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IgA VASCULITIS IN THE FORM OF CUTANEOUS PRESENTATION IN A 45-YEAR-OLD WOMAN TREATED WITH CORTICOSTEROID: CASE REPORT

VASCULITE POR IgA SOB FORMA DE APRESENTAÇÃO CUTÂNEA EM MULHER DE 45 ANOS TRATADA COM CORTICOSTEROIDE: RELATO DE CASO

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ABSTRACT

Immunoglobulin A vasculitis (IgAV) is known to be a systemic vasculitis of small vessels. Although is possible that this condition sets up in any age group, the epidemiological profile of this disease mostly affects the pediatric population between 2 and 10 years of age. In this sense, this descriptive article aims to report the rare case of an adult patient who evolved with IgAV, presented in a cutaneous form, and treated with corticosteroids. For that, after the patient's free and informed consent, the study was approved by the Research Ethics Committee of the University of Rio Verde (UniRV, Goiás, Brazil), whose Certificate of Presentation for Ethical Consideration is 36621920.0.0000.507. Therefore, it was found that the patient evolved with clinical compatible with IgAV of probable cutaneous involvement due to lesions in the form of purpura and renal involvement, presenting a good therapeutic response with corticosteroid therapy at non-immunosuppressive doses and after 1 year and 3 months of follow-up, there was remission of the clinical picture without new episodes. There are some reports in the literature about the good therapeutic response of this clinical with corticosteroid therapy at non-immunosuppressive doses, however, clinical trials are needed.

Keywords: Corticosteroids. Purpura. Schoenlein-Henoch purpura. Vasculitis.

RESUMO

A Vasculite por imunoglobulina A (IgAV) é conhecida por ser uma vasculite sistêmica de pequenos vasos. Apesar desta condição ser capaz de se estabelecer em qualquer faixa etária, o perfil epidemiológico desta doença afeta, majoritariamente, a população pediátrica situada entre 2 e 10 anos de idade. Nesse sentido, este artigo descritivo, tem como objetivo relatar o raro caso de uma paciente adulta que evoluiu com a IgAV, apresentação de forma cutânea, sendo tratada com corticosteroide. Para tanto, após consentimento livre e esclarecido da paciente foi aprovado pelo Comitê de Ética em Pesquisa da Universidade de Rio Verde (UniRV – GO), cujo Certificado de Apresentação para Apreciação Ética é 36621920.0.0000.507. Logo, descobriu-se que a paciente evoluiu com clínica compatível para IgAV de provável acometimento cutâneo pelas lesões em forma de púrpura e pelo envolvimento renal, apresentando boa resposta terapêutica com a corticoterapia em doses não imunossupressoras e após 1 ano e 3 meses de *follow-up* houve remissão do quadro sem novos episódios. Encontra-se na literatura alguns relatos sobre a boa resposta terapêutica desta forma clínica com a corticoterapia em doses não imunossupressoras, no entanto ensaios clínicos são necessários.

Palavras-chave: Corticosteroides. Púrpura. Púrpura de Schoenlein-Henoch. Vasculite.

INTRODUCTION

Immunoglobulin A vasculitis (IgAV) is known to be a systemic vasculitis of small vessels. According to Rigante *et al.* (2013), it is found that the main clinical manifestations occur due to the deposition of IgA in small vessels in some organs and/or systems and cause signs and symptoms such as nonthrombocytopenic purpura, arthritis, abdominal pain, digestive hemorrhage, and renal involvement (hematuria/proteinuria) (ROBERT *et al.*, 2007; SAULSBURY, 2010).

It is known that IgAV was called Henoch-Schönlein Purpura (PHS) or anaphylactoid purpura, and its nomenclature changed after the 2012 International Chapel Hill Consensus Conference, given the predominant deposit of galactose-deficient IgA1 in the wall of the affected vessels, being the most characteristic in the pathophysiology of the disease (JENNETTE *et al.*, 2013).

Although IgAV can establish itself in any age group, the epidemiological profile of this disease mostly affects the pediatric population between 2 and 10 years of age (AGRAHARKAR *et al.*, 2000; GEDALIA, 2004). Studies show that the average annual incidence was reported in 6.2 - 20.4 cases / 100,000 children, with men affected twice as often as women, being less common in black children compared to white or Asian children (GEDALIA, 2004; ROBERT *et al*., 2007). On the other hand, its occurrence in adults is quite uncommon and estimated at 13 cases per million inhabitants (YANG; YU; CHIANG, 2014).

It is noticed that the etiopathogenesis of IgAV combines hereditary changes and both environmental (such as infections) and immunological factors. The abnormality of the immune system involves humoral and cellular immunity, cytokine release, coagulation mechanisms, genetic susceptibility, and inflammatory status. However, it is inferred that research using animal models with IgAV is currently limited. Thus, the etiology, pathophysiology, clinical treatment, and development of drugs for this disease are still insufficient (KAWASAKI; HOSOYA; SUZUKI, 2005).

In general, IgAV with gastrointestinal (GI) involvement represents about 85% of cases and ranges from mild symptoms to fatal complications, such as perforation and acute mesenteric ischemia, and it is found that the disease affects the duodenum, especially the second duodenal portion. (MOSLEY; DESAI; GUPTA, 1990; COLLINS; DUKE, 1995; SZER, 1996; EBERT, 2008). Furthermore, the relationship between IgAV and Epstein-Barr virus (EBV) infection is known, in which this virus has been implicated in the pathogenesis of different vasculitic syndromes, such as polyarteritis nodosa and vasculitis associated with antineutrophil cytoplasmic antibodies (ANCA) (CALDEIRA *et al*., 2007; YAMAGUCHI *et al*., 2014).

Commonly, for rare skin complications such as hemorrhagic bullous lesions, corticosteroid immunosuppression is continuously helpful to control inflammation and limit the extent of necrosis and subsequent scarring. Typically, this treatment causes a dramatic improvement in clinical symptoms, observed within a few days (BOER *et al*., 2010; DHANJAL *et al*., 2017). Despite this, there is no evidence of the efficacy of corticosteroids on renal or gastrointestinal prognosis and the duration of purpura. In the presence of renal involvement, specific therapy is relevant (HUBER *et al*., 2004; WEISS *et al*., 2007; HAHN *et al*., 2015).

In this sense, this descriptive article aims to report the rare case of an adult patient who evolved with IgAV, presented in a cutaneous form, and treated with corticosteroids.

CASE REPORT

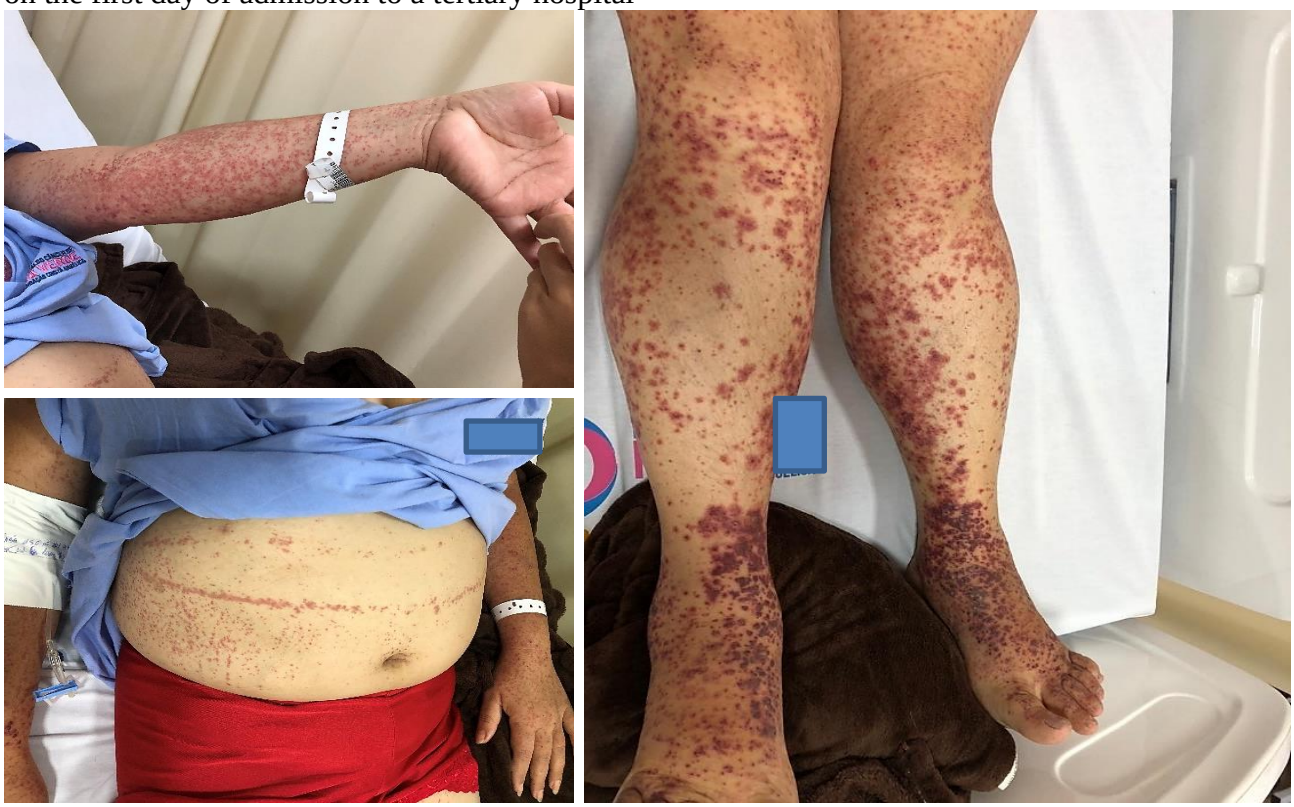
Descriptive study, in the form of a case report, from a tertiary philanthropic hospital in the Brazilian Midwest, after the patient's consent through the Free and Informed Consent Term (FICF), the project was submitted and approved by the Research Ethics Committee by the University of Rio Verde (UniRV – GO), whose Certificate of Presentation for Ethical Consideration is 36621920.0.0000.507.

A 45-year-old female patient came to the Emergency Care Unit (UPA) reporting that she had noticed 10 days earlier the appearance of erythematous papules and purpura starting in the topography of the calcaneus and radiating to the whole body, sparing only the face and breasts. She also mentioned pain in the lower limbs (LL), of the throbbing type, scored in 7 (numerical pain scale 0-10), heat, redness, edema, pruritus, paresthesia, and impaired walking distance. Hands are also swollen. She reports that she had sought the UPA on another occasion for dysuria, polyuria, when 1 tablet (cp) of Ciprofloxacin 500 mg every 12 hours was prescribed and in use for 2 days. She used Silver Sulfadiazine herself in the skin lesions without clinical improvement. Denies: nausea, vomiting, fever, shivering, urinary urgency, and abdominal pain. Denies drug allergy. After two days, the patient was transferred to a tertiary hospital for investigation of the condition in a ward bed by the Medical Clinic.

In her past pathological history, she mentions systemic arterial hypertension (SAH) in regular use of Losartan 50 mg/day; Hydrochlorothiazide 25 mg/day; type 2 diabetes mellitus in regular use of Metformin 850 mg, 1 tablet every 8 hours; Glibenclamide 5 mg/day; Insulin NPH 2 IU at night; depressive disorder in use of fluoxetine 20 mg/day. Denies: dyslipidemias and thyroid diseases. She reported a laparoscopic cholecystectomy 6 months earlier and a tubal ligation 18 years ago. Denies: alcoholism and smoking.

On physical examination, the patient was in good general condition, euthymic and with normal attention span, lucid and oriented in time and space, acyanotic, anicteric, ruddy, and hydrated. 98% saturation in ambient air; heart rate (HR) 80 bpm; respiratory rate (RR) 16 bpm; axillary temperature (TAX) 35.7°C. Cardiorespiratory and abdominal semiology without alterations. Upper limbs (UL) and lower limbs (LL): symmetrical depressive edema (++ / 4+), elastic and warm. Diffuse skin lesions all over the body with erythematous papules and purpura are shown in Figure 1.

Figure 1 - Diffuse skin lesions all over the body in the form of purpura and erythematous pustules on the first day of admission to a tertiary hospital

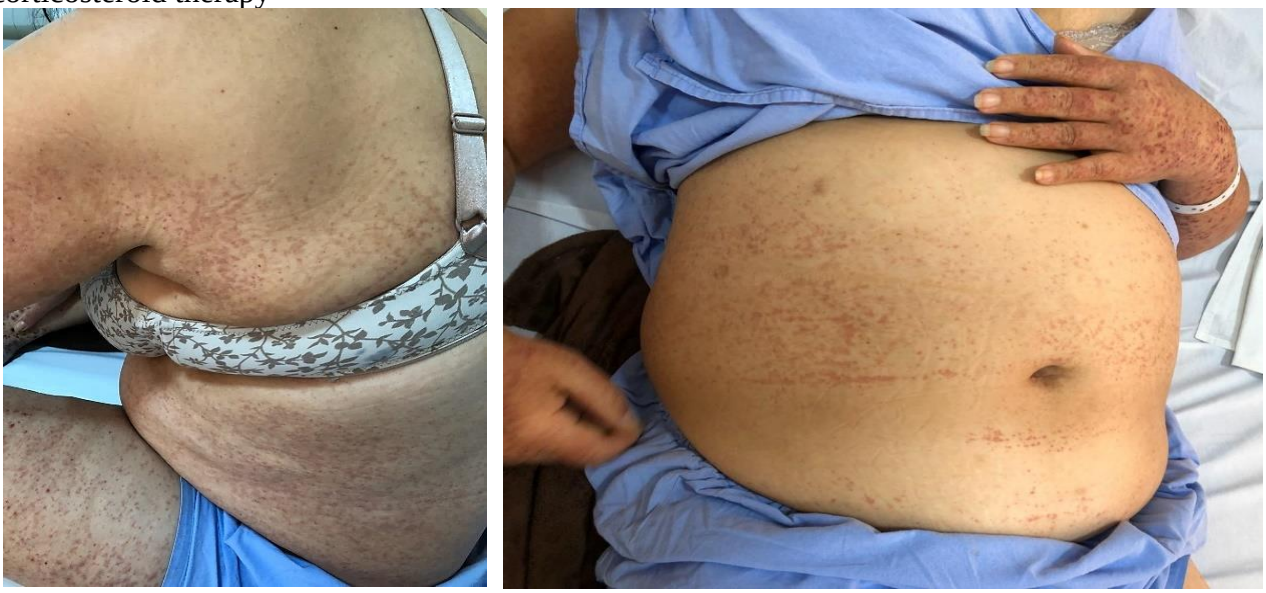


Source: the authors.

It was requested an evaluation and the opinion of a hematologist, who made a clinical diagnosis of immunoglobulin A vasculitis (IgAV) and requested the exams listed in Table 1 to

exclude other differential diagnoses. Antibiotic therapy with intravenous Ciprofloxacin was maintained for seven days. From the second day of hospitalization, treatment with Prednisolone 20 mg/day (non-immunosuppressive dose) was started until the sixteenth day of hospitalization, evolving with a good therapeutic response, without complications during this period, in addition to the regression of all skin lesions leaving some hypochromic macules and other crusted ones seen below in Figure 2.

Figure 2 - Patient with hypochromic macules and crusts on the left lower limb after 14 days on corticosteroid therapy



Source: the authors.

Table 1 - Laboratory tests

Test	Reference Values	Date (MM/DD/YY)				
		08/06/19	**08/07/19**	**08/08/19**	**08/12/19**	**08/19/19**
Erythrogram	RBC 4 – 5.5 Ht 37-47 Hb 12-16 Plat 150,000 – 400,000	RBC 4.18 Ht 27.7% Hb 9.2 Plat 324,000	RBC 4.62 HT 30.7% Hb 9.0 Plat 369,000	RBC 3.8 Ht 26% HB 8 Plat 271,000 OBS: anisocytosis +, microcytosis +, e hypochromia +	RBC 4.21 Ht 28% Hb 8.3 Plat 394,000	RBC 4.4 Ht 29.3% Hb 8.8 Plat 417,000
Leukogram	Leuco 4,500 – 10,000 Bast 0-100 Seg 2250 - 6000	Leuco 6,500 Bast 1% Seg 74%	Leuco 6,410 Bast 4% Seg 70%	Leuco 4,500 Bast 2% Seg 61%	Leuco 9,540 Bast 2% Seg 78%	Leuco13,680 Bast 1% Seg 63%
Protein electrophoresis	Albumin 3.5 – 4.85; Alpha1: 0.22 – 0.4 Alpha2: 0.55 – 1.08 Beta1: 0.32 – 0.54 Beta2: 0.24 – 0.54 Gama 0.74 – 1.75 Total ptn: 6.5-8.2			Albumin 2.89; Alpha1: 0.30; Alpha2: 0.72; Beta1: 0.49; Beta2: 0.40; Gama: 1.51; Relation A/G:0.85 Total ptn: 6.30 Absent monoclonal PTN		

Notes: red blood cells (RBC); hematocrit (Ht); hemoglobin (Hb); platelets (Plat); leukocytes (Leuco); rods (Bast); segmented (Seg); non-reactive (NR).
Source: the authors.

Table 2 - Urinalyses Type I

	Reference Values	Result 08/08/2019	Result 08/19/2019
Urinalysis	Citrine yellow color, clear, no deposit, PH 1015-1025, negative nitrite, rare epithelial cells, Leuco up to 10/field, red blood cells up to 8/field, absence of mucus filament.	Dark yellow, cloudy, Deposits +++, nitrite -, proteins ++, Hb +++, mod. epithelial cells, Leuco >50, Countless red blood cells, mucus filament ++, increased bacterial flora.	Citrine yellow, cloudy, deposits present, density 1020, nitrite +, proteins +, bilirubin absent, hb +, frequent epithelial cells, leuco > 100000, red cells 25mil/ml, mucus filaments present ++, intense bacterial flora, crystals absent, leukocyte cylinders. Presence of renal epithelial cells.

Source: the authors.

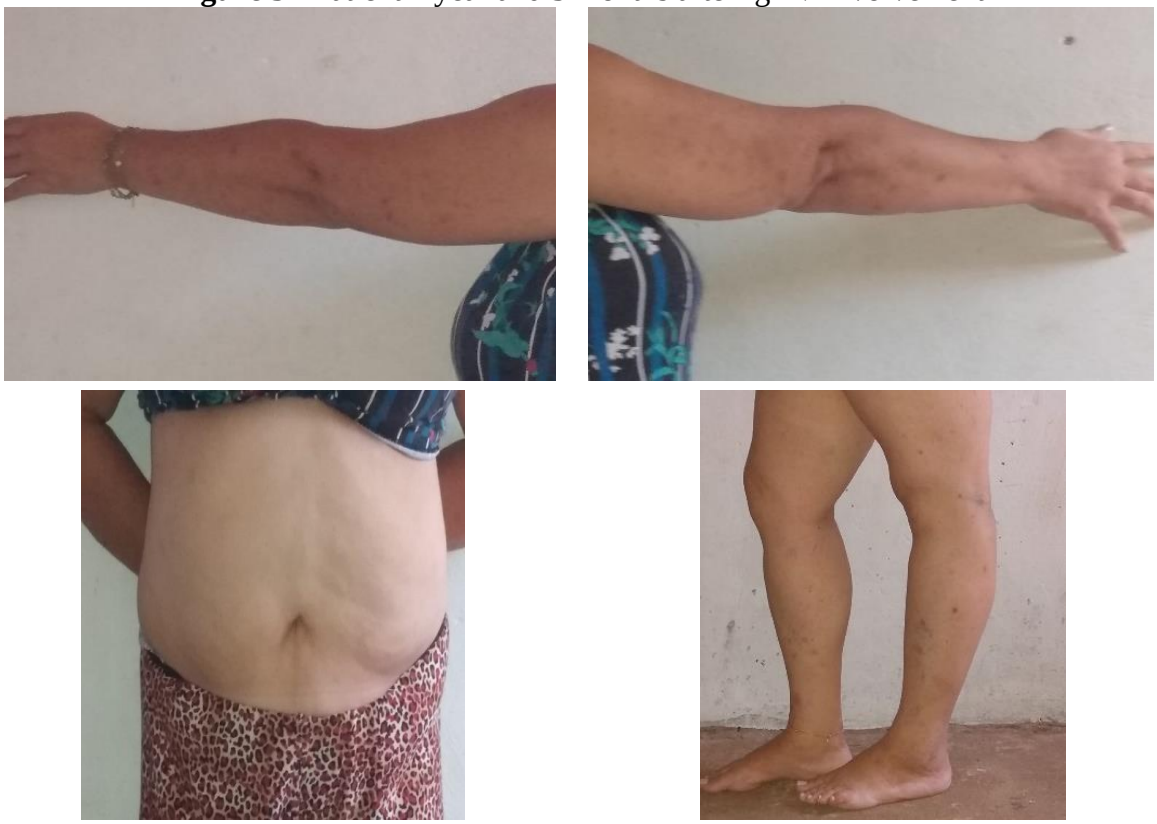
Table 3 - Serologies

	Reference values	Result
Latex Rheumatoid Factor	> 8 positive	< 8,0 (Negative)
Syphilis (total and specific CA)	Non-reactive	Non-reactive
Antinuclear Factor	Nucleus, Nucleolus, Cytoplasm, mitotic, Metaphase plate - All non-reactive	Core + reagent + metaphase plate fine dotted pattern - Nucleus, cytoplasm and mitotic apparatus NR
HBsAg	NR	NR
Anti-HIV	NR	NR
Anti-HCV	NR	NR
ASO	+ >200	135

Source: the authors.

There was a period of 1 year and 3 months of follow-up to check the patient's health status and whether there were new episodes of IgAV exacerbation. In the meantime, there were no clinical or cutaneous manifestations, as can also be seen in Figure 3.

Figure 3 - Patient 1 year and 3 months after IgAV involvement



Source: the authors.

DISCUSSION

IgAV is often diagnosed clinically. It is known that there are no specific diagnostic tests to assess or diagnose it. However, general laboratory tests can be done and even be useful to exclude other diagnoses and assess the impact of vasculitis in other systems. Because IgA has an essential involvement in the etiopathogenesis of IgAV, it is interesting to measure the serum level of IgA. Serum laboratory data, particularly normal platelet count and coagulation studies, will help to rule out two differential diagnoses: idiopathic thrombocytopenic purpura and thrombotic thrombocytopenic purpura (PILLEBOUT; SUNDERKÖTTER, 2021).

It is currently known that the accepted classification criteria of the European League Against Rheumatism (EULAR) to establish the diagnosis require the presence of purpura and at least one of the following four clinical aspects: abdominal pain, arthritis, renal involvement, or biopsy histology reflecting leukocytoclastic vasculitis or IgA deposition. Nevertheless, it is necessary to understand that this classification has not been validated or designed for application in adult populations (YONG; LEE; TAY, 2015). Interestingly, the patient in this case presented, in addition to the lesions in the form of purpura, renal involvement on the urinalysis performed after her admission to the hospital, demonstrating the following findings: the presence of deposits; of positive nitrite; of present proteins; microscopic hematuria; considerable leukocyturia; intense bacterial flora; leukocyte casts; and presence of renal epithelial cells.

In patients with systemic IgAV or kidney-limited IgA nephropathy (IgAN), IgA1 in serum and tissue deposits reduced glycosylation in the terminal region (SUZUKI *et al*., 2011). In addition, there is also emerging information demonstrating that patients with IgAV and IgAN have abnormally glycosylated IgA1 in circulation and possibly glycan-specific IgG antibodies that form anti-IgA1 immune complexes IgA1-IgG (SUZUKI *et al*., 2009). IgG antibodies directed against abnormal glycosylation bind respectively to IgA1 molecules and are found in the vessel walls, causing inflammation (JENNETTE *et al*., 2013). Analogous to this, our patient presented a urinalysis with alterations, which may determine probable renal involvement, as mentioned above.

As with other vasculitides, IgAV can manifest as a single organ vasculitis. The unique form of cutaneous IgAV is analogous to IgAN without a systemic disease, although patients with kidney-limited IgAN or single-organ cutaneous IgAV may later develop systemic IgAV. It can be associated with and probably caused by other diseases such as liver disease, inflammatory bowel disease, and spondylitis. The onset of symptomatic IgAV is often associated with a GIT or upper respiratory tract infection (JENNETTE *et al*., 2013).

Commonly, for rare skin complications such as hemorrhagic bullous lesions, corticosteroid immunosuppression is continuously helpful to control inflammation and limit the extent of necrosis and subsequent scarring. Typically, this treatment causes a dramatic improvement in clinical symptoms, observed within a few days (BOER *et al*., 2010; DHANJAL *et al*., 2017). Despite this, there is no evidence of the efficacy of corticosteroids on renal or gastrointestinal prognosis and the duration of purpura. In the presence of renal involvement, specific therapy is relevant (HUBER *et al*., 2004; WEISS *et al*., 2007; HAHN *et al*., 2015). In fact, the patient in this report evolved well with mitigation of diffuse skin lesions after using corticosteroid therapy in non-immunosuppressive doses for fourteen days.

In turn, another case report showed a 15-year-old male patient with bullous IgAV who presented severe abdominal pain and hemorrhagic bullous lesions in the lower limbs. He was treated with a corticosteroid and his symptoms improved dramatically after use. No side effects were identified after follow-up, although there were only scars. In this same report, a review of the literature related to bullous IgAV was made and 39 cases were found, most of which were treated with corticosteroids since 1985 (HUNG-WEN; CHIU-YU; YEE-HSUAN, 2018).

However, there is a case report of an 11-year-old female child diagnosed with IgAV who developed severe skin lesions and did not respond well to standard corticosteroid therapy after one month of use. As a result, she was treated with intravenous immunoglobulin (IVIG) (2 g/kg), in

addition to oral prednisone (1 mg/kg/daily), the latter being progressively reduced. This combination induced rapid and persistent resolution of symptoms within 90 days of follow-up (MAURO *et al*., 2019).

Two studies reported the use of azathioprine with corticosteroids in two patients due to uncontrolled skin lesions and progressive heavy proteinuria (TRAPANI *et al*., 2010; MEHRA *et al*., 2014).

We see a case report of IgAV in a 5-year-old schoolboy whose biopsy confirmed leukocytoclastic vasculitis with IgA deposition inside the walls of small vessels in line with the presence of severe and chronic (more than 6 months of evolution) cutaneous manifestations that were resistant to the use of corticosteroids. In the meantime, the corticosteroid therapy was discontinued and the use of colchicine (1 mg/day, administered orally) was chosen, which was introduced 10 months after the onset of the disease, with a marked improvement in skin manifestations within a few days. Consequently, no adverse effects were seen in the following 3 years and the treatment with colchicine was finally stopped when the patient was 8 years old, without any recurrence in the 6 months of follow-up (SLIMANE *et al*., 2016).

In contrast, in adults, colchicine is recommended as first-line therapy for chronic or recurrent cutaneous leukocytoclastic vasculitis, but the only randomized controlled trial with published results showed no significant effect (SAIS *et al*., 1996). However, colchicine was also used in a patient for a carrier of chronic hepatitis B (CHAN; TANG; LO, 2007).

In contrast, one study reviewed published case reports of IgAV treated with dapsone and compared them with 2 similar cases they found. Seventeen patients (between 22 months and 16 years of age) with severe or persistent clinical signs of IgAV were included. Dapsone has shown good results in the resolution of skin lesions, but not in renal manifestations. Complications (methhemoglobinemia) were observed in 1 patient. Half of the patients relapsed after discontinuing treatment. The difference between the time interval before initiation and the duration of treatment was not significant. Therefore, it was suggested in this study that dapsone may have a positive effect on chronic IgA vasculitis when cutaneous manifestations last longer than 6 weeks at a dose of 1-2 mg/kg once a day for one week (ROMAN *et al*., 2019). Similarly, we identify three case reports of children, aged 4, 5, and 16 years, diagnosed with IgAV associated with infection by *Mycoplasma pneumoniae* or *Chlamydia pneumoniae*. In all cases presented, persistent cutaneous manifestations and abdominal pain were resistant to antibiotics and corticosteroids. Despite this, they were resolved 48 hours after the introduction of dapsone and no adverse effects of the treatment were observed. Thus, the administration of dapsone should be considered in the cutaneous forms of IgAV (VOLEJNIKOVA; HORACEK; KOPRIVA, 2018).

CONCLUSION

The patient in this study evolved with clinically compatible immunoglobulin A vasculitis (IgAV) in the form of probable cutaneous involvement by lesions in the form of purpura and renal involvement, in addition to a good therapeutic response to corticosteroid therapy at non-immunosuppressive doses and after 1 and 3 months of follow-up, there was remission of the condition without new episodes. There are some reports in the literature about the good therapeutic response of this clinical with corticosteroid therapy at non-immunosuppressive doses, however, clinical trials are needed.

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