Characteristics of an Internet-Based, International Cohort of Patients with a Self-Reported Diagnosis of Urticarial Vasculitis

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INTRODUCTION

Urticarial vasculitis (UV) is a rare disease (annual incidence of < 1 per million). This rarity makes it difficult to conduct large trials.

AIMS

To describe and validate an international cohort of patients with UV based on patient-reported data.

METHODS

- Internet-based longitudinal registry
- Enrollment period: 11/2014-2/2022
- All patients with a self-reported diagnosis of urticarial vasculitis were included
- Standardized online data collection forms
- 2012 Chapel Hill Consensus Conference (CHCC) definition of hypocomplementemic urticarial vasculitis (HUVS) met if all three of the following were present:
  a) Biopsy showing vasculitis
  b) Low serum levels of C3 or C4
  c) Any of the following manifestations: kidney, joint, obstructive lung disease, or ocular

RESULTS

Baseline Demographics	
Enrollment	102
Female (%)	92 (90)
Mean age at symptom onset (SD)	40.5 (15.0)
Mean age at diagnosis (SD)	44.1 (13.2)
Low C3 or C4 (%)	31/59 (53)
Skin biopsy done (%)	89/99 (90)
Skin biopsy with vasculitis (%)	77/89 (87)
Met CHCC criteria for HUVS (%)	23/31 (74)

Symptoms/Manifestations	N (%)
Hive-like rash	102/102 (100)
Severe joint pain or swelling	77/93 (83)
Severe muscle pain	73/94 (78)
Nerve damage	56/77 (73)
Abdominal pain	49/79 (62)
Fever	32/78 (41)
Inflammation in one or both eyes	32/87 (37)
Lung involvement	28/84 (33)
COPD or Asthma	37/91 (41)
Weight loss	19/92 (21)
Kidney involvement	17/88 (19)
CNS involvement	6/80 (8)
Pericarditis	5/74 (7)
Thrombosis	5/87 (6)
Any systemic manifestation	85/102 (83)
Positive ANA	33/64 (52)

Means of Diagnosis	N (%)
Biopsy results	84 (82)
Based on symptoms	58 (57)
Laboratory testing of the blood	45 (44)
Radiographic testing	7 (7)
Other	4 (4)
Unsure	2 (2)

Medication Use (Ever)	N (%)
Glucocorticoids	89 (87)
Dapsone	27 (26)
Hydroxychloroquine	26 (25)
Colchicine	23 (23)
Methotrexate	19 (19)
Antihistamines	18 (18)
Azathioprine	16 (16)
Omalizumab	8 (8)
Aspirin	8 (8)
Mycophenolate mofetil	7 (7)
Cyclosporine	5 (5)
Rituximab	4 (4)
TNF inhibitors	3 (3)
Plasmapheresis	1 (1)

Physician Managing Vasculitis (Top 4)	n (%)
Rheumatologist	52 (51)
Primary Care Physician	35 (34)
Dermatologist	26 (25)
Allergist/Immunologist	16 (16)

DISCUSSION

- The diagnosis of urticarial vasculitis is clinicopathological, based on presence of urticaria and histologic confirmation
- All patients in this cohort reported urticarial lesions, the majority of which were confirmed by biopsy
- All clinical manifestations reported are consistent with physician-reported cohorts

CONCLUSIONS

Internet-based cohorts incorporating patient-reported data are a means for future conduct of clinical trials in extremely rare diseases, such as urticarial

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