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Case Report

Acute nephritic syndrome as the sentinel presentation of class III A lupus nephritis in a young patient

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ABSTRACT

Lupus nephritis (LN) is one of the most serious manifestations of Systemic lupus erythematosus (SLE), occurring in 50-75% of children with SLE, with over 90% developing renal involvement within two years of diagnosis. We describe a diagnostically challenging case of a 12-year-old girl presenting with nephritic syndrome, in whom further evaluation revealed an underlying diagnosis of Systemic lupus erythematosus (SLE). Investigations revealed hypocomplementemia (C3 21.0 mg/dl, C4 1.6 mg/dl), positive antinuclear antibody (ANA IF titre 1:80, ANA ELISA 7.9), strongly positive anti-double-stranded DNA (anti-dsDNA +++), significant proteinuria on 24-hour collection (888 mg/day), and microscopic haematuria. She fulfilled three of eleven American college of rheumatology (ACR) 1997 criteria for SLE. Light microscopy demonstrated focal segmental endocapillary hypercellularity with focal mesangial prominence. Renal biopsy confirmed focal proliferative lupus nephritis (ISN/RPS Class IIIA) with a NIH activity index of 3/24 and a characteristic 'full house' immunofluorescence pattern comprising deposits of IgG (2+), IgA (3+), IgM (1+/2+), C3 (1+/2+), C1q (3+), kappa (1+/2+), and lambda (2+). This case underscores the importance of maintaining a high index of suspicion for SLE in children with nephritic features and hypocomplementemia. Renal biopsy is essential for definitive classification and treatment planning even when ACR criteria are not fully met. Early histopathological diagnosis and class-guided immunosuppressive therapy are critical to preventing progression to end-stage renal disease in paediatric lupus nephritis.

Keywords: Systemic lupus erythematosus, Lupus nephritis, Paediatric, ISN/RPS class III, Full house immunofluorescence, Renal biopsy, Nephritic syndrome

INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic, episodic, multisystem autoimmune disease characterised by widespread inflammatory involvement of blood vessels and connective tissue, accompanied by the production of antinuclear antibodies (ANA), particularly antibodies to double-stranded DNA (dsDNA).[1,2] Paediatric-onset SLE accounts for approximately 15-20% of all SLE cases and tends to follow a more aggressive course than adult-onset disease, with greater renal and haematological involvement.[9] Lupus nephritis (LN) develops in 50-75% of children with SLE and remains the most significant determinant of long-term morbidity and mortality in this population, with more than 90% developing renal involvement within two years of diagnosis.[1] The glomerulus bears the brunt of immune complex deposition and clinical findings alone cannot reliably predict histopathological severity or prognosis.[2,3] Renal biopsy therefore remains indispensable for diagnosis, classification and therapeutic decision-making.[2,3] The international society of nephrology/renal

pathology society (ISN/RPS) classification stratifies LN into six classes, guiding tailored immunosuppressive regimens.[3]

We report a 12-year-old girl diagnosed with focal proliferative lupus nephritis (ISN/RPS Class IIIA) who presented as nephritic syndrome and fulfilled only three of eleven ACR 1997 criteria, highlighting the limitations of a criteria-only diagnostic approach and the pivotal role of early renal biopsy.

CASE REPORT

A 12-year-old girl presented with follow up of nephritic syndrome in the form of mild edema and hypertension. There was no history of preceding throat infection, skin rash, joint pain, photosensitivity, oral ulcers or a family history of autoimmune disease.

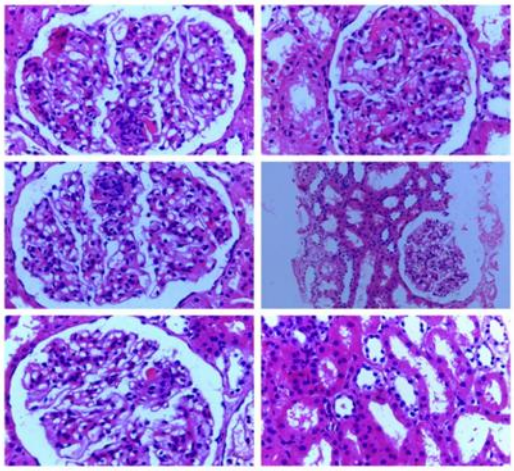

Figure 1: Light microscopy showed focal segmental glomerular endocapillary hypercellularity with focal mesangial prominence.

On examination, she was not sick looking with periorbital oedema. There were no rashes or oral ulcers. Blood pressure was elevated for age. Systemic examination including cardiovascular, respiratory and abdominal examination was within normal limits.

Initial investigations revealed haemoglobin 12.4 g/dl, white blood cell count 6400/mm^3, and platelet count 1.2×10^9/ml. Inflammatory markers were elevated: ESR 120 mm/hr and CRP 0.63 mg/dl. Lipid profile showed total cholesterol 216 mg/dl, LDL 148 mg/dl and triglycerides 155 mg/dl. Serum creatinine was 0.4 mg/dl and serum albumin was 2.9 g/dl, indicating mild hypoalbuminaemia. ASO titre was 32.8 IU/l making post-streptococcal glomerulonephritis less likely.

Immunological workup revealed markedly reduced complement levels with C3 21.0 mg/dl and C4 1.6 mg/dl. ANA by immunofluorescence was positive at titre 1:80, ANA ELISA was 7.9, and anti-dsDNA was strongly positive (3+). Urine examination showed 2+ albuminuria with 4-6 red blood cells per high-power field. The 24-hour urinary protein excretion was 888 mg/day. Ultrasound abdomen showed mildly bulky bilateral kidneys with increased cortical echogenicity and maintained corticomedullary differentiation. Echocardiography was normal.

Based on the clinical and immunological profile, a diagnosis of SLE with lupus nephritis was strongly suspected. The patient fulfilled three of eleven ACR 1997 criteria: renal disorder (proteinuria and microscopic haematuria), positive ANA and strongly positive anti-dsDNA.[4] Renal biopsy was performed for histopathological classification.

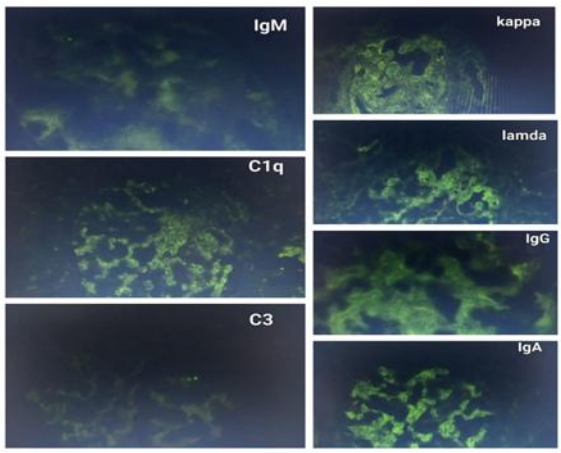

Figure 2: Immunofluorescence demonstrated a characteristic 'full house' pattern with granular capillary wall and mesangial deposits.

Light microscopy showed focal segmental glomerular endocapillary hypercellularity with focal mesangial prominence (Figure 1). Immunofluorescence demonstrated a characteristic 'full house' pattern with granular capillary wall and mesangial deposits of IgG (2+), IgA (3+), IgM (1+/2+), C3 (1+/2+), C1q (3+), kappa (1+/2+) and lambda (2+) (Figure 2). The NIH activity index was 3/24. A final diagnosis of focal proliferative lupus nephritis, ISN/RPS Class IIIA, was established.

The child was given IV methylprednisolone for 3 days followed by oral steroids at 1 mg/kg/day. In view of biopsy-confirmed LN, hydroxychloroquine and mycophenolate mofetil (MMF) at 15 mg/kg/day were initiated. Comprehensive dermatological and ophthalmological evaluations were performed prior to discharge, and symptomatic management was advised. The clinical course was challenging, with delayed remission despite high-dose corticosteroid therapy. Multiple attempts at tapering steroids were associated with relapses, necessitating prolonged immunosuppression and close monitoring.

During follow-up, the MMF dosage was gradually escalated to the maximum permissible dose of 2 g/day over the course of one year. She attained remission after 21 months of therapy.

DISCUSSION

SLE is a complex autoimmune disorder with protean clinical manifestations. Pediatric-onset SLE tends to be more severe at presentation compared to adult-onset disease, with a higher frequency of renal, haematological and central nervous system involvement.[9] LN is the most feared renal complication of SLE, and its early recognition and accurate classification are paramount to prevent irreversible renal damage.[1-7]

The ACR 1997 classification criteria require fulfilment of at least four of eleven criteria for the diagnosis of SLE [4]. However, as this case illustrates, children may develop biopsy-confirmed, histologically significant lupus nephritis while satisfying fewer than four criteria. Our patient met only three: renal disorder, positive ANA and positive anti-dsDNA. This underscores the diagnostic limitations of a purely criteria-driven approach. The SLICC 2012 criteria, more sensitive than ACR 1997, permit diagnosis of SLE with biopsy-proven LN in the presence of positive ANA or anti-dsDNA alone, criteria our patient would readily fulfil.[14]

Hypocomplementemia, observed in our patient (C3 21.0 mg/dl, C4 1.6 mg/dl), reflects classical complement pathway activation by immune complex deposition in the glomeruli.[12] This, combined with strongly positive anti-dsDNA and urinary abnormalities, should prompt renal biopsy regardless of whether the threshold ACR criteria count is achieved.

The ‘full house’ immunofluorescence pattern simultaneous positivity for IgG, IgA, IgM, C3, and C1q is pathognomonic of lupus nephritis and reliably distinguishes it from other immune complex-mediated nephropathies.[3,12] The ISN/RPS 2003 classification categorises Class III as focal LN involving fewer than 50% of glomeruli, with subtype IIIA indicating purely active lesions. This subtype generally carries a better renal prognosis than the mixed active-chronic (IIIA/C) or chronic (IIIC) variants.[3]

Clinical parameters such as degree of proteinuria, haematuria, or hypertension do not reliably predict histopathological class, activity or chronicity indices.[2,3] A NIH activity index of 3/24 in our patient, while reflecting mild activity, necessitated timely institution of immunosuppressive therapy to prevent progression. Brunner et al. demonstrated that pediatric SLE patients exhibit more severe organ involvement at diagnosis, warranting an aggressive diagnostic approach including renal biopsy.[9]

The management of Class III LN follows international guidelines, which recommend induction therapy with high-dose corticosteroids combined with mycophenolate mofetil (MMF) or cyclophosphamide, followed by long-term maintenance.[10-12] MMF has demonstrated comparable efficacy to intravenous cyclophosphamide with a more favourable toxicity profile and is increasingly preferred in children.[10] ACR and KDIGO guidelines advocate treatment decisions based on histopathological activity and chronicity rather than clinical features alone.[11,12] Long-term follow-up studies confirm that early aggressive management significantly reduces the risk of progression to end-stage renal disease.[7,8]

This case carries three key clinical messages: SLE with significant LN can manifest even when the full complement of ACR diagnostic criteria is not met, necessitating a high index of clinical suspicion; renal biopsy is essential for definitive ISN/RPS classification and guides therapy and the ‘full house’ immunofluorescence pattern is virtually diagnostic of LN and should trigger prompt initiation of immunosuppression.

CONCLUSION

Pediatric lupus nephritis may present with fewer than four ACR diagnostic criteria and yet exhibit histopathologically significant disease. The clinical-biochemical triad of hypocomplementemia, positive anti-dsDNA and significant proteinuria should prompt early renal biopsy, irrespective of the total ACR criteria fulfilled. ISN/RPS histopathological classification is indispensable for guiding immunosuppressive therapy. The ‘full house’ immunofluorescence pattern is pathognomonic of LN. Early diagnosis and class-guided treatment are critical to preventing progression to end-stage renal disease and optimising long-term outcomes in affected children.

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