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ORIGINAL ARTICLE

Indications and efficiency of dapsone in IgA vasculitis (Henoch-Schonlein purpura): case series and a review of the literature

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Abstract

Immunoglobulin A (IgA) vasculitis (Henoch-Schonlein purpura (HSP)) is the most common vasculitis in children. It is characterized by purpuric rash, arthritis, gastrointestinal, and/or renal involvement. Spontaneous resolution is the typical outcome. In chronic cutaneous manifestations of IgA vasculitis, dapsone seems to show a good effectiveness. Multiple case reports and case series about dapsone in chronic IgA vasculitis are available. However, no clear evaluation of its indications, its effectiveness, or its usage guidelines (optimal dosage or duration of treatment) is available. We reviewed the published cases of IgA vasculitis treated by dapsone and compared them with 2 similar cases that we encountered. Seventeen patients (ranging from 22 months old to 16 years old) with severe or persistent clinical signs of IgA vasculitis were included. Dapsone showed good results on the resolution of cutaneous lesions but not on renal manifestations. Complications (methemoglobinemia) were observed on 1 patient. Half of the patients relapsed after treatment discontinuation. The difference between the time lapse before initiation and the duration of the treatment was not significant.

Conclusion: We suggest that dapsone can have a positive effect in chronic IgA vasculitis when cutaneous manifestations last more than 6 weeks at the dosage of 1–2 mg/kg once per day during 1 week.

What is Known:
- *IgA vasculitis or Henoch-Schonlein purpura is the most common vasculitis in children and affects mostly small vessels of the skin, kidney, and gastrointestinal tract. It resolves spontaneously in most of the cases. Exceptionally, cutaneous lesions can last several weeks.*
- *Dapsone is a bacteriostatic antibacterial sulfonamide drug found to be effective in the treatment of some inflammatory dermatological diseases like IgA vasculitis.*

What is New:
- *Dapsone is effective against chronic purpuric lesion (> 6 weeks) at the minimal dose of 1 mg/kg/day.*
- *Relapse occurs frequently after discontinuation but responds after a second course of treatment. A longer duration of treatment or a delay in treatment by dapsone does not seem to influence the relapse rate.*

Keywords Dapsone · Henoch - Schonlein purpura · IgA vasculitis · Child

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Abbreviations

ACE	Angiotensin-converting enzyme
CRP	C-reactive protein
DIF	Direct immunofluorescence
HSP	Henoch-Schonlein purpura
IgA	Immunoglobulin A
NSAIDs	Non-steroidal anti-inflammatory drugs

Introduction

IgA vasculitis is formerly called Henoch-Schonlein purpura. This vasculitis is the most common one found in children. Several theories about environmental and genetic factors that could explain its etiology are discussed in [15]. The IgA Fc receptor seems to play a part in this etiology [6]. The pathogenesis implies the formation of circulating IgA-containing complex. Epidemiological studies have shown an annual incidence of 10–22/100,000 children. Boys are usually more afflicted with a mean age of 6 years. IgA vasculitis is a multisystemic disease affecting skin, joints, kidneys, and/or gastrointestinal tract. However, other organs can be involved. Diagnosis requires the presence of palpable purpura associated with at least one of the following criteria: diffuse abdominal pain, arthritis or arthralgia, renal involvement (hematuria, proteinuria), or biopsy with predominant IgA deposition. Sign or history of upper respiratory tract disease can precede the initial IgA vasculitis onset by 1 or 2 weeks. Most cases of IgA vasculitis were resolve spontaneously. Only one-third of the patients are experiencing recurrence. The long-term prognosis depends on the occurrence of renal involvement [17, 19].

Early corticosteroid treatment is often used to reduce abdominal pain or renal involvement. Nevertheless, there is no evidence of steroid effectiveness on the renal or gastrointestinal prognosis and on the duration of the purpura. In the presence of renal involvement, specific therapy is required [5, 7, 21].

Exceptionally, cutaneous lesions can last several weeks. In such situations, we talk about chronic purpura, although it is not clearly defined. Dapsone and colchicine have been tried in some case reports but the effectiveness was never measured accurately and the risk of purpura recurrence after treatment discontinuation seems high. Anti-leukotriene agents and anti-CD20 monoclonal antibody seemed also to be effective on purpuric lesions in very small case series [1–3, 8, 12, 18, 20].

The aim of this paper is to evaluate the effectiveness of dapsone on chronic cutaneous lesions in HSP. We also compared dosage and duration of dapsone treatment by reviewing case reports.

Methods

We searched through PubMed with the following keywords: "IgA vasculitis" OR "HSP" OR "Henoch Schonlein" OR "Rhumatoid Purpura" associated to the keyword "dapsone" AND "children." We obtained 14 articles. Ten were excluded because 3 did not discuss dapsone's therapy, 2 were about hypocomplementemic urticarial vasculitis syndrome or allergic vasculitis, 2 were about palpable purpura, 1 was not a case series or a case report, 1 was focused on individuals older than 18 years, and 1 was lacking data. We included all case series and case reports concerning at least 1 child treated with dapsone for IgA vasculitis. IgA vasculitis was defined following the EULAR/PRINTO/PReS criteria as the presence of palpable purpura, with lower limb predominance associated with at least one of the following criteria: diffuse abdominal pain, arthritis or arthralgia, renal involvement (hematuria, proteinuria), or biopsy with predominant IgA deposition [13, 20]. If diagnosis did not fulfill these criteria or if there were uncertainties about the diagnosis due to the lack of data, the article was excluded (Fig. 1).

We analyzed demographic data, clinical manifestations, biological results (if available), disease duration before treatment by dapsone, the reason why it was started, the treatment itself (dosage, duration of therapy, adverse events…) and its effectiveness on the multiple manifestations of the disease.

Data availability The datasets used and/or analyzed are available as supplementary material.

Results

Case report 1

A 5-year-old boy developed a brutal abdominal pain and palpable purpuric rash on his legs leading to the IgA vasculitis diagnosis. All of the additional test (urine analysis, blood

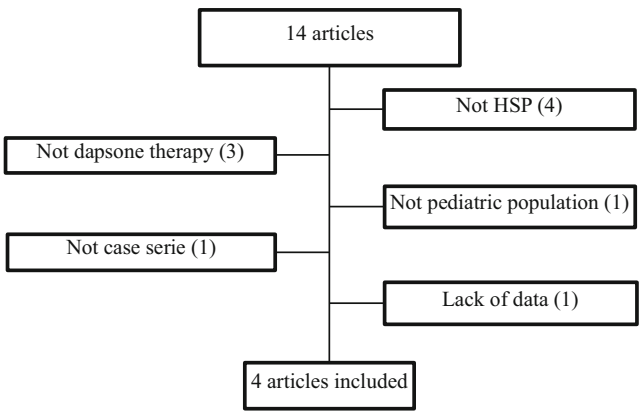

Fig. 1 Flowchart of literature review

sampling, and blood stool test) were normal. CRP was normal. He was admitted to the hospital 4 days after the first symptoms.

Due to severe abdominal involvement, intravenous steroids (1 mg/kg/day) were started. Despite the steroids, abdominal pain got worse and purpura extended to the legs, genital areas, and buttocks. Macroscopic hematuria also appeared. Abdominal ultrasonography showed multiple small bowel wall hematomas and spontaneously resolving ileoileal intussusceptions. On the eighth day, gastrointestinal bleeding appeared. Steroids were then increased to 2 mg/kg per day. He then stopped oral feeding and started parenteral nutrition.

Bilateral knee, ankle, and wrist arthritis appeared but resolved rapidly with non-steroidal anti-inflammatory drugs (NSAIDs). During this period, epididymitis occurred twice and resolved as well using NSAIDs. Abdominal manifestations improved after a few weeks. Corticosteroid could then be tapered.

After 2 months of evolution, slight proteinuria was detected (maximum 0.53 mg/mg creatinine). Purpuric rash on the legs remained persistent and painful. Skin lesion biopsy confirmed the leukocytoclastic vasculitis but IgA deposition could not be found by direct immunofluorescence (DIF). Negative DIF could be due to the sampling method. Moreover, IgA deposit is not required for IgA vasculitis's diagnosis [11, 13, 14].

We started dapsone (2 mg/kg daily) 7 weeks after the initial onset. Purpura resolved rapidly 7 days after the treatment was started. ACE inhibitors were started at the same time for renal involvement. Proteinuria disappeared after a few days.

Dapsone was progressively tapered after 6 months. He unfortunately relapsed with cutaneous manifestations 10 months after dapsone discontinuation. There was no other organ involved, including kidneys. After starting dapsone again with 2 mg/kg during 2 weeks, every lesion finally disappeared again without any relapse.

Case report 2

A 3-year-old girl developed an extensive purpuric rash on legs, elbow, abdomen, and buttock. She also complained about brutal abdominal pain and joints edema.

Abdominal pain persisted without hematochezia. Abdominal ultrasonography was performed and was normal. Abdominal pain and arthritis resolved spontaneously after 5 days without corticosteroids.

There was no renal involvement.

After 7 weeks, palpable purpuric rash persisted on the legs. A therapy by dapsone (2 mg/kg daily) was started 8 weeks after the initial purpuric rash. It disappeared rapidly in 10 days without further complications.

Dapsone was progressively tapered after 2 months without any relapses 8 months after dapsone discontinuation.

Case reports in the literature

Iqbal et al. [8] describes eight children treated with dapsone in a pool of 41 patients diagnosed with IgA vasculitis from January 1992 to May 2004. Those eight children have a severe or persistent purpuric rash. The median age is 8 years old. The treatment was started between 5 days and 14 months after the initial clinical signs with a daily dose ranging from 0.5 to 1.3 mg/kg. The first treatment's response appeared between 3 and 7 days under dapsone. But 6 of the 8 children relapsed after the discontinuation of dapsone. The duration of treatment was extremely variable: from a few days to 2 years. There was no information regarding the use of other therapies.

Mazille et al. [12] describes three teenagers with similar results. The initiation of dapsone treatment was limited to chronic purpuric rash (defined as more than 6 weeks). The median age of the patients was 15 years old. One teenager was first treated by corticosteroid without effect. The purpuric rash disappeared rapidly after the initial treatment but relapses were common when the dapsone was stopped, even if dapsone was continued during months. One teenager (16 years old) presented a methemoglobinemia. It is a well-known complication of dapsone. This complication is usually dose related (> 2 mg/kg/day) but this was not the case in this patient (daily dose 1.3 mg/kg). No effect on renal involvement was observed.

Chen et al. [3] describes an adolescent girl of 14 years old with bullous hemorrhagic IgA vasculitis. C-reactive protein (CRP) was mildly elevated (15 mg/dl). Severe purpuric rash was treated for the first time after 3 weeks of evolution by steroids and doxycycline. After 7 days of treatment, the lesions improved. Dapsone was introduced 10 days after the initial treatment to reduce steroids use. The purpuric rash disappeared after 4 weeks of bi-therapy by dapsone and steroids. Renal involvement persisted and was treated by angiotensin-converting enzyme (ACE) inhibitors. There is no information on relapse or complication.

Volejnikova et al. [20] describes three children treated by dapsone for recurrent or persistent purpura and abdominal pain. The median age was 5 years. All children presented an atypical bacterial infection associated with IgA vasculitis. CRP was moderately high in all cases (max 37 mg/dl). The treatment was started between 2 weeks and 6 months after the initial clinical signs. One patient was first treated by corticoids for abdominal pain. One child first received dapsone (0.5 mg/kg/day) without result. He was then treated by dapsone 1 mg/kg/day, as were the others. Using this dosage, all purpuric rashes disappeared in a few days. One child relapsed four times after treatment discontinuation. No complication was mentioned.

We found 17 children treated by dapsone with persistent or severe IgA vasculitis purpuric rash.

All infants had a persistent or severe rash. Arthritis or joint pain occurred in 70% of patients, abdominal involvement in 82%, and renal involvement in 47%. Abdominal involvement

was frequent: 100% in our study, 87.5% in Iqbal's study [8], 100% in Mazille's study [12], and 67% in Volejnikova's study [20]. IgA vasculitis bullous of Chen et al. [3] did not present abdominal manifestation. Renal involvement was present in 50% of our patient compared to 75% in Iqbal's study [8], 100% in Chen's study [3], and none in Mazille and Volejnikova's study [12, 20]. All the cases are described in Table 1.

Median age of patients in the five groups was significantly different because of Mazille and Chen's cases [3, 8]. These studies were based on adolescents, whereas the other groups studied children. No differences were found between the five studies in terms of clinical manifestation or relapse's frequency.

In the cases described, 65% of children were girls. The median age was 8.7 years old (centile 25 = 5 years; centile 75 = 14.5 years). A comparison of the clinical signs displayed by the studied patient and by the general population of IgA vasculitis–afflicted patient is detailed in Table 2. Abdominal manifestations were more frequent in chronic IgA vasculitis. Girls were more affected than in general population.

Rash was clearly the reason of treatment by dapsone in 9 children. In the study of Volejnikova, abdominal involvement was also a reason of treatment by dapsone in 2 children. In the study of Iqbal, the reason of starting a treatment by dapsone is not described completely.

Median duration of clinical signs before dapsone's initiation was 2 months (quartile 1 = 0.4 months, quartile 4 = 6 months). Nine patients (53%) were treated after a symptom progression of at least 6 weeks. Three (18%) received dapsone after 4 weeks of evolution. Four patients in Iqbal's study were treated within the first week of symptoms. Mean dose of dapsone was 1.2 mg/kg/day. Our dose of dapsone (2 mg/kg/day) was significantly higher than in the other group. Total duration of first course mean was extremely variable between 4 days and 6 months. Duration of first course of treatment by dapsone was shorter in Iqbal's case series (Table 3).

Cutaneous vasculitis responded in all cases (100%). Renal involvement did not respond in all cases. Relapses occurred in 53% of patients. The mean age is not significantly different when comparing the group who relapsed with the one who did not ($p = 0.83$). The time between the beginning of the disease and the dapsone's initiation is not significant between both groups either ($p = 0.63$) (Table 4).

No significant variation was found in the frequency of relapse regarding the duration of the dapsone treatment ($p = 0.5$). Four patients were treated ≤ 7 days and two of them relapsed (50%) compared with the 12 patients treated more than 7 days who relapsed in 58% (7/12) of cases.

In our cases, patients were followed during 8 and 12 months. The follow-up in Iqbal's study [8] was not indicated. In Mazille [12] and Chen [3], patients were followed between 6 and 12 months. In Volejnikova [20], one patient was followed during at least 1 year. This patient relapsed four times in 1 year without a second course of dapsone. In all other cases, after a second treatment by dapsone, all purpuric lesions disappeared again.

We noted one patient (6%) with a complication (methemoglobinemia), which does not seem to be dose-related.

Discussion

In children, IgA vasculitis appears mostly in young boys. It is more frequent around 5–6 years old. In our report, chronic purpuric lesions are significantly more frequent in girls. Median age is 8 years old. Abdominal involvement is also

Table 1 Demographic characteristics, presenting clinical feature, and treatment of the patients

	Our cases	Iqbal et al. [8]	Mazille et al. [12]	Chen et al. [3]	Volenjnikova et al. [20]
Number of cases	2	8	3	1	3
Age median (years)	4 (3, 5)	8 (5, 10)	15 (15, 16)	14	5 (4, 16)
Clinical					
Rash	2/2 (100%)	8/8 (100%)	3/3 (100%)	1/1 (100%)	3/3 (100%)
Joint	2/2 (100%)	6/8 (75%)	3/3 (100%)	1/1 (100%)	0/3
Abdominal	2/2 (100%)	7/8 (87.5%)	3/3 (100%)	0/1	2/3 (67%)
Renal	1/2 (50%)	6/8 (75%)	0/3	1/1 (100%)	0/3
Other organs	1/2 (50%)	1/8 (12.5%)	0/3	0/1	0/3
Presentation to treatment	7 to 8 weeks	5 days to 18 months	6 months	1 months	1 to 6 months
Dose	2 mg/kg/day	0.5 to 1.3 mg/kg/day	1.3 to 1.4 mg/kg/day	25 to 50 mg/day	0.5 to 1 mg/kg/day
Total duration of treatment (days)	60 to 180	4 to 28	21 to 365	> 30	28 to 35
Response	Yes	Yes	Yes	Yes	Yes
Relapse after discontinuation	1 (50%)	6 (75%)	1 (33%)	No	1 (33%)
Complication	No	No	Yes	No	No

Table 2 Comparison of clinical signs in our cases and IgA vasculitis in general population

	Case series	Population of IgA vasculitis [19]	*p* value
Age (years)			
Median	8 (5, 14.5)	5.3	
Mean	8.7 (±4.8)	6.1 (±2.7)	0.0008
Sex ratio (boys/girls)	1/1.5	1.8/1	
Boys (%)	35.3% ($n = 17$)	63% ($n = 150$)	0.05
Clinical manifestation			
Rash	100% ($n = 17$)	100% ($n = 150$)	NS
Joint	70% ($n = 17$)	74% ($n = 150$)	NS
Abdominal	82% ($n = 17$)	51% ($n = 150$)	0.03
Renal involvement	47% ($n = 17$)	54% ($n = 150$)	NS

significantly more frequent (84%) compared with the general IgA vasculitis population (51–73%). Arthritis or joint pain occurred in 70% of patients and renal involvement in 47% [17, 19].

A chronic purpuric rash in IgA vasculitis is rare. There is no clear definition of "chronic purpuric rash." We know that IgA vasculitis usually resolves spontaneously in 4 weeks and recurrence happens in one-third of the patient but subsides after 4 or 6 months [19]. Most authors agree to define a chronic purpuric rash as a rash lasts more than 6 weeks. When purpuric rash is persistent, dapsone and colchicine are the most commonly used treatment [1].

Dapsone is a bacteriostatic antibacterial sulfonamide drug found to be effective in the treatment of inflammatory dermatological disease. It was first used with IgA vasculitis in 1983 in patient with chronic purpura [9]. The exact mechanism of action in this indication is not clearly understood. We know that dapsone has anti-inflammatory properties and antioxidant scavenger effect. Dapsone inhibits chemotaxis and function of neutrophils. Dapsone inhibits the release of interleukine-8 and the production of leukotriene B4. Dapsone inhibits the generation of secondary messengers essential to the activation of beta2-integrin molecules and suppresses the generation of toxic free radicals in neutrophils by its action on the beta2-integrin [1, 4, 16, 22, 23].

In these five series of cases, 17 patients were treated by dapsone due to the severity or the persistence of purpuric rash in IgA vasculitis. Symptom duration before dapsone's initiation was approximatively 2 months compared with general population with IgA vasculitis spontaneously resolving in 4 to 6 weeks.

All chronic or severe purpuric lesions disappeared after a treatment by dapsone within a few days. Other symptoms did not respond. Dapsone is used in many dermatological diseases. The effects of dapsone on cutaneous lesions seem to be by its action on interleukine-8 and by suppressing free radical's secretion. Interleukine-8 secretion seemed to be increased in IgA vasculitis which could explain its effect on skin lesions in IgA vasculitis [10, 22].

In our observations, relapses occurred frequently (53% of cases) but do not seem to be related to the age of the patient, to the time between onset of disease and dapsone initiation or to the dose of dapsone used. After a second course of treatment by dapsone, all purpuric lesions disappeared again. Studies with a larger number of patients are needed to determine the reasons of relapses.

The duration of treatment was variable between cases from 4 days to 6 months. A longer total duration of treatment did not seem to be more effective on relapses but this could be due to a limited number of patients in this study. Randomized controlled trials should be initiated to study dapsone treatment in IgA vasculitis and confirm that a short course of dapsone is as effective as several weeks of treatment.

The most common adverse events are the hematological side effects such as hemolysis, methemoglobinemia, and

Table 3 Comparison of dapsone use in 5 studies

Mean of	Our cases	Iqbal et al. [8]	Mazille et al. [12]	Chen et al. [3]	Volejnikova et al. [20]	*p* value
Presentation to treatment (months)	2	5	7	1	3	NS
Dose (mg/kg/day)	2	1	1.3	/	1	< 0.001
Duration of treatment (days)	120	11	127	/	35	< 0.005

Table 4 Comparison of patients with and without relapse

	Relapse ($n = 8$)	No relapse ($n = 5$)	p
Age (mean)	8.1 years (± 3.6)	9.6 years (± 6)	NS
Presentation to treatment (mean)	5 months (± 8)	3.5 months (± 2.7)	NS
Dapsone dosage (mean)	1.12 mg/day	1.38 mg/day	NS
Duration of first course (mean)	35 days (± 55)	65 days (± 83)	NS

agranulocytosis. Only one patient had a methemoglobinemia. Before starting therapy, it is recommended to make a complete blood count, hepatic and renal function study, urine analysis, and to look for glucose-6-phosphate dehydrogenase level to prevent side effects [4, 16, 23].

Limitations

We analyzed data of cases series with a low number of patients. Duration of treatment, reason of initiation, and symptom duration before dapsone's initiation were extremely variable. Case reports are more often published when there is a positive response. All these conditions can influence our results.

There are no randomized controlled trials for treatment of chronic cutaneous lesions in IgA vasculitis by dapsone but only published case reports. Randomized controlled trials should be initiated to study dapsone treatment in IgA vasculitis and confirm its effectiveness. Randomized controlled trials are also necessary to determine the ideal dosage and duration of treatment.

Conclusion

These cases illustrated the benefit of dapsone therapy on chronic cutaneous lesions in IgA vasculitis encouraging a widespread use in severe or painful case. Dapsone seems to be effective against chronic or recurrent purpuric lesion in IgA vasculitis with purpura disappearing within a few days. The exact mechanism of action in this disease is not completely understood but is probably due to the inhibition of chemotactism of neutrophil or to its action on interleukin-8, beta-2 integrin, and leukotriene B4. Dapsone has a suspensive effect more than a curative effect. The side effects (methemoglobinemia and hemolysis) are rare. Knowledge of the liver function and a complete blood count of patient are recommended before initiating treatment.

We suggest using dapsone in IgA vasculitis when severe or painful purpura lesion did not disappear after 6 weeks at a dose of 1 to 2 mg/kg/day for a duration of 7 days. We want to emphasize the need of randomized control trials to confirm the best dosage and the duration of treatment in this indication.

If a relapse occurs after dapsone discontinuation, we suggest treating again with dapsone for a duration of 7 days. Relapses occur rarely after a second course of treatment.

Authors' contributions C.R. and OG wrote the manuscript. C.R., B.D., L.M., T.S. and O.G. contributed to collect clinical data and to the analyzis of data. O.G. supervised the work.

Compliance with ethical standards

Conflict of interest The authors declare that they have no competing interests.

Ethics approval and consent Not applicable.

Consent for publication Not applicable.

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