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CASE REPORT

Coexistence of idiopathic nephrotic syndrome, celiac disease, and dermatitis herpetiformis: a rare paediatric case report

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ABSTRACT

Idiopathic nephrotic syndrome (INS) is the most common cause of nephrotic syndrome in childhood. Celiac disease is an autoimmune gluten-dependent enteropathy, and dermatitis herpetiformis (Duhring's disease) represents its most frequent extraintestinal manifestation. The coexistence of INS, celiac disease, and dermatitis herpetiformis constitutes an exceptionally rare clinical finding. We present the case of a 13-year-old girl diagnosed with all three conditions. Following the introduction of a gluten-free diet, complete remission of both nephrological and dermatological symptoms was achieved. This case highlights the importance of an interdisciplinary diagnostic and therapeutic approach and the need to consider gluten-related disorders in children with INS of atypical presentation.

KEY WORDS:
children, proteinuria, nephrotic syndrome, dermatitis herpetiformis, celiac disease.

INTRODUCTION

Nephrotic syndrome is a constellation of symptoms resulting from glomerular injury, which occurs also in the paediatric population. It is classified as congenital, primary (including idiopathic), or secondary to systemic diseases. In children, it is most commonly idiopathic, with an average age of onset between 2–10 years [1, 2]. The aetiology of idiopathic nephrotic syndrome (INS) remains incompletely understood; genetic and immunological factors play a significant role [3, 4]. Pathophysiologically, the disease is associated with podocyte injury and disruption of the glomerular filtration barrier. While activation of Th17 lymphocytes and increased IL-17 levels may promote inflammation, IL-13-induced CD80 expression on podocytes is proposed as a primary mechanism increasing glomerular permeability, particularly in minimal-change disease. Circulating permeability factors that may injure podocytes include haemopexin, soluble urokinase plasminogen activator receptor, and cathepsin L [5–7]. Clinically, the disease manifests as nephrotic-range proteinuria, hypoalbuminaemia, and oedema. According to the IPNA 2023 guidelines, diagnosis requires nephrotic-range proteinuria (urine protein-to-creatinine ratio [UPCR] ≥ 2 mg/mg or ≥ 200 mg/mmol, or ≥ 3+ on urine dipstick), hypoalbuminaemia (< 3.0 g/dl), and oedema; hyperlipidaemia is a common feature but not required for diagnosis [8].

Celiac disease develops in genetically predisposed individuals carrying HLA-DQ2 or HLA-DQ8 antigens and is characterised by an abnormal immune response to gluten. The disease involves activation of $CD4^+$ T lymphocytes against deamidated gluten peptides generated by tissue transglutaminase type 2 (TG2). Autoantibodies against gluten and TG2 are produced, leading to villous atrophy in the small intestine [9–11]. Serological diag-

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nosis relies on measurement of TGA-IgA and EmA-IgA antibodies. A TGA-IgA titre ≥ 10× upper limit of normal (ULN) with positive EmA-IgA from a second blood sample allows diagnosis without intestinal biopsy according to European Society of Paediatric Gastroenterology, Hepatology, and Nutrition guidelines [12].

Dermatitis herpetiformis (Duhring's disease) is the most frequent extraintestinal manifestation of celiac disease. It presents as a polymorphic rash with papules, vesicles, and blisters. While typical locations include the elbows, knees, and buttocks, the lesions are also characteristically found on the lower back and sacral area. The disease is confirmed by granular IgA deposits in the dermal papillae [13–15]. It is worth noting that some paediatric patients may be seronegative for circulating antibodies due to the deposition of immune complexes in the skin [16, 17].

Few cases of nephrotic syndrome coexisting with dermatitis herpetiformis have been reported in the literature, most often in association with celiac disease [18–20]. The present report describes a rare case of concurrent INS, celiac disease, and dermatitis herpetiformis in a child, emphasising the importance of interdisciplinary diagnostic assessment.

CASE REPORT

A 13-year-old girl was admitted due to generalised oedema following multiple insect bites. Physical examination revealed facial and peripheral oedema, with no other abnormalities. Blood pressure was normal. Laboratory tests showed hypoalbuminaemia (1.74 g/dl), hypoproteinaemia (3.96 g/dl), and markedly elevated total cholesterol (596 mg/dl), low-density lipoprotein (428 mg/dl), and triglycerides (205 mg/dl). D-dimer level was 1536 ng/ml, with normal renal function (creatinine 0.41 mg/dl, eGFR 145 ml/min/1.73 m²). Urinalysis revealed massive proteinuria (3073 mg/dl) without haematuria. Immunoglobulin levels showed elevated IgA and IgM with decreased IgG. A first episode of INS was diagnosed. Intravenous methylprednisolone therapy (60 mg/m²/day, equivalent to prednisone) was initiated, followed by oral prednisone. Remission of proteinuria was achieved by day 5. Oral therapy was continued for six months with gradual tapering. The patient remained under nephrology, allergology (sensitisation to moulds, mites, animal dander, and pollen), and cardiology follow-up (QT interval – time from the start of the Q wave to the end of the T wave).

At age 16 years, after 2.5 years of steroids and without any recurrence of proteinuria – even during minor respiratory tract infections – the patient experienced a second relapse, and was readmitted with massive, symmetrical oedema of the face and lower limbs, ascites, and a papular rash on elbows and knees without pruritus. Weightgain of 9.3 kg was noted (46.7–56 kg). Laboratory results showed hypoalbuminaemia (2.4 g/dl), hypoproteinaemi a (4.7 g/dl),hypercholesterolaemia (275 mg/dl),

TABLE 1. Laboratory results on the day of admission according to the hospital laboratory of the Department of Paediatric Nephrology and Paediatrics

Parameters	Patient's result	Reference range
WBC [×10³/µl]	6.83	4.0–10.0
Hb [g/dl]	12.8	12.0–16.0
PLT [×10³/µl]	301	150–400
CRP [mg/dl]	0,12	0–0.5
Albumin [g/dl]	2.4	3.2–4.5
Total serum protein [g/dl]	4.7	6.0–8.0
Urinalysis	–	–
Protein [mg/dl]	Detected	None
RBC [cells/µl]	< 4	0–7
Leukocytes [cells/ µl]	18	0–10
Specific gravity [g/ml]	1.021	1.005–1.025
Creatinine [mg/dl]	0.65	0.50 – 0.90
eGFR (CKD-EPI ml/min/1.73^2)	132	90
Urea [mg/dl]	18.4	8.6–32.1
Na [mmol/l]	135	130–143
K [mmol/l]	4.0	3.5–5.1
Ca [mg/dl]	7.9	8.4–10.2
Total cholesterol [mg/dl]	275	106–224
Triglycerides [mg/dl]	111	34–165
APTT [s]	33.1	26.0–37.0
INR	0.91	0.90–1.20
Fibrinogen [g/l]	6.56	1.8–3.5
D-dimers	2543.0	170–550
ANA		
ANA DFS70	+++	–
ANA anti-centromere	++	–
ANA Mi-2ß	+	–
ANCA	Not detected	–
ASO [IU/ml]	< 13	< 200
RF [IU/ml]	< 9.25	–
QuantiFeron	Negative	–
Hbs antigen	Negative	–
Total anti-HBs [mIU/ml]	1.09	–
Anti-HCV	Negative	Negative
IgA [mg/dl]	292	85–194
IgG [mg/dl]	598	706–1440
IgM [mg/dl]	267	44–113
C3 [mg/dl]	106	90–180
C4 [mg/dl]	25.8	10–40

ANA – antinuclear antibodies, ANA DFS70 – antinuclear antibodies dense fine speckled 70, ANCA – anti-neutrophil cytoplasmic antibodies, anti-HCV – antibodies against hepatitis C virus, APTT – activated partial thromboplastin time, ASO – antistreptolysin O, C3 – complement component 3, C4 – complement component 4,Ca – calcium, CRP – C-reactive protein, eGFR (CKD-EPI) – estimated glomerular filtration rate (chronic kidney disease epidemiology collaboration), Hb – hemoglobin, HBs antigen – hepatitis B surface antigen, IgA – immunoglobulin A, IgG – immunoglobulin G, IgM – immunoglobulin M, INR – international normalized ratio, K – kalium, Na – natrium, PLT – platelets, RBC – red blood cells, RF – rheumatoid factor, total anti-HBs – total antibodies against hepatitis B surface antigen

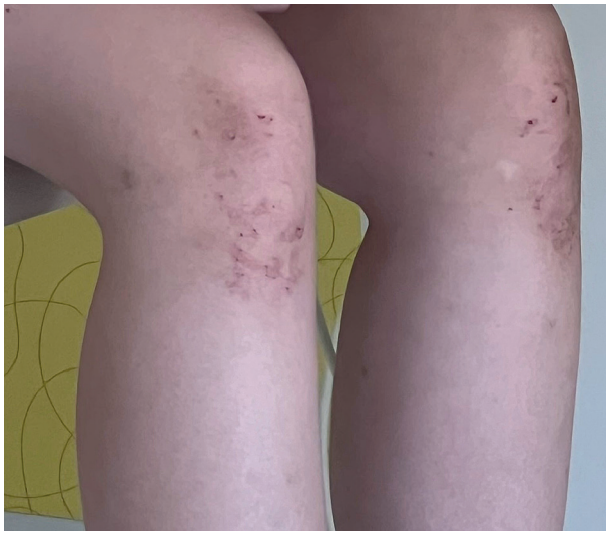

FIGURE 1. Symmetrical erythematous papular and vesicular lesions on the knees of a 16-year-old patient, subsequently confirmed as dermatitis herpetiformis

and nephrotic-rangeproteinuria(UPCR 32.2 mg/mg; 1932 mg/dl).Renal function was preserved (creatinine 0.65 mg/dl, eGFR 132 ml/min/1.73 m^2). Imaging studies (chest X-ray, abdominal ultrasound) were unremarkable. Laboratory findings are summarised in Table 1.

Methylprednisolone and subsequently oral prednisone (60 mg/m^2/day) were administered. Because of severe oedema and reduced diuresis, diuretics (hydrochlorothiazide, furosemide) and antithrombotic prophylaxis (acetylsalicylic acid) were introduced. Due to a respiratory tract infection caused by *Chlamydia pneumoniae* (IgM+), clarithromycin was added. Remission was achieved by day 12 of therapy. Owing to the atypical age of onset (> 10 years), elevated IgA, and positive nonspecific ANA, a kidney biopsy was performed, revealing minimal change disease, confirming idiopathic nephrotic syndrome.

In subsequent weeks, erythematous papular and vesicular skin lesions appeared on the hands, elbows, buttocks, and thighs, accompanied by itching (Figure 1). A skin biopsy, taken from perilesional healthy skin, demonstrated dermatitis herpetiformis (Duhring's disease) with granular IgA deposits. Serologic tests showed anti-tissue transglutaminase antibodies (TGA-IgA: 201.9 CU; > 10× ULN; IgG < 3.8 CU), confirming celiac disease. After the introduction of a gluten-free diet and dermatological therapy, complete remission of renal and skin symptoms was achieved.

DISCUSSION

This case underscores the importance of an interdisciplinary diagnostic and therapeutic approach in paediatric patients with INS of atypical presentation. The coexistence of celiac disease and dermatitis herpetiformis may result from a shared immunogenetic background, involving overproduction of IgA and the presence of HLA-DQ2 and HLA-DQ8 antigens typical of gluten-related disorders. In our patient, initiation of a gluten-free diet led to sustained remission of both nephrological and dermatological symptoms, supporting a potential link between gluten exposure and immune-mediated renal injury [21–23].

Similar observations were reported by Lemley *et al.* [21], who found that in children with steroid-resistant nephrotic syndrome, a gluten-free diet resulted in resolution of proteinuria and clinical improvement. The mechanism may involve modulation of T-cell responses and reduction of pro-inflammatory cytokines such as IL-2, IL-8, and TNF-α, which markedly increase after gluten re-exposure in celiac patients [22]. Furthermore, immunopathological studies have demonstrated deposits of IgA anti-TG2 antibodies in renal biopsies of patients with IgA nephropathy that disappeared following gluten withdrawal, suggesting a shared immunological mechanism in the pathogenesis of both conditions [23].

Although the coexistence of INS with celiac disease or dermatitis herpetiformis is rare, several case reports have been published [24–26]. Boonpheng *et al.* [26] emphasised that patients with celiac disease may develop various nephropathies, including IgA nephropathy and membranoproliferative glomerulonephritis, both of which improved after gluten elimination. Kaplan *et al.* [27] highlighted the autoimmune nature of dermatitis herpetiformis and its close association with celiac disease and other immune-mediated disorders.

Recent studies have also shown that a gluten-free diet can modulate gut microbiota, leading to reduced immune activation and attenuation of systemic inflammation [28]. Moreover, IgA deposits within glomeruli, as demonstrated by Vignon *et al.* [29], may play a pivotal role in glomerular barrier injury and progression of immune-mediated renal diseases. Biyikli *et al.* [30] reported a case of membranoproliferative glomerulonephritis associated with celiac disease, in which renal function improved after gluten withdrawal, further confirming the importance of dietary elimination in limiting renal inflammation.

The concordance of clinical observations and literature data suggests that early identification of gluten-related disorders in patients with INS and appropriate dietary intervention may represent an important component of supportive therapy, particularly in atypical or treatment-resistant cases, but current evidence is insufficient to support the routine use of a gluten-free diet in all patients with INS [21–23, 26, 30].

CONCLUSIONS

In the diagnostic evaluation of children with idiopathic nephrotic syndrome, gluten-related disorders should be considered. Early identification of celiac disease and dermatitis herpetiformis enables effective treatment,

preventing relapses and complications. This case emphasises the importance of collaboration among paediatric nephrologists, gastroenterologists, and dermatologists in the management of autoimmune-mediated diseases in children.

DISCLOSURES

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