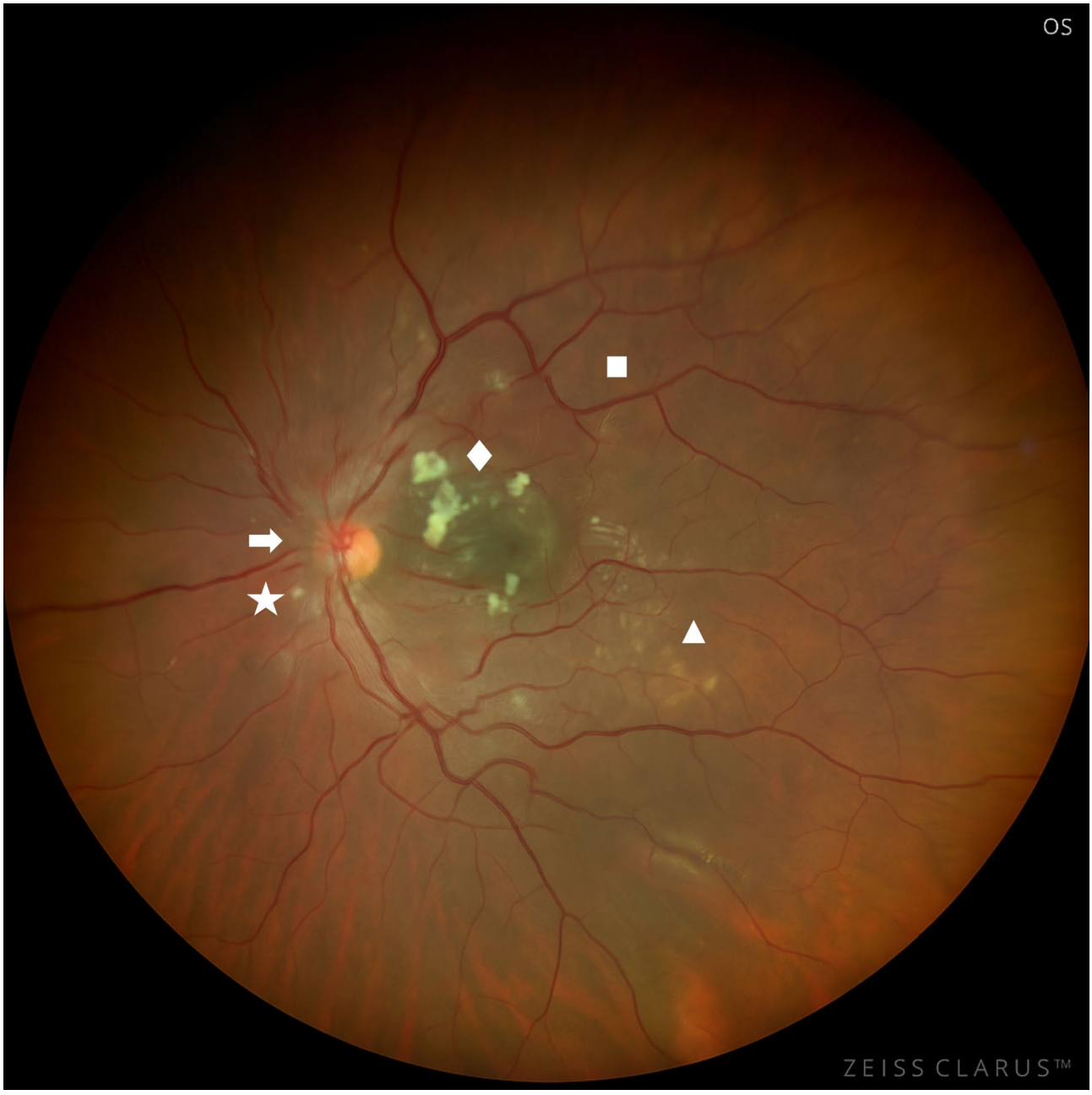

Fig. 2 Retinography of the left eye showing peripapillary hemorrhage (star), partial disc swelling on the nasal side (arrow), cotton wool spots (rhombus), pale/yellow spots of the choroid (triangle), macular edema, and multiple neurosensory detachments in middle periphery (square)

augmentation of the matrix and cellularity of the mesangial space, with associated basal membranes splitting; no signs of necrosis or crescents and focal signs of endocapillary hypercellularity were reported; mild tubular atrophy was observed. Direct immunofluorescence exam showed at the mesangial deposit on basal membrane of IgA, C3, k e λ chains. Considering the Oxford classification system 2016 the report was defined as M0 E1 S1 T1C0.

All the clinical data and the results of the biopsy, leaned to a diagnosis of IgA nephropathy.

After 2 days from the admission, the patient underwent a fluorescein angiography that showed posterior pole diffuse leakage, signal blockage of the cotton wool spots and late phase subretinal leakage in the neurosensory detachments. No signs of retinal ischemia were found (Fig. 6).

After the stabilization of the blood pressure level and the normalization of the creatinine blood levels, the patient was discharged with domiciliary therapy and

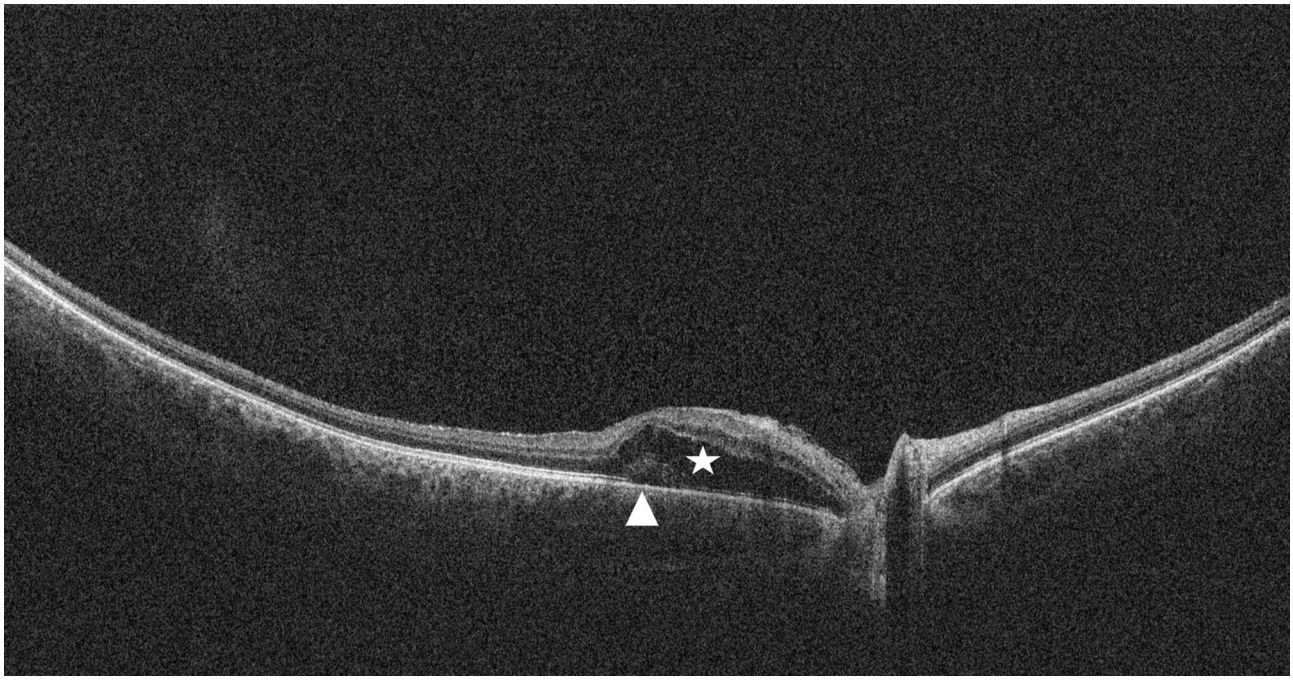

Fig. 3 OCT scan of the macula of the right eye showing macular edema with intraretinal cysts (star) and neurosensory detachment of the foveola (triangle)

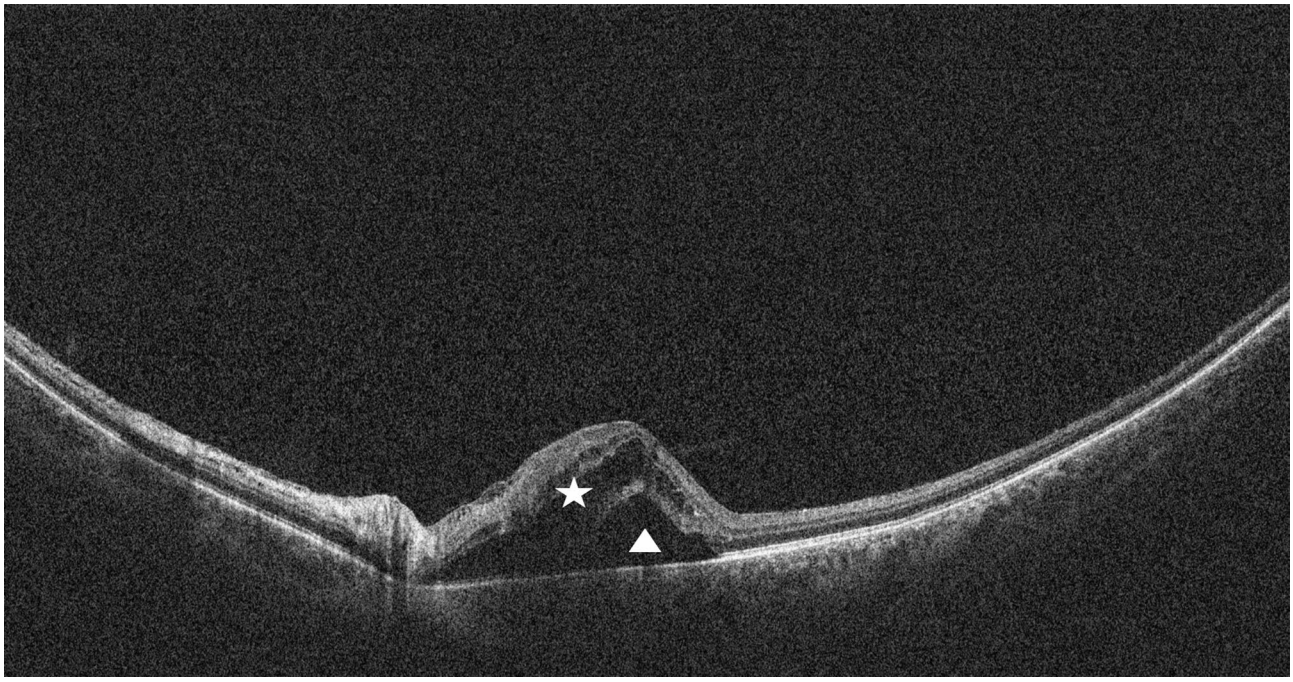

Fig. 4 OCT scan of the macula of the left eye showing macular edema with intraretinal cysts (star) and a big neurosensory detachment of the foveola (triangle)

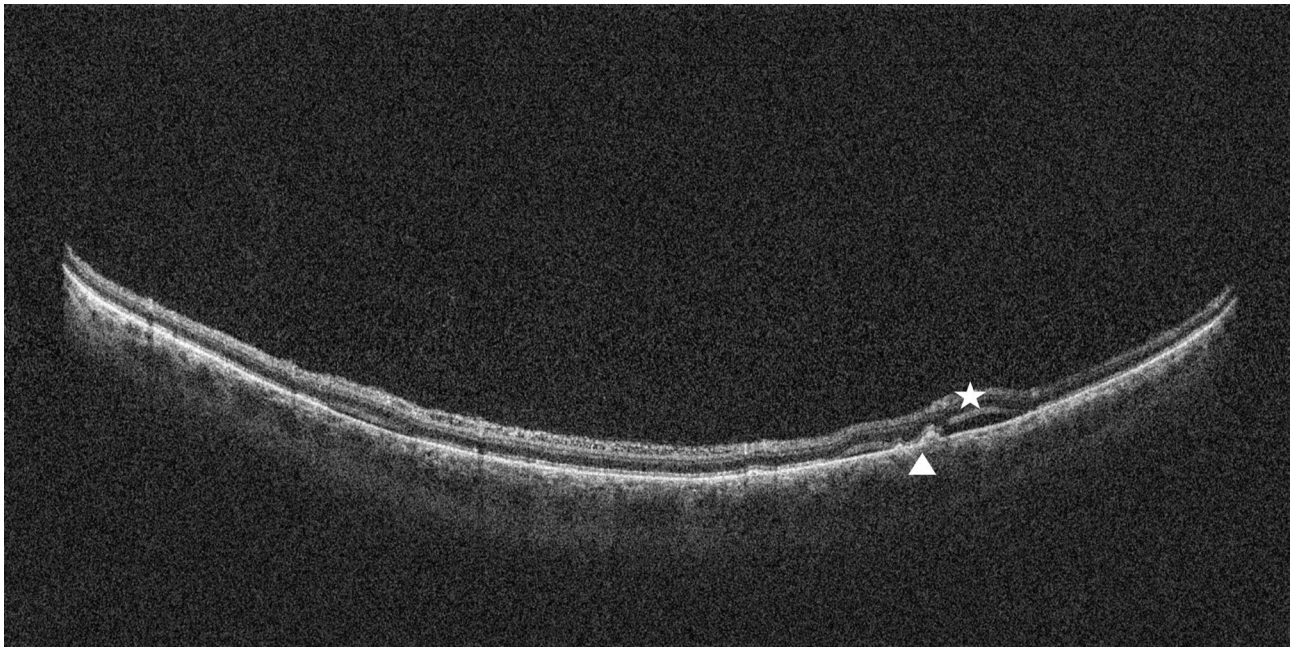

Fig. 5 Wide field OCT of the left eye centered on the super-temporal periphery showing the neurosensory detachment (star) overlying a little sub-RPE deposit (triangle)

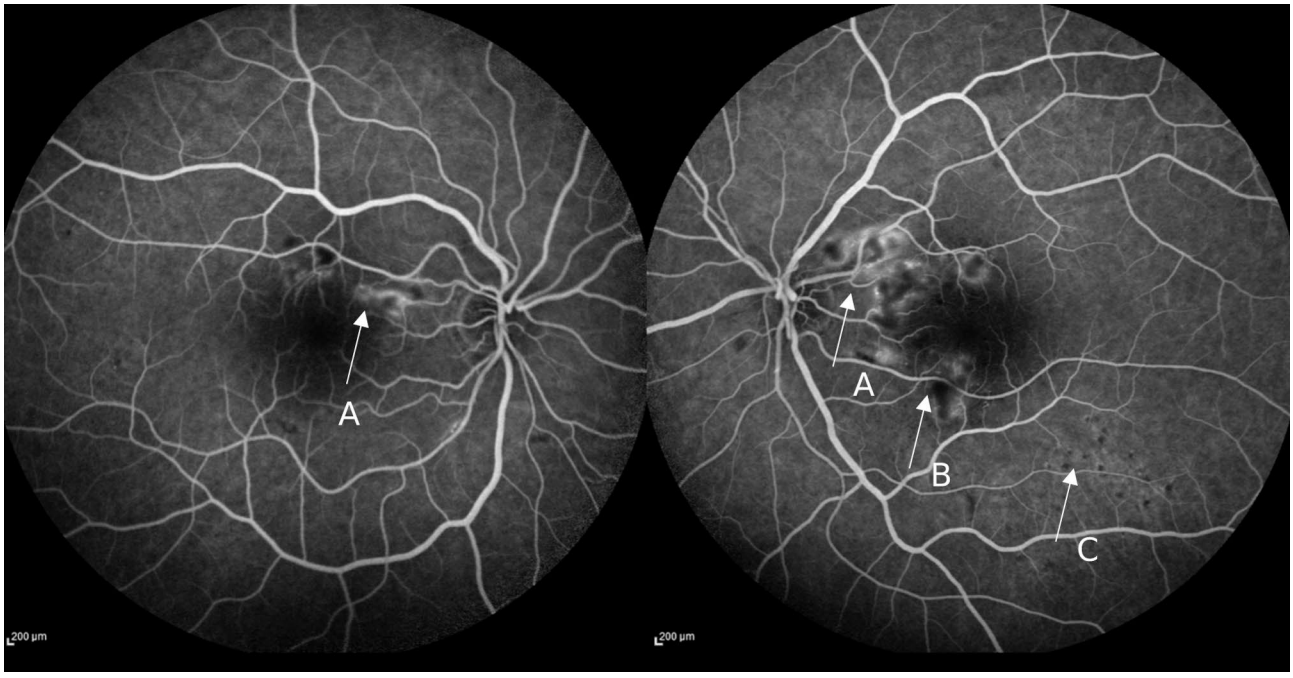

Fig. 6 Fluorescein angiography of the right and left eye, showing no signs of retinal ischemia, multiple points of fluorescein leakage(arrow A) and hypo-fluorescence in the areas of hard exudates (arrow B) and Elschnig spot (arrow C)

referred to the nephrology and ophthalmology outreach services.

After three weeks from the admission, the patient BCVA was 20/20 in both eyes and the fundus examination showed no macular edema, remission of most retinal hemorrhages and cotton wool spots, and complete absorption of neuroepithelium and RPE detachments.

Discussion

IgAN is the most frequent glomerulonephritis in the world and is commonly suspected and diagnosed after routine blood and urine test for screening purpose. Less frequently is associated with macrohematuria and fever, with or without upper high-airways syndrome [4].

In our case, the patient presented to the ER complaining only for a bilateral visual loss, with no other associated systemic signs or symptoms.

A similar presentation with ocular involvement as the first clinical manifestation is only described in a German case report not available in English. In this case report the authors found a patient that showed to emergency department complaining only of bilateral blurred vision. On fundus examination they found hemorrhages, cotton wool spots, macular edema, disc edema and peripapillary hemorrhages. No sign of Elschnig spots, Roth spot and RPE detachments were reported. Non mydriatic retinography and macular OCT were used to evaluate the patient [12].

Kwan JT et al. reported the case of a patient with ocular symptoms as first presentation of IgAN, although in their case the predominant feature was papilledema in the setting of a raised intracranial pressure [13].

Some other case reports have shown ocular clinical findings like those showed in our case but associated to a long history of kidney impairment from chronic IgA nephropathy and not an acute episode [14–17].

Moreover, in our report the patient underwent a complete ophthalmic evaluation that included multimodal imaging of the retina using ultra-wide field technology.

The retinal findings of our case are characterized by hemorrhages, Roth spot, cotton wool spots, macular edema and neuroretina and RPE detachments.

Retinal hemorrhages indicate a rupture of the inner blood retinal barrier as the consequence of vessels wall damage from sustained high blood pressure, permeating the leakage of erythrocytes to the inner retinal layers, some of them deep that are round and dark, and others superficial following the path of the nerve fibers layer that are lighter and flame shaped. These findings are frequent in the case of hypertensive crisis [18].

Roth spot is a flame shaped, round or oval retinal hemorrhage with a white center and are associated with various systemic illnesses. The white center is a fibrin thrombus with platelet aggregates, surrounded by red blood cells diffused in the inner retinal layers. This kind of hemorrhage is sign of a previous vessel rupture, and in our case, this could be caused by the elevated blood pressure of the patient. This finding has been reported in other cases of hypertensive retinopathy, but it's not usual [19]; moreover, this is the first time that such feature is described in the setting of a hypertensive crisis secondary to IgAN.

Cotton wools spots are focal signs of retinal infarction at the level of the nerve fiber layer as the consequence of the severe constriction of arterioles of the retina in response to the severely augmented blood pressure, accompanied by damage of their endothelium. The findings are also typical of hypertensive retinopathy [18].

The hypertensive choroidopathy is a known clinical entity during a hypertensive crisis and could be associated with the hypertensive retinopathy. Neurosensory detachments are signs of choroidal involvement and represent a sign of focal RPE pump failure due to choroidal focal infarction caused by elevated blood pressure and consequent constriction of the arterioles. The choroid pale/yellow spots are a sign that is usually found during hypertensive crisis in young patients, and there are known as Elschnig spots. These spots represent focal ischemia of the RPE in the non-perfused area of choriocapillaris. The Elschnig spots may appear on the OCT scans like small RPE detachments, occasionally overlaid by neurosensory detachments. Choroidal involvement is usually associated with a poor visual prognosis if left untreated [20].

Macular edema could be explained by the combination of the loss of function of the RPE and the rupture of the inner blood-retinal barrier, causing a loss of homeostasis in fluid entry and exit [21].

The optic disc edema swelling and the peripapillary hemorrhages are sign of optic disc involvement. The swelling of the optic disc is due to choroidal vessels involvement or to an increase in intracranial pressure. There are few published reports that communicate the concomitant presence of hypertensive retinopathy with disc swelling and augmented intracranial pressure in the setting of a malignant hypertension [13]. A characteristic finding in those cases is the bilateral swelling of the optic disc. In our case the swelling and the peripapillary hemorrhages were present only in the left eye. Additionally, no signs of raised intracranial pressure were found on cerebral imaging. In our case, we think that the optic disc swelling could be secondary to involvement and infarction of the peripapillary and choroidal vessels that supply the optic disc. Moreover, the unilateral disc involvement could explain the greater reduction in visual acuity in the left eye.

Conclusions

We presented the multimodal imaging findings of an uncommon clinical presentation of IgAN with acute renal failure and hypertensive crisis first manifesting with visual symptoms. Systemic disease could manifest only with ocular involvement as first manifestation and clinicians must be aware that a comprehensive general evaluation must be performed even in the case of isolated visual symptoms. A multidisciplinary approach is mandatory to treat these cases in order to preserve the renal and visual function.

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EP, MMAM and SD provided care to the patient and contributed to the writing of the manuscript. MMAZ contributed to the writing of the paper and provided internal review process.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
No ethical committee approval was necessary for case report.

Consent for publication
Written informed consent was obtained from the patient for publication of the case report.

Competing interests
The authors declare no competing interests.

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