Case Report

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Nephrotic Syndrome in Pediatrics and the Role of Medical History

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Abstract

We must always recognize the characteristics of complex pathologies; differentiate what is usual from what is not, even more so, when not all paraclinics are available 100%, then, interrogation and physical examination will always be our best guide in our Act. In this case, it is about a person with few symptoms when arriving at the consultation, but with an account of previous events that pointed to data on autoimmune disorders. In this way, together with the renal biopsy, the diagnosis could be reached and specific therapy started early.

Keywords: Nephrotic syndrome; Medical history; Kidney biopsy

Introduction

In cases of nephrotic syndrome, ideally we should have an arsenal of special diagnostic tests. However, none of them can substitute the value of the clinical history to generate diagnostic suspicions, being the best tool to decide the management at each step. Thus, it is important to recognize the characteristics of a habitual nephrotic syndrome, and to recognize when it is not. Here, the evolution of the patient, and a group of basic tests such as a general examination of urine, immunoglobulin and complement levels, in the context of a patient with nephrotic syndrome, gave us the clues to reach a not so obvious diagnosis initially.

Presentation of the Case

A 14-year-old male was diagnosed with velofacial syndrome, DiGorge syndrome, thrombocytopenic purpura, intra atrial communication, corrected at birth with a patch device, in addition to seizures, on follow-up by Pediatric Neurology, he was referred to the Pediatric Nephrology clinic, due to edema of the lower extremities only, without any other apparent symptoms. Doing the anamnesis of the case, he reported that the purpura data had been found 6 months ago, after a feverish infectious picture, which required antibiotics, with the appearance of lesions in the extremities, plateletpenia, with an improvement in the platelet count after steroid application. Edema of the lower extremities only a previous month, with no predominance of schedules, although sometimes it was greater at night or standing for a long time. On physical examination, with evidence of edema 2 crosses, without arterial hypertension, without other striking data. A general urine test was taken, EGO pH 6.5, 1030 Hb +, Prot +, erythrocytes 12/c, the absence of acute kidney injury was corroborated, with a GFR of 167 mL minm^2, so it was taken as proteinuria in a significant range with hematuria, without other data. ARA2 started. The initial approach was performed, negative infectious profile for HIV, toxoplasma, rubella, CMV, HSV, EBV, C3 94, C4 17.3, normal for age, IgA189, IgG 750, IgM110 IgE30.3, direct negative coombs, rheumatoid factor negative, Kidney ultrasound normal sizes and shapes. Due to the above and in the presence of persistent proteinuria, anti-neutrophil cytoplasm antibodies are taken, negative, and positive ANA in thick speckled 1:80, a weak value, but given the clinical history of a patient with debut of persistent non-nephrotic proteinuria, hematuria and the history of dermal lesions with platelet disease, in an adolescent patient, the possibility of autoimmunity was raised, and it was sent for evaluation by Rheumatology. A new approach is made in another analysis center, being also negative.

However, in a new evaluation at 3 months, he reported occasional headache, increased edema of the lower extremities, so immunological studies were again taken, with urine examination with proteinuria in the nephrotic range, with evidence of hypocomplementemia, hypogammaglobulinemia without deterioration of the renal function, reason why the possibility of vasculitis becomes stronger, and of raises taking of renal biosia. Which is performed with a report of glomerulonephritis by immune complexes, membranoproliferative pattern, with full house immunofluorescence, being characteristic of SLE, with an injury that would correspond to a class III (A/C)+V according to the ISN/RPS 2018 classification (with an activity index of 2/24 and chronicity of 4/12 respectively for class III). With which we were able to complete the diagnosis of systemic lupus erythematosus, the background being secondary to this entity, that is, the lesions of the skin, platelet disease, nephrotic proteinuria, and hematuria, even without deterioration of renal function.

Investigations

IgG 666 (low), C3 73, C4 13, low for age (**Figure 1 and Table 1**).

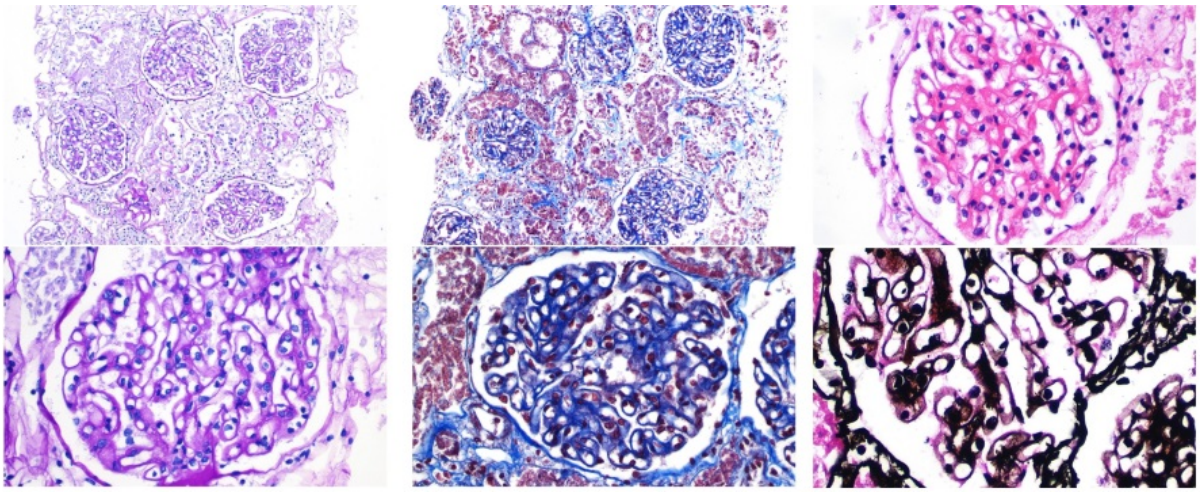

Figure 1: In the histological sections, 22 glomeruli per section in total, of which six (27.27%) present segmental scar-like sclerosing lesions that form synechiae between the capillary tangles and Bowman's capsules. The basement membranes are diffusely thickened, this change conditions a rigid appearance of the capillary loops, with the formation of some double duplication contours in which cell interposition can be seen, in addition, with the silver methenamine staining of Jones, they are identified spicules and abundant filling defects. Expansion of the mesangial matrix is observed with slight proliferation in few segments. In a glomerulus (4.55%), there is segmental endocapillary hypercellularity, leucostasis, and choriorexis. No subendothelial deposits in the form of "wire handles" or hyaline thrombi are identified. Nor are active extracapillary proliferative lesions (cellular crescents) observed. The interstitium has areas of fibrosis with associated tubular atrophy that affect approximately 10%-15% of the cortical surface (grade I). There is denudation of the tubular epithelium and in some fields intraluminal proteinaceous molds are identified. Preglomerular and interstitial arteriolar vessels are morphologically normal.

Table 1: Immunofluorescence.

Anticuerpos	Resultado
IgG	Positivo con patrn granular, global y difuso en membranas basales glomerulares (3+)
IgA	Positivo con patrn granular, global y difuso en membranas basales glomerulares (1+)
IgM	Positivo con patrn granular, global y difuso en membranas basales glomerulares (2+)
C1q	Positivo con patrn granular, global y difuso en membranas basales glomerulares (2+)
C3c	Positivo con patrn granular, global y difuso en membranas basales glomerulares (2+)
C4c	Positivo con patrn granular, global y difuso en membranas basales glomerulares (2+)
Fibringeno	Negativo
Albúmina	Negativo
Kappa	Positivo con patrn granular, global y difuso en membranas basales glomerulares (2+)
Lambda	Positive con patrn granular, global y difuso en membranas basales glomerulares (3+)

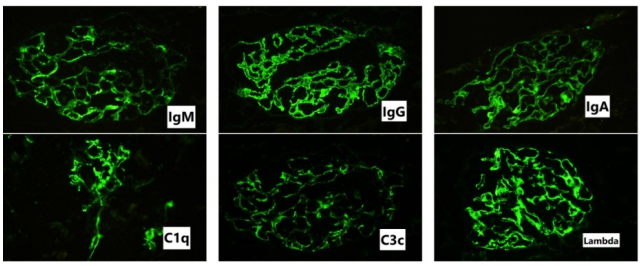

Figure 2: Study of direct Immunofluorescence in frozen tissue. Positivity for IgM, IgG, IgA, C1q, C3c, Lambda.

Differential diagnosis

The possibilities of nephrotic syndrome in this case were considered-Henoch Schonlein purple-IgA vasculitis-Shunt nephritis-Idiopathic nephrotic syndrome (**Figure 2**).

Treatment

In the presence of a nephrotic syndrome in the context of SLE, intravenous steroids were started, and later management was available in our hospital, with oral aziatioprine and prednosine, with resolution of proteinuria.

Outcome and follow-up

Currently with remission of the disease, with follow-up in consultation by rheumatology, without deterioration of renal function.

Discussion

The management of systemic lupus erythematosus (SLE) is a chronic inflammatory disease characterized by very diverse clinical manifestations and the presence in the serum of a variety of autoantibodies that react with different cellular components. The skin, joints, lungs, heart, kidneys, nervous system, and other organs are involved. The American Association of Rheumatology (AAR) developed diagnostic criteria for SLE to establish with certainty the diagnosis of SLE and distinguish it from other rheumatic diseases. The prevalence of SLE is approximately 40 in 100,000 in Europe and North America. SLE is three times more common in African Americans than Caucasians, but it is rare in African blacks. In more than 80% of cases, SLE affects women after puberty. In boys, the prevalence is 1 in 100,000 with girls affected more frequently than boys. The female/male ratio increases from 2: 1 in prepubescent to 4.5:1 in adolescents and 8:1 in adults. In children, the incidence occurs in schoolchildren and adolescents. Renal symptoms are found in 40 to 80% of patients, appearing in the first year. Renal involvement is variable, and in some patients they show minimal abnormalities and others show nephritic syndrome with rapidly progressive renal failure. Proteinuria e is the most frequent symptom, hematuria is associated with proteinuria [1-3].

The main goal of early appropriate treatment is to avoid long-term complications from delayed therapy, that is, irreversible organ damage or eventual death. In addition, a retrospective longitudinal matched cohort study of more than 9,000 SLE patients showed that patients diagnosed within 6 months of onset of clinical symptoms exhibited fewer outbreaks, as well as a lower hospitalization rate and overall costs. related to the LES than the paired ones. Patients diagnosed>6 months from the clinical start; This suggests that the earlier the intervention, the greater the protection against disease-related damage [4-7].

Conclusion

The selection of paraclinics will always be made based on clinical findings. We see patients, not laboratories. The manifestations in SLE can be as variable, from serious to subtle.

Clinical suspicion must always guide our procedure. In-depth questioning will always be the first step. In this case, inappropriate use of oral steroids for apparent thrombocytopenic purpura may have masked the evidence and frank symptoms of systemic lupus erythematosus.

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