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HENOCH-SCHONLEIN PURPLE DIAGNOSIS BY RENAL BIOPSY: CASE REPORT

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Abstract
Henoch-Schönlein Purpura is an inflammatory disease characterized by deposits of immunoglobulin A (IgA) in the walls of blood vessels, producing a series of symptoms that mainly affect the skin, joints, gastrointestinal system and kidneys. The exact cause is not completely known, but it is believed that there is an autoimmune component as well as a genetic predisposition. It affects children more frequently than adults. Larger studies are required to evaluate the effectiveness of corticosteroids and possibly develop an approach that includes guidelines for symptomatic treatment, immunosuppression, and immunomodulation.
Objective: To present a clinical case of a patient with Henoch-Schonlein purpura diagnosed by renal biopsy.
Methodology: This paper is based on a case report, the discussion of which will be complemented by a selection of relevant literature on the subject. This selection will be carried out through scientific search platforms such as PubMed, Springer, Elsevier and Scopus. To refine and broaden the search, bibliographic descriptors, such as DeCs/MeSH, as well as Boolean operators, such as "NOT," "OR," and "AND," in English and Spanish, will be used. Articles published in the period between 2017 and 2023 that have access to the full text will be considered.
Results: We describe the case of a 59-year-old female patient who presented with respiratory distress, abdominal pain, purpura in the lower limbs, and headache. After diagnostic testing, significant findings were identified including hematuria, proteinuria, and the presence of IgA deposits with a granular and diffuse pattern on a renal biopsy. As a result of these findings, he was diagnosed with Henoch-Schönlein purpura, with a possible infectious origin related to pneumonitis.
Discussion: The indexed bibliography on the presumed etiological factors, clinical manifestations, diagnosis and therapeutic options is analyzed.
Conclusion: The presentation of a clinical case and exhaustive literature review on IgA vasculitis is of medical interest due to its impact on health. Early detection and proper management are crucial to address effectively, preventing kidney deterioration and improving the quality of life of those affected.

Keywords: Iga vasculitis, Henoch-Schönlein purpura, iga deposits, kidney biopsy.

INTRODUCTION

IgA vasculitis is an inflammation of small vessels related to tissue deposits of immunoglobulin A (IgA). It can present as systemic vasculitis (IgAV - Schönlein-Henoch purpura (HSP) or as a skin-only variant (skin-limited IgAV), while IgA nephropathy presents a kidney-restricted variant in the spectrum of IgA-related vasculitis. Henoch Schönlein purpura affects children more often than adults and is characterized by a combination of cutaneous vasculitis, arthritis, and gastrointestinal and renal involvement (1).

The disease was first described in 1802 by William Heberden in a case report of a five-year-old boy with hematuria, abdominal pain, purpuric rash, and vomiting. In 1837 Johann Schönlein He describes it as the association between purpura and arthralgias, and finally in 1874 Eduard Heinrish Henoch Add gastrointestinal and kidney symptoms (2).

The exact cause of IgA vasculitis is not fully understood, but it is believed that there is an autoimmune component to its development. It may be related to previous viral or bacterial infections, as well as genetic predisposition. Between 30% to 50% of cases are due to respiratory tract infections caused by Streptococcus pyogenes. The annual incidence is 3 to 26 cases per 100,000 people. This condition is most common in children, between 3 and 15 years of age, but it can also affect adults. It affects blacks less frequently compared to other groups (3).

Diagnosis is based on clinical symptoms and observation of palpable purpura on the skin. Blood and urine tests, as well as a skin or kidney biopsy, can help confirm the diagnosis and assess the severity. One of the most serious complications of Henoch-Schönlein purpura is kidney damage. Some patients develop IgA glomerulonephritis, an inflammation in the renal glomeruli that can lead to chronic kidney failure (1). Treatment of IgA vasculitis should be tailored to the severity of symptoms and the individual response of each patient. Some patients may improve without the need for specific treatment, while others may require potent immunosuppressive therapies (4).

The clinical case presentation and exhaustive literature review on IgA vasculitis is of medical interest due to its impact on patients' health and its diagnostic and therapeutic challenges. Early detection and proper management are crucial to address effectively, preventing kidney deterioration and improving the quality of life of those affected.

WHAT IS HENOCH-SCHONLEIN PURPLE?

Immunoglobin A (IgA) vasculitis, also known as Henoch-Schönlein purpura (HSP), is autoimmune, self-limited small vessel leukocytoclastic vasculitis characterized by a predominant immune deposition of IgA1 in the walls of blood vessels. Typical symptoms include palpable purpura, arthritis or arthralgia, abdominal pain, and hematuria or proteinuria (4).

EPIDEMIOLOGY

This condition occurs at any age, but is most common in children between the ages of 3 and 15. According to the study by Pillebout and Sunderkötter (1), the incidence in children is 1/4880 of children per year, being lower in adults with an index of 1/1 million. It is more common in men than in women with a ratio of 2:1 or up to 5:1 respectively. It is most prevalent during the autumn and winter seasons. It is reported in every country in the world, but its distribution is variable. It appears to be more prevalent in Japan, Southeast Asia, Europe, and Australia than in North America and South Africa. It is seen in all ethnic groups, but is rarer in black subjects (5).

ETIOLOGY AND PATHOGENESIS.

Importantly, IgA vasculitis is a complex, multifactorial disease, and no single causative factor has been identified. The interaction between genetic, environmental, and immunological factors likely contributes to the development of this disease.

Environmental Factors.
Upper respiratory tract infections usually precede most cases of Henoch-Schönlein purple (PHS). Streptococcal strains, parainfluenza virus, and more recently SARS-CoV-2 are the most commonly associated pathogens, and in children human parvovirus B19 is a common viral trigger. The interaction between leukocytes and vascular endothelial cells along with the vascular deposition of IgA1-containing immune complexes suggests that HSP is an antigen-mediated dysregulated immune response to an antigen. Through the binding and activation of complement factors, IgA cross-reacts with endothelial cells and damages the cells. The dysregulated immune response can lead to inflammation and vasculitis without a granulomatous reaction (4).

A high frequency of sinusitis and focal infections of the oral cavity was also found in children with IgA vasculitis This pathology may also be related to exposure to antigens from drugs or vaccines for measles, mumps, rubella, influenza, or hepatitis B as an immune response to a vaccine antigen as can trigger immune responses through molecular mimicry, increasing intestinal permeability and abnormal IgA1 production resulting in subsequent immune dysfunction (6). In adults, it has been linked to cancer, which may reflect an immune response elicited by the tumor. In a study conducted by Mitsui, et al (7), reported that of 53 patients with a mean age of 41.3 years with HSP, 24 had malignancies.

Genetic factors.
Certain alleles HLA-DRB1 01, HLA-DRB1 11, HLA-B35, and HLA-A11 associated with an increased risk of IgA vasculitis have been studied (4). Genetic polymorphisms have been identified in immune-related genes, such as the cytokine gene IL-1β, that may be involved in susceptibility to IgA vasculitis (8). Several recent studies established a link between IgA vasculitis and mutations in the familial Mediterranean fever (MEFV) gene. IgA vasculitis was reported in up to 7% of patients with the disease. Children with IgA vasculitis in Israel, Turkey, Iran, and China showed higher-than-expected rates of carriers of MEFV gene mutations (9).

Immunological factors.
In IgA vasculitis, there is an abnormal buildup of immunoglobulin A (IgA) in the blood vessels. IgA is an antibody that is part of the immune system and is commonly found in the mucous membranes of the body. IgA is deposited on the walls of blood vessels, triggering an inflammatory response. Abnormal IgA is thought to activate the immune system, triggering an exaggerated inflammatory response. This response includes the release of pro-inflammatory cytokines and the activation of inflammatory cells, such as neutrophils. The accumulation of IgA and the resulting inflammation can damage blood vessel walls, leading to vasculitis and its clinical manifestations, such as rash, abdominal pain, and arthritis (4).

CLINICAL MANIFESTATIONS.
Symptoms of Henoch-Schönlein purpura can vary in intensity and may include:

Skin manifestations.
The main symptom of IgA vasculitis is a palpable, round or oval, retiform purpura with a predilection for the lower legs. Caused by the destruction of vessel walls, where erythrocytes are subsequently extravasated (10). Perivascular inflammatory infiltrate and extravasate erythrocytes cause palpable lesions, while the round, oval, or especially retiform pattern derives from the anatomical arrangement of the damaged vessels. In severe cases, complete destruction of vessels or inflammation-induced intravascular thrombosis results in hemorrhagic blistering and necrosis (1).

The predilection for the lower legs is likely due to the slower, gravity-related blood flow in the dilated venules and, consequently, the easier deposition of IgA on the vessel walls (1).

Joint manifestations.
Joint manifestations are present in two-thirds of cases and cause arthralgias that mainly affect the ankles and knees. They are of variable intensity and usually resolve within a short time. One or more joints may be affected, simultaneously or successively. The pain may be associated with periarticular swelling, most often related to synovitis, which does not destroy the joint (11).

Gastrointestinal manifestations.
Abdominal pain is usually moderate (86%), but can also be severe postprandial. Gastrointestinal bleeding can be mild (66%) and only detectable by a positive fecal occult blood test, but can sometimes be severe (20%) and life-threatening. Abdominal ultrasound and/or CT are useful in detecting thickening of the intestinal wall with inflammatory infiltration of the mesentery, or parietal or submucosal hematomas, sometimes stenosing (1).

Endoscopy of the upper and lower digestive tract allows for an accurate topographic diagnosis of the lesions, their extent, and thus the selection of sites for biopsies. Lesions predominate in the second portion of the duodenum. They show mucosal erythema with petechial purpura, erosions and even true necrotic areas of the digestive wall (1).

Renal manifestations.
Manifestations range from microscopic hematuria and mild proteinuria to nephrotic and nephritic syndrome and renal failure. Hypertension may develop at the onset or during recovery from PHS. The most common finding is isolated microscopic hematuria that usually develops within the first few weeks of disease onset. Most cases of nephritic Henoch-Schönlein purpura are mild and the chances of recovery are good (11).

Neurological manifestations
Henoch-Schönlein purpura, although rare, can have neurological manifestations. The symptoms appear between the second and fourth week of the onset of the disease. The most common symptoms are headache, seizures, and other poorly specified changes in the central nervous system that lead to emotional instability characterized by irritability, dizziness, and behavioral changes. Other rare complications include ataxia, intracranial hemorrhage, mononeuropathy, and acute motor sensory axonal neuropathy (11).

DIAGNOSIS.
The diagnosis of Henoch-Schönlein purpura is based on the patient's medical history, the symptoms present, and the findings on physical examination. An almost pathognomonic sign of cutaneous vasculitis due to immune complexes is an erythematous, round or oval retiform purpura with predominance in the lower limbs combined with articular, gastrointestinal and renal manifestations (12).

The immunoglobulin A (IgA) test is usually elevated in 60% of cases, however, it is not specific for diagnosis. On urine examination, pre-otheinuria and hematuria are present. On the other hand, histological analysis of biopsies confirms the diagnosis when there are findings of a leukocytoclastic vasculitis of the postcapillary venules of the skin or proliferative endocapillary glomerulonephritis, each associated with the presence of IgA deposits in the cutaneous postcapillary venules and in the mesangium of all glomeruli, and sometimes along the walls of the glomerular capillaries (1).

EULAR/PRINTO/PRES criteria for Henoch-Schönlein purpura
Diagnosis is made if there is the presence of purpura or petechiae with predisposition in the lower limbs and if at least one of the following 4 criteria is present: (1) abdominal pain, (2) presence of IgA in histology, (3) arthritis or arthralgia, (4) renal failure (13).

TREATMENT.

Symptomatic treatment.

Rest reduces the spread of cutaneous purpura, but does not improve digestive or renal symptoms (1). Compression with elastic bandages and the use of antihistamines reduces vascular dilation, increasing blood flow and thus decreasing the deposition of immunoglobulins (5). Paracetamol and its derivatives are efficient for joint and muscle pain. The use of non-steroidal anti-inflammatory drugs (NSAIDs) is indicated only for joint pain resistant to simple analgesics and due to their antiplatelet effect they should be avoided in patients with renal insufficiency and gastrointestinal bleeding (1).

To relieve abdominal pain, antispasmodics are the medications of choice (1). In cases of remitting vasculitis limited to the skin, the use of Dapsone 50-150 mg/day or Colchicine 1mg/day for 6 months is indicated (14). In patients with renal insufficiency, a renin-angiotensin system blocker should be used as a nephroprotection measure for the control of artenial pressure and proteins in urine. As long as symptoms continue, monitoring should be continuous and once symptoms have been controlled, annual follow-up is recommended (5).

Specific treatment.

Corticosteroids are very useful for reducing abdominal and joint pain. Methylprednisolone infusions accompanied by a short-acting oral corticosteroid is the best treatment of choice when the proteinuria/creatinuria ratio is >1g/g after treatment with renin-angiotensin system blockers (1). In a retrospective study by Maritati, et al (15), with 22 patients, 20 of whom achieved remission after the use of rituximab, however, more studies are needed.

Patient Education.

Information about the disease, treatment, and the importance of adhering to medical indications. Promotion of a healthy lifestyle, including a balanced diet and regular physical activity.

CASE REPORT

A 59-year-old female patient with a 2-year history of clinical symptoms characterized by respiratory distress, hemoptysis and hematuria was performed, at which time a renal biopsy was performed, of which there is no report and the family does not remember, a treatment was instituted for 6 months that the patient and the family do not remember either. Currently, the patient is admitted to the Mount Sinai Hospital in the company of her daughter referring to a 1-month evolution of respiratory distress, abdominal pain and non-productive cough, so she goes to the doctor who diagnoses her with a respiratory infectious process, however, the patient presents exacerbation of the symptoms of two days of evolution characterized by productive cough with blood, Chills and shortness of breath, which is why she went to the emergency department for a comprehensive evaluation.

Vital signs on admission: BP 140/90, HR 82, SAT 81% with 2lt of oxygen through nasal tips, T 37.8. On physical examination, the patient presented generalized pallor, hypoventilated pulmonary fields, rales and crackles from apex to base in both fields with diminished vesicular murmur. Abdomen RHA present, soft, depressible and not painful on palpation. Symmetrical limbs, with preserved muscle tone and strength, without edema with the presence of retiform round palpable purpura in the lower limbs (Image 1). Neurological examination: Oriented, vigilant and collaborative.

It was decided to perform a simple and contrasted computed tomography of the chest whose images were compatible with interstitial pneumonia, however, due to the patient's condition it was not specified as an infectious process, so due to her history of 2 years ago, we proceeded to investigate an autoimmune disease where the markers of autoimmune diseases associated with the patient's clinic were all negative. The only conclusive of all the tests performed was the urine test, which presented severe hematuria and proteinuria in the subnephrotic range. For this reason, consultation with the

nephrology and rheumatology service is requested to evaluate the need for a renal biopsy for both the current condition and the condition of 2 years ago.

During his hospital stay, a simple and contrasted CT scan of the abdomen and a renal and bladder ultrasound with normal parameters were performed. After the clinical evaluation and examinations, both the nephrology and rheumatology services concluded that a kidney biopsy was needed to reach a diagnosis.

Under sedation, a renal biopsy is performed, approximately 25 minutes where 3 fragments of renal biospia are taken without complications, the patient after the procedure is clinically stable. Based on the patient's clinical history, the pulmonology, rheumatology and nephrology departments agree on initiating pulse corticosteroid therapy until the histological result of the renal biopsy is obtained.

1.1 LABORATORY TESTS

Table 1. *Summary of laboratory tests during the hospital stay.*

Day 1: 30/05/2023	
BLOOD BIOMETRY	White blood cells, neutrophils 74.1, lymphocytes 10.3, hemoglobin 15.3, hematocrit 44.3, platelets 23,900
BLOOD CHEMISTRY	Glucose 116, Urea 27.9, Creatinine 0.96, LDH 241, Total Protein 7.96, Albumin 4.21, Globulin 3.75, TGO 20.5, TGP 11.7, Alkaline Phosphatase 141, BT 0.48, BD 0.22, BI 0.26, Amylsa 75, Lipase 46.2
ELECTROLYTES	Na 143, K 4.1, Cl 112, Ca 9.33, Mg 2.09
ARTERIAL BLOOD GASES	PH 7.35, PCO2 36.7, PO2 59, BEECF-5, HCO3 20.6.
TUMOR MARKERS	CEA 3.83, AFP 2.76, CA 125 10.28, CA 15-3 16.03, CA 19-9 17.96
Day 2: 31/05/2023	
BLOOD BIOMETRY	White blood cells 7.48, neutrophils 90.7, lymphocytes 4.4, hemoglobin 12.9, hematocrit 37.4, platelets 217,000, ESR 80.
BLOOD CHEMISTRY	Glucose 191, Urea 31.7, Creatinine 1.09, Total Protein 6.95, Albumin 3.59, Globulin 3.36, TGO 20.8, TGP 11. GGT 11.8, BT 0.39, BD 0.21, BI 0.18, total CK 98.
ELECTROLYTES	Na 138, K 4.5, Cl 111
ARTERIAL BLOOD GAS	PH 7.36, PCO2 38.8, PO2 71, BEECF -3, HCO3 22.4
IMMUNOLOGY	ANTI SCL 70 2.2, ANTI LA SS-B 1.5, ANTI RO SS-A 0.7, ANA 0.44, ANTI PR3 1, ANTI MPO 0.4, C3 143, C4 24, ANCAS NEGATIVE
SEROLOGY	ANTI CCP 3.67, PCR 16.22, Procalcitonin 0.2, FR 13.6.
EMO	PH 5, Density 1.020, Proteins ++, Ketone Bodies +, Nitrites Negative Red blood cells 30-40/field, Leukocytes 8-10/field, Bacteria +, Erythrocyte dysmorphism 10%
MOLECULAR BIOLOGY	AF SARS COV2 negative, Influenza type A and B negative, RSV negative.
Day 3: 01/06/2023	
HEMOSTASIS	TP 13, INR 1.11, TPT 33.3
BLOOD CHEMISTRY	Urea 33.3, Creatinine 0.89
UROLOGY	Protein in urine in 24 hours 576. Urine volume in 24 hours 1,500
Day 4: 02/06/2023	
BLOOD BIOMETRY	White Blood Cells 10.8, Neutrophils 92.8, Lymphocytes 4.2, Hemoglobin 12.5, Hematocrit 35.8, Platelets 299,000.
BLOOD CHEMISTRY	Urea 32.8, Creatinine 0.77

Source: Mount Sinai Hospital Laboratory. Author: María Belén Carrasco Jaramillo

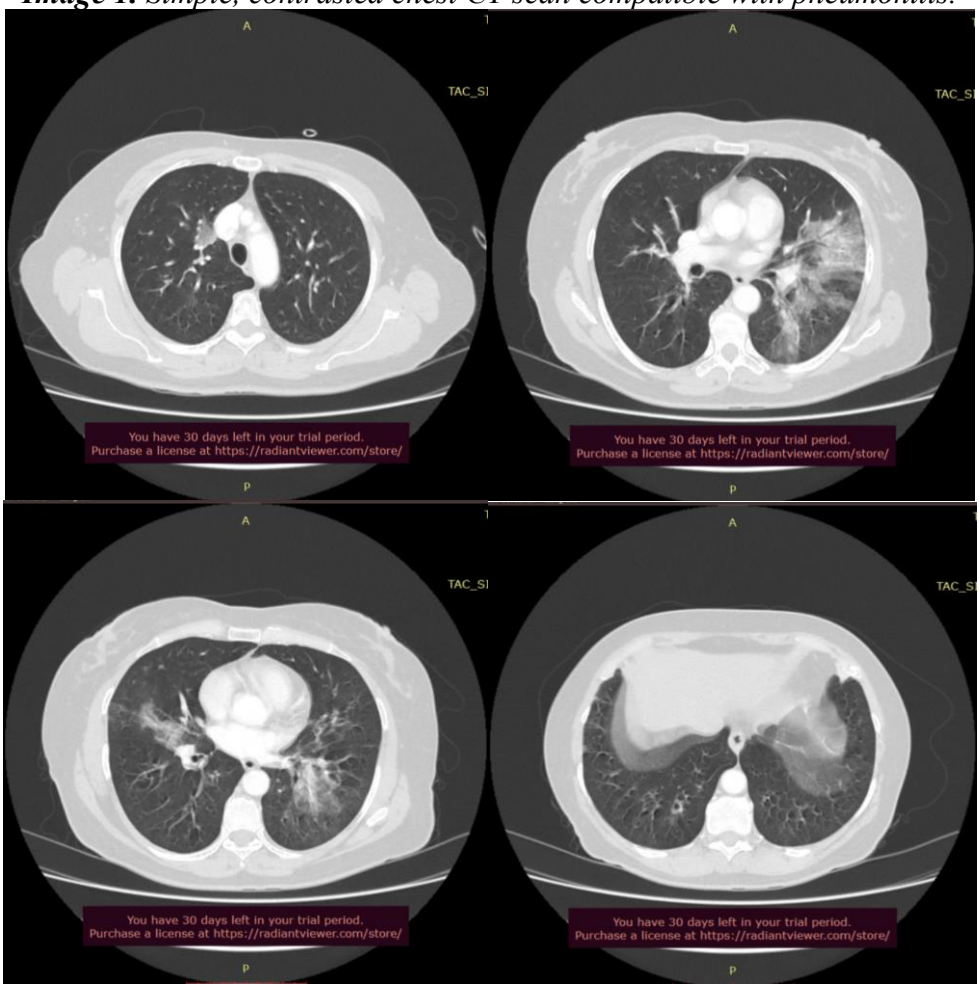
Imaging Exams

Simple, contrasted CT scan of the chest

With the parenchymal window, small volume interstitial infiltrates are observed at the level of the anterior segment of the right upper lobe, large volume in the left posterior apical segment and apical segment of the left lower lobe. A similar image is observed at the level of the lateral segment of the middle lobe, no nodular images are observed. There are small areas of subpleural fibrosis in the linguula.

Diagnosis: Imaging compatible with pneumonitis (interstitial pneumonia)

Image 1. *Simple, contrasted chest CT scan compatible with pneumonitis.*



Source: Mount Sinai Hospital Imaging. Author: María Belén Carrasco Jaramillo

BIOPSY REPORT

In the histological sections of the sample destined for light microscopy, two fragments of renal parenchyma (cortex and medulla) are observed, which together have 11 glomeruli per section in total. Two of these (18.18% of the total glomeruli in the sample) had segmental sclerosing lesions that caused synechiae between the capillary tangles and the Bowman's capsules (S1), located at the glomerular tip. All glomeruli have folded segments of capillary loops, with the occasional formation of double contours by duplication of basement membranes, with cell interposition. There is diffuse mesangial proliferation with widening of the matrix (M1), which causes the glomeruli to acquire a hyperlobed appearance.

Some segments with endocapillary hypercellularity (E0) are not observed. There are no active extracapillary proliferative lesions (cell crescents), (C0). There are no hyaline thrombi or subendothelial deposits that form "wire loops". The podocytes are hypertrophic and form crowns

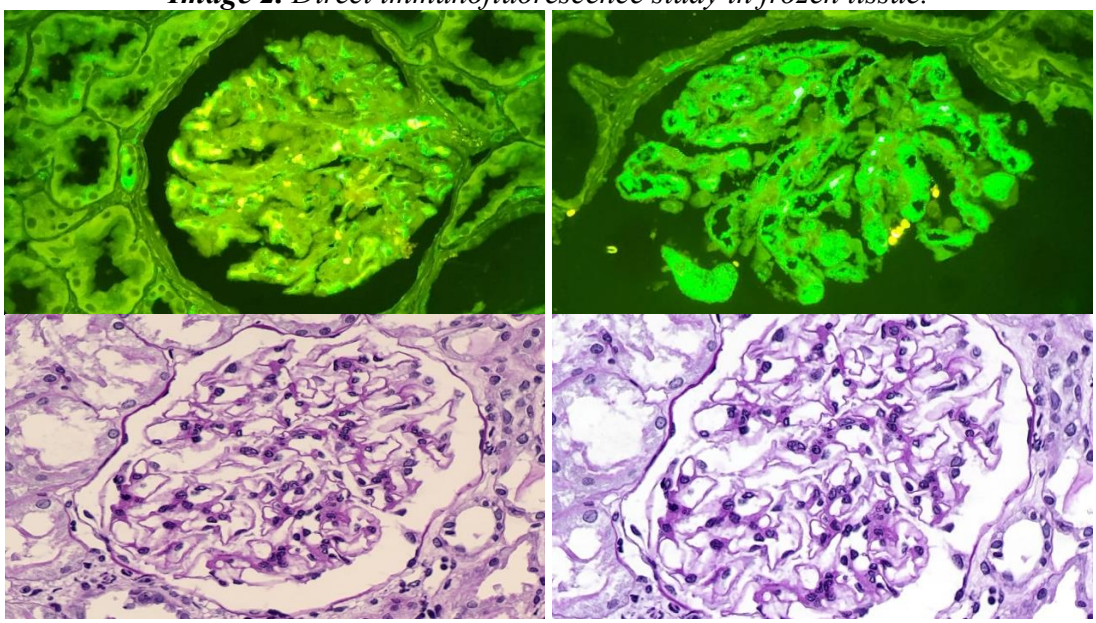
around the tangles. Some podocytes show vacuoles of protein resorption. Interstitial fibrosis with tubular atrophy is present in approximately 10-15% of the tissue evaluated (T0).

An inflammatory infiltrate consisting of lymphocytes, plasma cells and few neutrophils is observed in the interstitium with extension to the tubular walls. The tubular epithelium has moderate regenerative changes, fine cytoplasmic vacuolization, with flattening and multifocal loss of lining cell continuity. Proteinaceous, hematic and tubular intraluminal cellular remains were identified. The preglomerular and interstitial arteriolar vessels are morphologically normal.

Direct immunofluorescence study in frozen tissue.
In the sections of the material intended for the direct immunofluorescence study, a fragment of renal parenchyma is observed, which has 6 glomeruli per section in total.

The result of the study was as follows:
- IgG: Negative.
- IgA: Positive with granular, global and diffuse pattern in the mesangium (3+).
- IgM: Positive with granular, global and diffuse pattern in the mesangium (1+).
- C3c: Positive with granular, global and diffuse pattern in the mesangium (1+).
- Cq: Negative.
- Fib: Negative.
- K: Positive with granular, global and diffuse pattern in the mesangium (2+).

Image 2. *Direct immunofluorescence study in frozen tissue.*



Source: Mount Sinai Hospital Imaging. Author: María Belén Carrasco Jaramillo

DIAGNOSIS:
Based on the clinical symptoms of the patient with a picture characterized by purpura in the lower limbs, headache, joint pain, abdominal pain, hematuria and proteinuria and, after performing imaging and laboratory examinations in conjunction with a new biopsy and direct fluorescence immunofluorescence examination with findings of dominant IgA deposits, the diagnosis of IgA is concluded:

- Henoch-Schönlein purpura (dominant deposits of IgA. M1, E0, S1, T0, C0. Podocyte hypertrophy and spike-like lesions)

- Active tubulo-interstitial nephritis, with acute multifocal tubular lesion and moderate regenerative changes of the epithelium
- Interstitial pneumonia whose infectious process may have triggered IgA vasculitis.

TREATMENT

The pulmonology, rheumatology and nephrology service indicated that the patient receive punctures of 1 g of methylpredsolone on the first day and 1 g IV on the second. At the time of discharge, the patient was sent with prednisone 1g/kg/day PO, losartan 50mg/12h for control of proteinuria and arterial hypertension.

After a month and a half of control, the dose of prednisone was reduced by half by 30mg VO QD for 15 more days and subsequently the dose was decreased, at the time with 5mg QD PO. Renal function, clinical and symptoms of arthralgia and headache controlled, the patient is discussed to maintain control every 2 months.

EVOLUTION

The patient is currently having a favorable clinical evolution, symptoms are kept under control and comes for regular evaluation.

DISCUSSION

Henoch Schönlein purpura in adults is an uncommon entity. According to the EULAR/PRINTO/PRES Criteria for PHS (13), Diagnosis is made if there is the presence of purpura or petechiae with predisposition in the lower limbs and, at least one of the symptoms of In the classic diagnostic tetrad, our patient presented with three symptoms: abdominal pain, arthralgia and presence of IgA in histology. Abdominal pain may be accompanied by nausea, vomiting, diarrhea, rectal bleeding, and melena. However, in the case of our patient, these latter findings were not observed.

According to Piram (4), Infectious processes have previously been identified in up to 50% of patients, especially related to the upper respiratory tract. In addition to microbiological data, the seasonal pattern supports the presence of this triggering infection. The most commonly implicated infectious agent is *streptococcus*, *Mycoplasma pneumoniae, Legionella and Yersinia*, which is related to the case of this patient who presents with pneumonitis.

According to Pillebout, et al (1) In patients presenting with purpura and mild arthralgias without clinical renal involvement, or in those with microscopic hematuria, mild proteinuria, and normal renal function, corticosteroids do not appear to be indicated and these forms are usually treated symptomatically. However, despite the fact that the patient had purpura, mild arthralgias, and normal renal function, the proteinuria values in a 24-hour sample were 576 with a result of "++", indicating the need for corticosteroids.

An immunosuppressive treatment based on corticosteroids was administered, with a favorable response with effective blood pressure control. The prognosis will be conditioned by the possible recurrence of the disease and the evolution of kidney damage, so it is crucial to carry out a prolonged follow-up.

CONCLUSION

The treatment of IgA vasculitis focuses primarily on relieving symptoms in the cutaneous and/or joint forms. However, the treatment of the most severe forms is considerably more complex and remains the subject of debate. Larger studies are required to assess the effectiveness of corticosteroids and possibly develop an approach that includes guidelines for symptomatic treatment, immunosuppression and immunomodulation.

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