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Severe IgA Vasculitis Nephritis in a 12-Year-Old Senegalese Child

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Abstract

IgA vasculitis is the most common childhood vasculitis. Rare among African descendants, renal manifestations remain poorly documented in children in Senegal. We report the case of a 12-year-old Senegalese adolescent who presented with a 3-week history of persistent acute nephritic syndrome associated with symmetrical purpura of lower limbs, polyarthralgia and abdominal pain. Laboratory tests revealed glomerular proteinuria associated with acute renal failure KDIGO Stage 3. Renal biopsy revealed immune deposits of IgA and C3, as well as 95% crescentic fibrocellular glomerulonephritis. The patient was treated with intravenous pulses of methylprednisolone, oral prednisolone and Azathioprine. The outcome was favorable, and the glomerular filtration rate was fully restored after three months of treatment. IgA vasculitis nephritis can be severe and affect vital prognosis and renal function. In our case, early diagnosis and prompt appropriate treatment restored kidney function and that of other affected organs.

Keywords

Acute Renal Failure, Children, Hematuria, IgA Vasculitis, Glomerulonephritis

1. Background

Henoch-Schönlein purpura or IgA vasculitis (IgAV) is a leukocytoclastic vasculitis of small vessels [1]. It is caused by acute perivascular deposition of immuno-

globulin A (IgA) and sometimes is associated with C3, IgG, IgM, fibrin and properdin deposition in the capillaries wall of the dermis, glomeruli and the mesangium [1] [2].

About 90% of IgA vasculitis cases occur between the ages of 3 and 15 years old with a peak incidence at 6 years old [2]. Worldwide, Afro-Caribbeans and African Americans have the lowest incidence, while Asians and Caucasians have the highest incidence [3]. Positive clinical diagnosis is based on the European League Against Rheumatism (EULAR) clinical criteria associating a clinical tetrad of a palpable non-thrombocytopenic purpura, typically non-migratory and nondestructive polyarthralgias, gastrointestinal involvement and renal dysfunction [4] [5]. Renal manifestations of IgA vasculitis occur in 30% to 50% of cases. Most of the time, they are self-limiting. However, if they persist and remain undiagnosed, they can progress to chronic or end stage renal disease [6]. In Africa and Senegal in particular, the epidemiological description and diagnosis of IgA vasculitis nephritis remain a huge challenge, mainly due to late diagnosis and paucity of data.

We report a case of IgA vasculitis nephritis (IgAVN) complicated by KDIGO stage 3 acute renal failure (ARF) in a 12-year-old Senegalese adolescent.

2. Case Report

We report the case of a 12-year-old adolescent who consulted our pediatric nephrology department with a 3-week history of generalised skin rash, diffuse abdominal pain, polyarthralgia, facial and lower limbs swellings as well as dark brown urine.

The history started with a sore throat followed by a pruritic papular eruption distributed over the upper limbs, then extending to the trunk, abdomen and both lower limbs. He first sought consultation at a primary health care facility, where an upper respiratory tract infection and urticaria were diagnosed. He received symptomatic treatment without clinical improvement after five days of treatment. This was followed by the onset of generalised and progressive colicky abdominal pain, throbbing polyarthralgia mainly in the knees and ankles, progressive swelling of the feet, early morning facial puffiness, intermittent high grade fever, anorexia and fatigue. All these elements motivated him to consult our pediatric nephrology department, where he was finally admitted.

His past medical history was relevant for 3 episodes per year of recurrent pharyngitis. He reported family history of rheumatoid arthritis. Physical examination revealed a conscious patient, low-grade fever (T 38˚C), tachycardia (HR 130 b/min), normal respiratory rate (RR 20 C/min), stage 2 hypertension (BP 140/100 mmHg), normal blood glucose (1.09 g/dL). His urine was dark brown in color. Urinary output was 1.98 mL/kg/hr. His height and weight were 140 cm and 28 kg, respectively, giving a BMI for age of 14.28 (between -2 and -3 WHO Z score). The adolescent presented with periorbital edema, mild pitting edema of the dorsum of the feet and symmetrical hyper pigmented macular rashes on lower limbs and

trunk (Figure 1). Abdominal examination revealed diffuse tenderness with no evidence of peritoneal irritation or organomegaly. The rest of physical examination was normal particularly at the level of the joints.

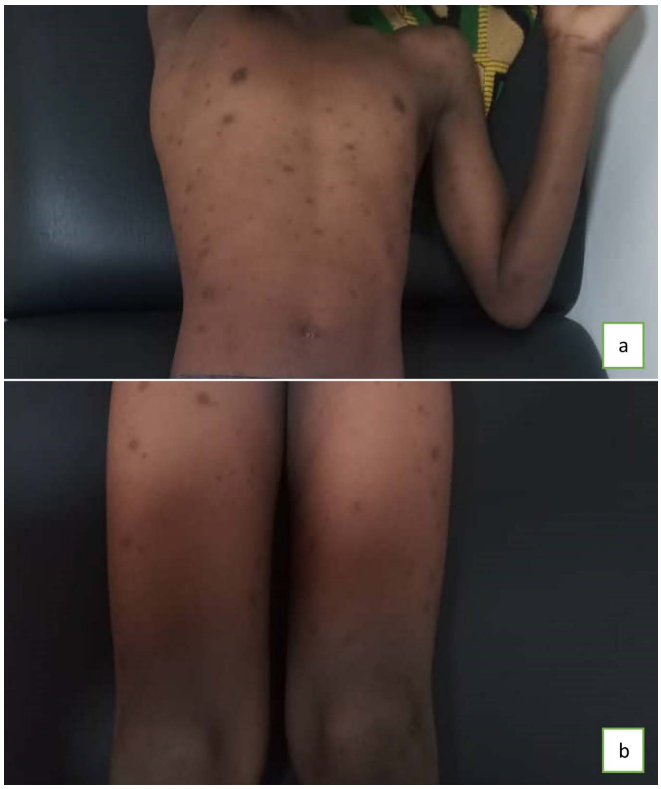


Figure 1. Skin rashes on admission. (a) Hyper pigmented macular rashes on the thorax abdomen and upper limbs; (b) Symmetrical hyper pigmented macular rashes on both thighs.

The urine dipstick showed 2++ proteinuria and 2++ hematuria. Laboratory tests revealed a urine Protein/Creatinine ratio (PCR) at 200 mg/mmol. Serum creatinine level was 19 mg/L. The estimated glomerular filtration rate (eGFR) was 30 mL/min/1.73m^2 (Schwartz). Anti-streptolysin O antibodies (ASLO) were negative. Serum C3 was normal. Other laboratory investigations are reported in Table 1. Abdominal ultrasound was normal. Esophagogastroduodenoscopy reported congestive pangastritis without ulcer. The chest X-ray was normal as well as the electrocardiogram result. The renal biopsy result showed crescentic fibrocellular glomerulonephritis involving 95% of the glomeruli, corresponding to IgAVN grade V of the International Study of Kidney Disease in Children (ISKDC) classification (Figure 2). IgA deposits were also found on immunohistochemistry, as shown in Figure 3.

Based on the clinical picture, laboratory results, IgA deposits and crescentic fibrocellular glomerulonephritis on renal biopsy, the diagnosis of IgA vasculitis nephritis associated with acute renal failure KDIGO stage 3 was established as per EULAR classification criteria.

Table 1. Laboratory results at admission.

Laboratory Tests	Admission results
Full blood count:	
GB (10^3/uL)	26.37
NEUT (10^3/uL)	2.221
PLT (10^3/uL)	167
HB (g/dL)	8.4
MCV (fL)	80.6
MCHC (pg)	34.6
CRP (mg/L)	189.3
Malaria Rapid Diagnostic test	Negative
Blood culture	Negative
ASLO	Negative
Hepatitis BsAg	Negative
Anti Hbc antibodies	Negative
HIV test	Negative
Serum C3 g/L	1.51
Serum C4 g/L	0.21
Serum CH50 U/mL	60
Anti-native DNA	Negative
Total protein g/L	54.4
Blood electrolytes	
Na+ (meq/L)	127
K+ (meq/L)	3.6
Cl– (meq/L)	114
Phosphorus (mg/L)	69.9
Magnesium (mg/L)	19.9
Calcium (mg/L)	93.5
Blood group	O+
HB electrophoresis	AA
Coagulation profile:	
Prothrombin time	100
INR	1.37
aPTT	31s
Renal	
Urine dipstick	Protein ++ Haematuria ++
Urine protein/creatinine ratio mg/mmol	200
Creatinine mg/L	19
Urea g/L	1.12
eGFR (Schwartz)	30.4 ml/min/1.73

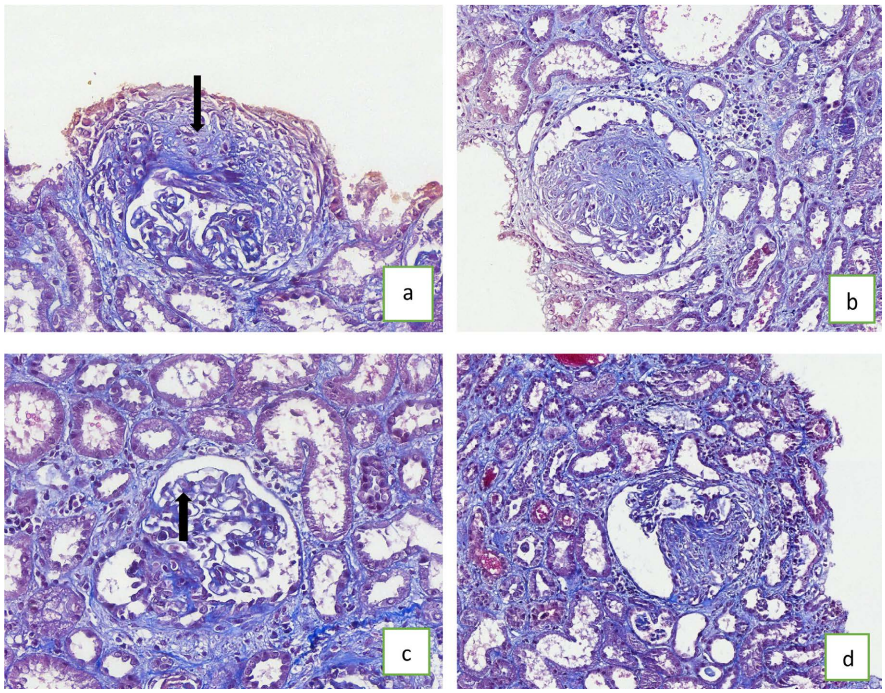
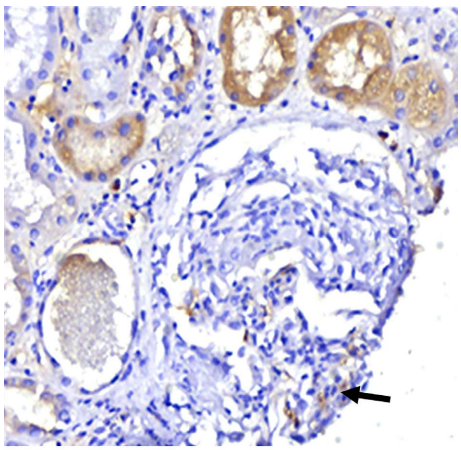


Figure 2. Crescentic glomerulonephritis. (a) Extracapillary fibrocellular proliferation around the flocculus. Masson trichrome Gx250; (b) Extracapillary proliferation and fibrosis of the flocculus. Masson trichrome Gx250; (c) Segmental cellular crescent. Masson trichrome Gx250; (d) Fibrocellular crescent. Masson trichrome Gx250.



IgA deposits on a mesangium (black arrow)

Figure 3. IgA deposits on mesangium.

The patient received 3 intravenous pulses of methylprednisolone at 1 g/1.73m^2 every 48 hr for one week combined with oral prednisone at 2 mg/kg/day and oral Azathioprine at 2 mg/kg/day for 3 months. Upon admission, he also received supportive treatment. After two weeks of oral prednisone and azathioprine, there was regression of extrarenal symptoms and gradual restoration of renal function without residual proteinuria. The patient's renal outcome remained favorable after six months of follow-up, as shown in **Figure 4**.

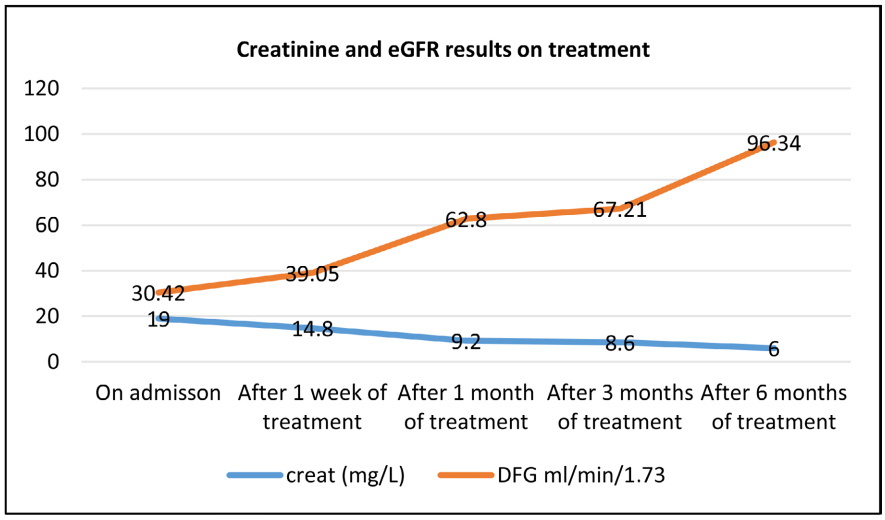


Figure 4. Creatinine and GFR evolution on corticosteroids + AZA treatment.

3. Discussion

IgA vasculitis is the wide form of systemic vasculitis seen in children [5]. It can be encountered in all age groups but in 90% of cases, it occurs between 3 and 15 years of age, with a peak incidence between 4 and 6 years of age [7]. A slight male predominance is also described [2] [8]. IgA vasculitis affects all ethnicities but is rare in African Americans [6]. As Hamani *et al.* point out in their study, our patient was also of African descent [9]. Thus, although rare in black ethnicities, this highlights the genetic and geographic variability of IgA vasculitis [10].

Although cause-effect relationship of IgA vasculitis is difficult to establish, in 40% to 50% of cases, upper respiratory tract infections (URTIs) precede the onset of the disease [6] [7] [11]. Our patient reported recurrent pharyngitis thus suggesting a potential infectious trigger. He also reported generalised pruritic macular rashes. In our setting, this usually suggests diagnoses such as chickenpox, scabies and urticaria, which are much more frequent thus delaying early recognition of IgAV. In more than 95% of cases, the rash is an early sign of IgA vasculitis in children [5] [9] [12] [13]. The characteristic rash is a palpable no thrombocytopenic purpura that progresses from erythema to papules and then to palpable non blanching purpura [1] [5]. This may vary among patients. Atypical skin lesions may appear to healthcare personnel outside of flare-ups, contributing to IgAV misdiagnosis and requiring biopsy whenever possible [4]. In this case, skin biopsy was not available.

The diagnosis of IgA vasculitis is primarily clinical. Besides the rash, other extra renal manifestations of IgAV in this case included abdominal and joint pain. These findings are similar to the study by Maha Oudrhiri *et al.* in Morocco who found joint and gastrointestinal manifestations in 56.5% and 65.5.8% of cases, respectively [14]. Sylvia Gomez *et al.* also reported gastrointestinal and joints involvement in 54 % and 82% of cases, respectively [15]. Gastrointestinal manifestations (GI) are present in 50% - 80% of patients with IgAV presenting primarily

as diffuse abdominal pain [11]. In the presence of severe abdominal pain, it is mandatory to exclude an acute surgical abdomen. Abdominal ultrasound of our patient was normal, while esophagogastroduodenoscopy showed non erosive congestive pangastritis. The spectrum of endoscopic findings is based on the severity of the vasculitis [16]. Usually, irregular, ulcerative nodular lesions or hematoma-like protuberances are characteristic of IgA vasculitis in the duodenum [16]. Joint involvement in IgA vasculitis consists of a transient non migratory symmetrical polyarthralgia affecting mainly the large joints (especially the knees and ankles) [5] [16].

Even at this stage of clinical presentation, the diagnosis of IgA vasculitis remains challenging in our setting, considered rare, particularly in the presence of an atypical skin rash associated with abdominal and joint pain. It is therefore necessary for our health care practitioners, especially those in primary care, to include IgA vasculitis among their differential diagnoses in the presence of cutaneous, joint, and digestive manifestations in a child.

This case also highlighted renal involvement of IgA vasculitis. Patient presented with a nephritic syndrome combining macroscopic hematuria, edema and grade 2 hypertension. Moderate glomerular proteinuria was also reported. Given the clinical course of this patient, it is legitimate to first consider post infectious glomerulonephritis which is much more common in our context. Therefore, laboratory investigations should search as much as possible post infectious causes of glomerulonephritis. In this clinical case, the infectious screening was negative and complement C3 and CH50 assays were normal. In the course of multiple organ symptoms and dysfunction, lupus nephritis can also be considered as a differential diagnosis and screened for. Anti-native DNA antibodies were negative in this case.

Manifestations IgA vascular nephritis are variable. They range from hematuria and or proteinuria to chronic renal failure including nephritic and nephrotic syndrome [17]. In case of impaired estimated glomerular filtration rate (eGFR), persistent proteinuria, nephrotic or nephritic syndrome, renal biopsy is mandatory [1] [2] [9] [10]. Our patient had an altered eGFR of 30 ml/min/1.73m^2 (Schwartz), corresponding to Stage 3 acute renal failure according to the KDIGO classification.

The performed renal biopsy reported morphological changes, immune deposits of IgA and C3 and identified severe crescentic glomerulonephritis with 95% glomerular involvement corresponding to ISKDC grade V [18]. This finding is similar to that of Sarina Butzner *et al.* who reported 100% mesangial IgA deposits on renal biopsy of all patients [19]. The pathological hallmark of IgAVN is the deposition of IgA-containing immune complexes in the renal mesangium. The deposited immune complexes activate the alternative complement pathway (with C3 deposition) and recruit inflammatory cells causing glomerulonephritis [6]. Currently, the International Study of Kidney Disease in Children (ISKDC) classification is mainly used for the histological analysis of IgAVN [4].

To date, there are few prospective, randomized, controlled studies on the treat-

ment of IgAVN and their results remain controversial [12] [20] [21]. Some studies highlight a potential beneficial impact of corticosteroids. Oral prednisolone and/or pulsed methylprednisolone should be used as early treatment in mild to moderate cases [4]. Severe forms, corticosteroid-dependent and corticosteroid-resistant forms of IgAVN may require immunosuppressive treatments, such as calcineurin inhibitors (CNIs), azathioprine (AZA), cyclophosphamide, mycophenolate (MMF), rituximab and plasmapheresis [4] [20] [22]. AZA was used in our patient in combination with oral prednisolone for 3 months.

The clinical outcome was favorable, with complete recovery of renal function. AZA has been shown to be effective in combination with oral corticosteroids in severe forms of IgAVN. No formal guidelines are available regarding the duration of treatment. In many studies, no patient experienced adverse effects associated with AZA treatment [12].

4. Conclusion

We reported in a resource-limited country a case of IgA vasculitis nephritis in an adolescent successfully managed despite late diagnosis. The combination of corticosteroids and Azathioprine resulted in a complete recovery of renal function and of regression of extra renal symptoms. This case therefore encourages awareness of the diagnosis of IgA vasculitis nephritis in the presence of acute nephritic syndrome associated with extra renal signs in order to improve prompt management and obtain a favorable outcome.

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Ethical Consideration

Informed consent form and permission for publication were granted from the patient and their parents.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Pillebout, E. and Sunderkötter, C. (2021) IgA Vasculitis. *Seminars in Immunopathology*, **43**, 729-738. https://doi.org/10.1007/s00281-021-00874-9

[2] Parums, D.V. (2024) A Review of Iga Vasculitis (Henoch-Schönlein Purpura) Past, Present, and Future. *Medical Science Monitor*, **30**, e943912. https://doi.org/10.12659/msm.943912

[3] Xu, L., Li, Y. and Wu, X. (2022) IgA Vasculitis Update: Epidemiology, Pathogenesis, and Biomarkers. *Frontiers in Immunology*, **13**, Article 921864. https://doi.org/10.3389/fimmu.2022.921864

[4] Ozen, S., Marks, S.D., Brogan, P., Groot, N., de Graeff, N., Avcin, T., *et al.* (2019) European Consensus-Based Recommendations for Diagnosis and Treatment of Immunoglobulin a Vasculitis—The SHARE Initiative. *Rheumatology*, **58**, 1607-1616. https://doi.org/10.1093/rheumatology/kez041

[5] Leung, A.K.C. (2020) Henoch-Schönlein Purpura in Children: An Updated Review. *Current Pediatric Reviews*, **16**, 265-276. https://doi.org/10.2174/18756336mta2lndyc2

[6] Dyga, K. and Szczepańska, M. (2020) IgA Vasculitis with Nephritis in Children. *Advances in Clinical and Experimental Medicine*, **29**, 513-519. https://doi.org/10.17219/acem/112566

[7] Reamy, B.V., Servey, J.T. and Williams, P.M. (2020) Henoch-Schönlein Purpura (IgA Vasculitis): Rapid Evidence Review. *American Family Physician*, **102**, 229-233.

[8] Vasküliti, I., Purpurası, H.S., Pediatrik, O., Epidemiyolojik, H., Ve, K., Bulgularının, L., *et al.* (2024) Evaluation of Epidemiological, Clinical, and Laboratory Findings in Pediatric Patients with IgA Vasculitis (Henoch-Schönlein Purpura). *Türkiye Çocuk Hastalıkları Dergisi*, **18**, 111-116.

[9] Hamami, A., Babakhouya, A., El Ouali, A., Ghannam, A., Rkain, M. and Benajiba, N. (2021) P062 Vasculitis in Children: Epidemiological, Clinical and Para Clinical Profile in the Oriental Region of Morocco—About 33 Cases. *Rheumatology*, **60**, keab722.054. https://doi.org/10.1093/rheumatology/keab722.054

[10] Gardner-Medwin, J.M., Dolezalova, P., Cummins, C. and Southwood, T.R. (2002) Incidence of Henoch-Schonlein Purpura, Kawasaki Disease, and Rare Vasculitides in Children of Different Ethnic Origins. *The Lancet*, **360**, 1197-1202. https://doi.org/10.1016/s0140-6736(02)11279-7

[11] Amatruda, M., Carucci, N.S., Chimenz, R. and Conti, G. (2023) Immunoglobulin a Vasculitis Nephritis: Current Understanding of Pathogenesis and Treatment. *World Journal of Nephrology*, **12**, 82-92. https://doi.org/10.5527/wjn.v12.i4.82

[12] Stone, H.K., Mitsnefes, M., Dickinson, K., Burrows, E.K., Razzaghi, H., Luna, I.Y., *et al.* (2023) Clinical Course and Management of Children with Iga Vasculitis with Nephritis. *Pediatric Nephrology*, **38**, 3721-3733. https://doi.org/10.1007/s00467-023-06023-8

[13] Marro, J., Williams, C., Pain, C.E. and Oni, L. (2023) A Case Series on Recurrent and Persisting IgA Vasculitis (Henoch Schonlein Purpura) in Children. *Pediatric Rheumatology*, **21**, Article No. 85. https://doi.org/10.1186/s12969-023-00872-1

[14] Oudrhiri, M., Bentahila, A. and Chkirate, B. (2020) Purpura rhumatoide de l'enfant: À propos de 46 cas. *PAMJ Clinical Medicine*, **2**, Article 149. https://doi.org/10.11604/pamj-cm.2020.2.149.21147

[15] Gómez, S., Pérez, M., *et al.* (2020) Henoch-Schonlein Purpura in Pediatrics: Ten Years of Experience at a Moderate Risk Office of a General Hospital. *Archivos Argentinos de Pediatria*, **118**, 31-37.

[16] Jelusic, M., Sestan, M., Giani, T. and Cimaz, R. (2022) New Insights and Challenges Associated with IgA Vasculitis and Iga Vasculitis with Nephritis—Is It Time to Change the Paradigm of the Most Common Systemic Vasculitis in Childhood? *Frontiers in Pediatrics*, **10**, Article 853724. https://doi.org/10.3389/fped.2022.853724

[17] Williams, C.E.C., Lamond, M., Marro, J., Chetwynd, A.J. and Oni, L. (2023) A Narrative Review of Potential Drug Treatments for Nephritis in Children with Iga Vasculitis (HSP). *Clinical Rheumatology*, **42**, 3189-3200. https://doi.org/10.1007/s10067-023-06781-8

[18] Jelusic, M., Sestan, M., Cimaz, R. and Ozen, S. (2019) Different Histological Classifications for Henoch-Schönlein Purpura Nephritis: Which One Should Be Used? *Pediatric Rheumatology*, **17**, Article No. 10. https://doi.org/10.1186/s12969-019-0311-z

[19] Butzer, S., Hennies, I., Gimpel, C., Gellermann, J., Schalk, G., König, S., *et al.* (2022) Early Clinical Course of Biopsy-Proven IgA Vasculitis Nephritis. *BMC Pediatrics*, **22**, Article No. 570. https://doi.org/10.1186/s12887-022-03611-9

[20] Delbet, J., Parmentier, C., Herbez Rea, C., Mouche, A. and Ulinski, T. (2021) Management of IgA Vasculitis with Nephritis. *Pediatric Drugs*, **23**, 425-435. https://doi.org/10.1007/s40272-021-00464-0

[21] Namgoong, M. (2020) Management of IgA Vasculitis Nephritis (Henoch-Schonlein Purpura Nephritis) in Children. *Childhood Kidney Diseases*, **24**, 1-13. https://doi.org/10.3339/jkspn.2020.24.1.1

[22] Rohner, K., Marlais, M., Ahn, Y.H., Ali, A., Alsharief, A., Novak, A.B., *et al.* (2024) Outcome of Immunosuppression in Children with Iga Vasculitis-Related Nephritis. *Nephrology Dialysis Transplantation*, **39**, 1299-1309. https://doi.org/10.1093/ndt/gfae009