



## COEXISTENCE OF LUPUS NEPHRITIS (CLASS V) AND KERATOACANTHOMA AS THE INITIAL PRESENTATION OF PEDIATRIC SYSTEMIC LUPUS ERYTHEMATOSUS: A RARE CASE REPORT

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**ABSTRACT** **Introduction:** Systemic lupus erythematosus (SLE) is a chronic, multisystem autoimmune disorder with variable presentations. While lupus nephritis is a common and serious manifestation, its occurrence alongside keratoacanthoma as the initial presentation in a PEDIATRIC patient is exceedingly rare. **Case:** We report the case of an 8-year-old male who presented with gross hematuria, proteinuria, purpuric rash, weight loss, hypertension, and generalized edema. Initial evaluation suggested nephrotic syndrome, and the patient was started on corticosteroids and antihypertensives. Renal biopsy revealed mesangio-capillary proliferation with subendothelial deposits, and direct immunofluorescence demonstrated weak positivity for IgG, IgA, IgM, kappa, and lambda in the glomerular basement membrane and mesangium, consistent with Class V lupus nephritis (membranous type). Skin biopsy showed features of keratoacanthoma, with direct immunofluorescence revealing weak IgG positivity in the epidermis. ANA testing was weakly nucleolar positive. **Discussion:** This case illustrates the rare coexistence of lupus nephritis and keratoacanthoma at initial presentation in a PEDIATRIC patient with SLE. Such atypical dual manifestations emphasize the importance of comprehensive evaluation, including both renal and cutaneous biopsies, in suspected cases of childhood lupus. Early recognition is crucial for guiding prognosis and management. **Conclusion:** The unusual coexistence of lupus nephritis and keratoacanthoma in PEDIATRIC SLE expands the clinical spectrum of the disease and underlines the need for timely biopsy and a multidisciplinary approach for accurate diagnosis and optimal treatment.

**KEYWORDS :** systemic lupus erythematosus, PEDIATRIC lupus, lupus nephritis, keratoacanthoma, case report

### INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease characterized by the production of autoantibodies and immune complex deposition, which result in variable clinical manifestations affecting multiple organ systems [1]. Paediatric -onset SLE (cSLE) represents approximately 10 to 20 percent of all lupus cases and is usually associated with a more aggressive disease course compared to adult-onset SLE [2]. Children frequently present with renal, hematological, and neurological manifestations, which are important determinants of morbidity and mortality [3].

Renal involvement in pediatric SLE is both common and clinically significant. Lupus nephritis develops in approximately 60 to 80 percent of children with SLE and represents a major cause of long-term renal impairment [4]. The International Society of Nephrology/Renal Pathology Society (ISN/RPS) classification system defines six histological classes of lupus nephritis. Among these, Class V (membranous) is less common in children than proliferative forms, but it still carries risks of nephrotic-range proteinuria and progression to end-stage renal disease if not treated effectively [5,8].

Keratoacanthoma is a rapidly growing cutaneous neoplasm that shares histological features with squamous cell carcinoma. It typically arises in older adults and is rarely reported in association with SLE [6]. Only a few cases in the literature describe keratoacanthoma complicating lupus, and pediatric reports are exceedingly rare [7].

Here, we describe an eight-year-old boy who presented simultaneously with lupus nephritis Class V and keratoacanthoma as the first manifestation of SLE. To our knowledge, such a presentation has not been reported previously. This case highlights the importance of considering unusual cutaneous lesions in SLE and the need for comprehensive evaluation, including both renal and cutaneous biopsies, in pediatric patients.

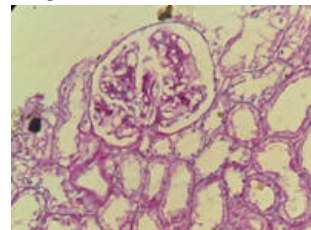
### Case Presentation

An eight-year-old boy presented to our hospital with an eight-day history of gross hematuria, marked proteinuria, purpuric rash, weight loss, hypertension, and generalized edema. Based on these features, nephrotic syndrome was initially suspected, and the child was started on oral corticosteroids and antihypertensive therapy. Owing to persistent hypertension, he subsequently received pulse intravenous methylprednisolone.

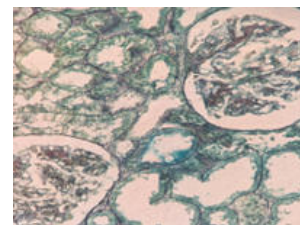
Laboratory investigations revealed proteinuria, hematuria, and mildly deranged renal function with elevated creatinine levels. Complement levels were reduced, with decreased serum C3 and C4. Antinuclear

antibody testing showed weak nucleolar positivity.

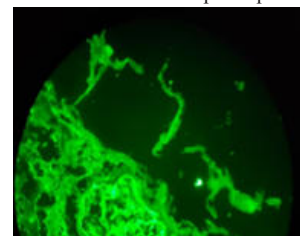
A renal biopsy was performed, which on light microscopy revealed mesangio-capillary proliferation and glomerular expansion. Direct immunofluorescence demonstrated weak linear positivity for IgG, IgA, IgM, kappa, and lambda light chains along the glomerular basement membrane and mesangium. These findings were consistent with Class V lupus nephritis, the membranous variant. [FIGURE 1-5]



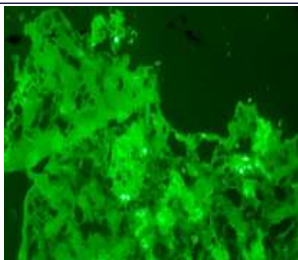
**Figure 1:** Renal biopsy (Periodic Acid-Schiff stain, ×400) showing mesangio-capillary proliferation with expansion of the glomerular tufts.



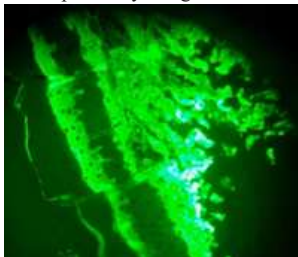
**Figure 2:** Renal biopsy (Silver methenamine stain, ×400) highlighting thickened glomerular basement membranes with subendothelial deposits, consistent with membranous lupus nephritis (Class V).



**Figure 3:** Direct immunofluorescence of renal biopsy showing linear and granular positivity for IgG along the glomerular basement membrane.



**Figure 4:** Direct immunofluorescence of renal biopsy showing mesangial deposits with positivity for IgA.

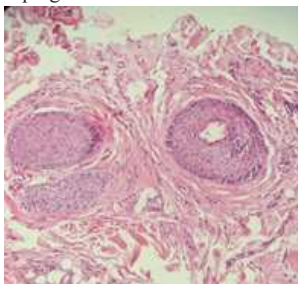


**Figure 5:** Direct immunofluorescence of skin biopsy showing weak linear and granular IgG deposition in the epidermis.

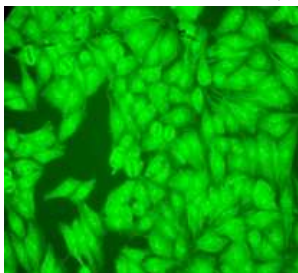
A skin biopsy of the purpuric lesion demonstrated a dome-shaped tumor with a central keratin plug, diagnostic of keratoacanthoma. Direct immunofluorescence of the skin showed granular and linear IgG positivity in the epidermis. [FIGURE 6-8]



**Figure 6-** Keratoacanthoma: a rapidly growing, dome-shaped nodule with central keratin plug



**Figure 7-** Histopathology of keratoacanthoma showing squamous epithelial lobules with central keratin-filled crater (H&E stain, ×40)



**Figure 8-** Direct immunofluorescence demonstrating weak nucleolar antinuclear antibody (ANA) positivity in the epidermal nuclei.

The patient was continued on systemic corticosteroids and supportive therapy. He was closely monitored through multidisciplinary collaboration with pediatric nephrology and dermatology.

## DISCUSSION

Childhood-onset SLE is characterized by higher disease activity and

cumulative organ damage compared to adult-onset SLE [2,3]. Renal disease is particularly common in children, with lupus nephritis occurring in up to 80 percent of patients [4]. Class V lupus nephritis, although less aggressive than proliferative classes III and IV, still poses a risk of persistent proteinuria, treatment resistance, and eventual renal damage if not managed promptly [5,8].

The concurrent finding of keratoacanthoma in a child with SLE is extraordinary. Keratoacanthoma is a low-grade epithelial neoplasm that usually arises in sun-exposed areas in adults. Its association with lupus is rare and has been described mainly in the context of immunosuppression [6,9]. In children, keratoacanthoma has been reported only in isolated cases, such as one involving a teenager with discoid lupus erythematosus [7]. The present case represents a novel dual manifestation of SLE, with renal and cutaneous lesions presenting simultaneously at disease onset.

The pathogenesis underlying this association is unclear. Lupus nephritis results from immune complex deposition in glomeruli, leading to complement activation, inflammation, and structural damage [10]. In keratoacanthoma, dysregulated immune surveillance and chronic inflammation may play a role in promoting abnormal epithelial proliferation [6]. It is plausible that systemic immune dysregulation in lupus may predispose to simultaneous renal and cutaneous involvement.

This case emphasizes the importance of comprehensive evaluation in pediatric SLE. Renal biopsy remains the gold standard for lupus nephritis classification and prognosis. Similarly, cutaneous biopsy in atypical skin lesions provides critical diagnostic information and may reveal rare associations such as keratoacanthoma. The timely use of histopathology and immunofluorescence in both renal and skin tissues facilitated accurate diagnosis and guided treatment in this child.

## CONCLUSION

The present case describes the rare coexistence of Class V lupus nephritis and keratoacanthoma as the initial manifestation of pediatric SLE. This unusual presentation expands the clinical spectrum of childhood lupus and highlights the importance of considering atypical cutaneous lesions in the evaluation of suspected SLE. Early and comprehensive biopsy-based diagnosis, together with multidisciplinary collaboration, is essential to optimize management and prognosis in children with complex lupus presentations.

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