Damage assessment in adult IgA vasculitis. Cross-sectional results of a multicentre cohort

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ABSTRACT

Objective. Immunoglobulin (Ig) A vasculitis affects children more commonly than adults and previous literature lacks any formal damage assessment. Our aim in this study is to investigate the disease course, relapse rates and prognostic factors in adults with IgA vasculitis and to evaluate the disease-related damage.

Methods. We assembled a retrospective cohort of adult IgA vasculitis from six tertiary Rheumatology Centres in Turkey. The demographics, clinical characteristics, treatment and outcomes of patients were abstracted from medical records.

Results. The study included 130 (male/ female: 85/45) patients and the mean age was 42.2±17 years. Cutaneous manifestations and arthritis/arthralgia were the most common clinical manifestations. One hundred thirteen patients (86.9%) were treated with oral glucocorticoids (GC). As additional immunosuppressive (IS) agents, azathioprine was given to 44 (34.9%) and pulse cyclophosphamide to 18 (12.6%) patients. Seventy-nine patients (60%) had follow-up of median 15 (IQR 7-40) months. Twelve (15%) patients relapsed during follow-up. The mean VDI score was 0.4 in the last visit. Nineteen (24.7%) patients had at least one damage item at the end of follow-up. Most frequent damage items were renal 11 (42%), ocular 4 (15%) and cardiovascular 4 (15%).

Conclusion. In this cohort the most frequent damage item was renal and was related to the disease itself. Damage score was higher in patients with more severe disease and treated more aggressively. Our results suggest that more effective treatment options are needed in a subgroup of patients with IgA vasculitis to prevent the damage related with the vasculitis, especially with more severe disease.

Introduction

Immunoglobulin (Ig) A vasculitis, formerly known as Henoch Schönlein purpura (HSP), is an acute, multi-systemic, leukocytoclastic vasculitis characterised by deposition of IgA-containing immune complexes and complement components in small-vessel walls (1). The disease is characterised by palpable purpura, arthritis/arthralgia, abdominal pain and renal impairment (2). Children are affected more commonly, as it is the most common vasculitis of childhood. The incidence is 13.5-20.4/100,000 in children (3-4), which declines to 13-14 cases/100,000 in adults (5-6). The aetiology is poorly understood. IgA1 accumulation in the vascular wall and renal mesangium plays a crucial role in pathogenesis. Possible triggers of IgA vasculitis include infectious agents (especially respiratory tract infections), drugs and vaccines (7). Various genetic loci, such as MHC, IL-1r antagonist and IL-8 are implicated in the pathogenesis (5).

The prognosis is usually favourable in children, yet relapses occur with a frequency of 2.7-51.7% (9, 10). While encountered less frequently in adults, the disease tends to be more severe (11). The severity of the illness is considered to be associated with gastrointestinal and renal involvement. Earlier studies revealed that renal involvement is observed in 45 to 85% of adult-onset IgA vasculitis cases, with a 30% risk of progression to renal failure. Disease flare is usually unique and relapses occur in roughly 20% of cases in adults (12-14). The evidence is scarce about prognosis in adults and previous literature lacks any formal damage assessment. Our aim in this study is to retrospectively investigate the disease course, relapse rates and prognostic factors in adult patients with IgA vasculitis and to evaluate the disease or treatment-related damage.

Competing interests: none declared.
Methods

Patients and registry
Data were collected retrospectively from a registry of patients with IgA vasculitis, followed in 6 tertiary rheumatology clinics across Turkey. The study included 130 patients who were at least 18 years old, diagnosed with IgA vasculitis according to the American College of Rheumatology classification criteria or 2012 International Chapel Hill Consensus (1). While 124 of the patients fulfilled the 1990 ACR criteria, and 6 of them were classified according to Chapel Hill Consensus Criteria definition as IgA vasculitis.

Clinical and laboratory data
All clinical, demographic and laboratory data were acquired from the patient charts. The standard patient form including new clinical features, laboratory, biopsy and treatment changes were filed for the diagnosis, first remission and the last follow-up visits. If relapse developed, a relapse visit was also performed. Renal failure was defined as a glomerular filtration rate (GFR) of <60 mL/minute/1.73 m2 assessed with Modified Diet in Renal Disease (MDRD) equation. Proteinuria was defined as >0.3 mg/day detected with 24 hour urine collection. Haematuria was defined as ≥5 red cells/mm³. Results of skin and kidney biopsies were noted. The initial glucocorticoid dose (GC) and the additional immunosuppressive (IS) therapies and other medications during follow-up were recorded.

Definitions
Activity was assessed according to the Physician Global Assessment (PGA, 0-10) and Birmingham Vasculitis Activity Score (BVAS) (17). The absence of any symptoms or signs of clinical activity (BVAS=0) was defined as remission. The disease was considered refractory if there was no improvement in clinical signs or symptoms, or if any new clinical finding occurred.

A relapse was defined as the reappearance of clinical signs of vasculitis, occurring after a period that was free of symptoms (BVAS>0).

Major relapse was characterised by organ or life-threatening manifestations such as at least 30% increase in creatinine value, pulmonary haemorrhage, retinal vasculitis, acute limb ischaemia, gangrene, and mononeuritis multiplex. If none of the major relapse criteria were evident in a patient with relapse, the patient was classified as having a minor relapse.

The clinical findings which persisted to the second visit and at least three months were evaluated to document the damage with vasculitis damage index (VDI) (15). Five factor score (FFS); which include proteinuria >1 g/d, renal insufficiency (stabilised peak creatinine 140 µmol/L), cardiomyopathy, severe gastrointestinal manifestations and central nervous system involvement (29), were recorded in terms of the severity of the disease at baseline visit and at the relapse visits.

Statistics
We calculated the means of continuous variables and standard deviation when the data were distributed normally. Me-
diagnosed and first to third quartile interval were calculated for data that did not have a normal distribution.

We analysed group differences for dependent categorical variables with the chi-square or Fishers’s exact test. We compared continuous variables with student t-test or Mann-Whitney U when data were skewed. A two-tailed p-value <0.05 was considered significant.

**Results**

One hundred and thirty patients were included in the study. The majority of patients (n=85, 65%) were male, and the mean age at diagnosis was 42±17 years. The baseline demographic and laboratory characteristics of the patients are presented in Table I.

The most common clinical features were constitutional (n=118, 91%), cutaneous (n=129 99.2%), musculoskeletal (n=111, 85%) and gastrointestinal manifestations (n=89, 68.5%). There were two (1.6%) patients with pulmonary involvement at the baseline and they presented with pulmonary haemorrhage. In these two patients, the diagnosis of ANCA-associated vasculitis (AAV) was also excluded. Eleven (8.5%) patients had renal failure at first admission. Clinical features are displayed in Table I.

Sixty (46%) patients had proteinuria and the median level of proteinuria was 1 g/day (Q1-Q3: 0.5-2.0). Sixty (42%) patients had haematuria at baseline. ANA positivity rate was 11.8% (n= 21/181) and hepatitis B presence 7.3% (n= 9/123).

Fifty-two (40%) patients had a preceding infection within the last six weeks (40 had upper respiratory tract, 3 urinary tract, 6 gastrointestinal and 3 other infections). Forty (33%) patients had at least one kind of drug exposure (26 antibiotics, 5 non-steroidal anti-inflammatory, and 9 other).

During diagnosis, 113 (87%) patients were treated with oral GCs. Pulse GC treatment was also given to 41 (31.5%) patients. As additional IS agents, azathioprine was given to 44 (34.9%) and pulse cyclophosphamide to 18 (13.8%) patients. Twelve (9.2%) patients received colchicine, and five (3.8%) patients did not receive any treatment.

The clinical features of the patients were evaluated according to the initial treatments. Those initially treated with ISs plus GCs had more renal involvement (35% vs. 65% p=0.0001) and more GIS involvement (47% vs. 53% p=0.048) than those treated with GCs alone. Other basal clinical features were similar in the treatment groups. At the first visit mean BVAS and PGA was 6 (SD=4) and 7.3 (SD= 2.1), respectively. In the basal evaluation, there were 84 (64.6%) patients with FFS=0, 29 (22.3%) patients with FFS=1, 13 (10%) with FFS=2, and 4 (3.1%) with FFS=3.

A total of 122 patients underwent a biopsy. Of these, 97 (79.5%) had skin, 23 (19%) had renal and 2 (1.6%) had gastrointestinal biopsies. 105 of these 122 patients had histologically confirmed IgA vasculitis in biopsy specimens.

**Follow-up**

Follow-up data were available for 79 (59.2%) patients. Median follow-up duration was 15 months (Q1-Q3: 7-40). Twelve (15%) patients relapsed during follow-up. Six were major, and six were minor relapses. Nine (75%) patients were still on GC treatment during the relapse. Seven (58%) patients were under IS agents, and 5 (41%) were taking both GC and ISs during the relapse.

Factors that may be related to the relapses were evaluated in patients with follow-up data. When the basal clinical characteristics of the patients with and without relapse were compared, it was determined that patients with relapse had higher acute phase responses (p=0.041). The relapse rate was 22.5% in GC alone treatment patient group, whereas the rate of relapse was 8% in GC+IS group (p=0.069), not reaching significance. There was also no statistically significant difference in other basal clinical and laboratory characteristics between the two groups.

At the last visit, disease status was judged as active or treatment failure by the treating physician in 13 patients (15%). Among 79 patients who had follow-up data, 43 (55%) patients were still undergoing GC treatment at the last visit. Eleven (25.5%) of them received a low dose (>7.5 mg/day prednisone equivalent), and 32 (74.5%) a medium or high-dose (>7.5 mg but ≤100 mg prednisone equivalent daily) GC therapy. The rate of chronic renal failure was 8.8% (n=7). Regarding the factors related to renal damage initial renal failure (p=0.001), proteinuria (p=0.004), haematuria (p=0.005) and IS treatment (p=0.003) were found to be associated with the development of renal insufficiency.

One patient died during follow-up due to pneumonia. At the end of the follow-up, the mean VDI score was 0.4, and 18 (23.5%) patients had at least one damage item. The most frequent damage item was renal 11 (42%). The rate of damage related with GC treatment...
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is 30%, non-related with GC treatment 70%. Distribution of damage items is shown in Figure 1.

In addition, VDI scores of the patients who were followed up at the last visit were evaluated according to the initial treatments. The rate of VDI score ≥1 was 10% in patients receiving GCs alone, whereas the rate of VDI score ≥1 of patients receiving steroid plus ISs was 38.5% (p=0.005). VDI score of those receiving GC+ISs (mean: 0.71±1.1) was also higher than those receiving GCs alone (mean: 0.11±0.31) (p=0.003).

Male gender (p=0.01), older age (p=0.006), initial renal failure (p=0.012), and proteinuria (p=0.012) were more common among patients who had a VDI score ≥1.

Discussion

Although generally accepted to be a mild vasculitis, IgA vasculitis can cause permanent organ damage, especially renal failure in adults. The present study is the largest patient series of adult IgA vasculitis from Turkey and to our knowledge, the first study including an overall damage assessment with VDI for IgA vasculitis in literature so far.

Similar to the previous reports, there was male dominance and the disease onset in fifth decade in our study (13, 18-23). When we compare our patient group with two previously published large series by Calvo-Rio et al. and Audemard-Verger et al. almost all of the patients had skin involvement as in other series. Constitutional symptom rate was greater in our patients (89% vs. 33%-20%). In addition, the proportion of GIS (90% vs. 53%-64%) and musculoskeletal (85% vs. 61%-60%) manifestations were higher in our series (17, 22). The number of patients presenting with renal insufficiency was lower than the Audemard-Verger et al. study (49% vs. 70%) but similar to the other studies (30-60%). This difference may be relevant as patients were enrolled only by rheumatologists in our study, whereas the French registry included many patients enrolled by nephrologists (18-24).

IgA vasculitis is thought as a self-limiting disease, particularly in children (25). In a recent cohort from Mayo Clinic, Villatoro-Villar et al. reported that adult patients with IgA vasculitis had more frequent skin ulcers and greater frequency of severe proteinuria compared to children (11). The treatment of severe manifestations, including severe gastrointestinal complications or proliferative glomerulonephritis, is still controversial due to lack of evidence that GCs or IS agents improve long-term outcome (26). In a prospective open-label trial by Pillebout et al. and in the retrospective study of the French vasculitis group, there were no differences between GCs plus cyclophosphamide and GCs alone regarding renal outcome and mortality. The rate of achieving partial or complete remission were also similar between the groups (23, 27).

Ninety six percent of all patients in our cohort initially received GC therapy. Initially 54% of patients received GC treatment alone in this study, whereas the ratios in the series of Audemard-Verger et al. and Calvo-Rio et al. were 47% and 35%, respectively. In addition, the rate of IS+GC use was higher in our series than in other two studies (45% vs. 5%-13%) (18, 23). Additional IS therapy (mainly azathioprine) was more given to patients with renal and GIS involvement as these manifestations are accepted as more severe spectrum at baseline. A higher percentage of patients with symptomatic disease may account for a higher GC use. Additional IS treatments may be caused by a higher inclination towards GC sparing agents by rheumatologists.

Relapses can complicate IgA vasculitis. In previous reports, relapse rate ranges between 20-30% in adult IgA vasculitis. In our study, relapse rate was found in lower range as 15% (9, 18, 23). Higher use of GCs and other IS agents at baseline or a high rate of missing follow-up data may explain this finding.

Rigante et al. reported that persistent purpura is a significant predictor of relapses (28). Similarly, Shin et al. and Byun et al. observed that adult age, persistent purpura, severe abdominal pain, the presence of haematuria and severe leukocytoclastic vasculitis were associated with relapses (9, 29). In a large series (315 children and 102 adult patient) by Calvo-Rio et al., while previous infections seemed to be protective for relapses, the predictors of relapse were gastrointestinal and joint involvement in the whole group. However, when they analysed adults alone, only gastrointestinal manifestations were predictive for relapses (30). In our subjects initial acute phase elevation and constitutional symptoms increase the risk of relapse slightly. There was no other potential predictor for relapses in our group.

The most important severe manifestation of IgA vasculitis in adults is renal failure. At the last visit, the rate of chronic renal failure was 8.8%. Related factors were renal failure at presentation, proteinuria, haematuria, and usage of IS treatments. Patients who were initially given additional IS therapy were more likely to have renal involvement and renal failure more developing in these patients. Kang et al. and Pillebout et al. also found that renal insufficiency at presentation and proteinuria were associated with renal damage. (6, 12). In an another study by García-Porrúa et al., investigating factors predicting renal sequelae in adult IgA vasculitis, haematuria at disease onset and persistence of renal manifestations during the course of the disease were found to be significant indicators for renal sequelae (31). In our series, haematuria was also predictive for renal involvement. Presence of IL-1 receptor antagonist allele 2 was associated with a higher risk of severe renal manifestations and renal sequelae (32). Polymorphisms in IL-8 gene were also associated with susceptibility to renal and possibly to GI manifestations in IgA vasculitis patients (33). In addition, Hong et al. indicated that late-onset disease was a significant independent prognostic factor for chronic renal insufficiency (34). Diseases such as diabetes mellitus and hypertension are common in this age group and may affect renal failure. In our study, we did not find any association between age and renal involvement.

It is well known that FFS score ≥1 is predictor of increased mortality in sys-
nymic necrotising vasculitides such as AAV (35). In our study, very low morta-
ality rate (1.2%) was observed despite the high percentage of FFS’s.

A formal damage assessment is lack-
ing in previous studies of IgA vascul-
itis. Damage assessment is becoming a part of routine management of sys-
temic necrotising vasculitides such as PAN and AAV. In previous reports, the
mean VDI score was found 3 in AAV and 2.2 in PAN. The most frequently
positive VDI items were GC-use re-
lated damage items in PAN and AAV,
such as hypertension, osteoporosis and
cataracta (16, 36, 37). In our study, the
most common VDI item was renal
damage, despite the high rate of GC
use in the last visit. A lower rate of
GC-related damage may be explained by
a short duration of GCs and young
aged patient population. As we ex-
pected, the factors affecting VDI in-
cludes renal failure, proteinuria and a
high initial BVAS. VDI score was also
higher (mainly disease related) in pa-
tients treated IS plus GCs than patients
receiving GCs alone.

The main limitation of the study is its
retrospective design. The participants
were only from rheumatology clin-
ics, which may lead to an inclusion of
disorders with lower renal involvement
rate. A short duration of a follow-up
period and lack of follow-up data for
some patients may also limit findings
and more precise information.

In conclusion, the main cause of dam-
age in IgA vasculitis appears to be the
disease itself. Although GC treatment
is given to the majority of patients, GC-
related damage develops infrequently
in early years of follow-up. Damage
score is also higher in patients with
more severe disease and treated more
aggressively (IS+GCs). Despite the be-
nign course of IgA vasculitis and some
patients undergoing remission without
treatment, high dose GCs and addi-
tional IS agents may be required in a
small group of IgA vasculitis patients.

Our results suggest that more effective
treatment options are needed in a sub-
group of patients with IgA vasculitis
to prevent the disease-related damage,
especially in patients with more severe
disease.

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