damage indices at presentation. One rare but life-threatening complication of AAV is diffuse alveolar hemorrhage (DAH).

Objectives: To study the clinical features, course and response to therapy in Hispanic American patients with AAV and DAH seen at a large public teaching hospital that serves medically underserved and low income patients with chronic poor health maintenance.

Methods: Among 60 Hispanic American patients with AAV, we retrospectively studied 13 patients with DAH from 2003-2017. Clinical features, laboratory results, diagnostic modalities and treatments given were studied.

Results: A total of 13 patients were identified; 6 men and 7 women, mean age of 51 ± 11 years. Anti-PR3 was found in 5 patients, anti-MPO in 8. Nine patients were diagnosed with microscopic polyangiitis and 4 with granulomatous polyangiitis based on ACR criteria or Chapel Hill Consensus Conference definitions. DAH was a presenting manifestation in 12 patients. Gross hemoptysis with hypoxia, drop in hematocrit and chest X-ray infiltrates were seen in all patients diagnosed with DAH. Bronchoscopy with BAL washings confirming diagnosis was done in 10 patients. All patients had multi-organ involvement including nephritis ($n=8$), peripheral neuropathy ($n=1$), palpable purpura ($n=2$), upper respiratory tract involvement ($n=4$) and eye involvement ($n=2$). Serum C3 was normal in 12 patients. Renal biopsy in 9 patients showed pauci-immune crescentic glomerulonephritis. Mean BVAS at time of DAH was 21.2 ± 5.1 .

All patients underwent induction-remission therapy with pulse methylprednisolone followed by tapering oral prednisone. Induction therapy included IV cyclophosphamide ($n=6$), rituximab ($n=6$) and IV cyclophosphamide/rituximab combination therapy ($n=1$). Nine patients underwent plasma exchange and 5 received IV gamma globulin. Eleven patients required mechanical ventilation; 6 patients needed hemodialysis, 2 temporarily.

All 13 patients responded to induction therapy; mean post-induction therapy BVAS was 9.3 ± 3.5 . Pulmonary fibrosis after DAH was noted in 2 patients. No early deaths (<6 months) were observed, however 1 patient died within 3 years due to infection. Mean follow-up period was 69 ± 54 months. Maintenance therapy used included prednisone ($n=13$), mycophenolate mofetil ($n=2$), azathioprine ($n=6$) and IV rituximab ($n=8$).

Five patients had clinical relapse, 3 with recurrent DAH. Two patients had minor relapse, defined by their treating physician as requiring increased prednisone dose or worsening proteinuria. Mean BVAS at relapse was 15.2 ± 5.5 .

Conclusion: This first in-depth study of DAH in Hispanic American AAV patients shows that in this socioeconomically disadvantaged patient population, the response to induction therapy is not significantly different from other ethnic groups reported in the literature. DAH remains a severe clinical manifestation of AAV and should be recognized without delay. If treated promptly following EULAR/ERA-EDTA recommendations, good short term response is to be expected.

REFERENCES

- [1] Yates M, Watts RA, Bajema IM, et al. EULAR/ERA-EDTA recommendations for the management of ANCA-associated vasculitis. *Ann Rheum Dis* 2016;75:1583-1594
- [2] Jennette JC, Falk RJ, Bacon PA, et al. 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. *Arthritis Rheum* 2013;65:1-11

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.444

AB0617

CUTANEOUS POLYARTERITIS NODOSA TREATMENT: A RETROSPECTIVE CASE SERIES

Mohan Stewart^{1,2}, Ada Lo³, Kamran Shojania^{2,4}, Sheila Au⁵, Jan Dutz⁵, Michael Seidman⁶, Jon Chan⁴. ¹University of British Columbia, Internal Medicine, Vancouver, Canada; ²Arthritis Research Canada, Richmond, Canada; ³University of British Columbia, Vancouver, Canada; ⁴University of British Columbia, Rheumatology, Vancouver, Canada; ⁵University of British Columbia, Dermatology, Vancouver, Canada; ⁶University of British Columbia, Pathology, Vancouver, Canada

Background: Cutaneous polyarteritis nodosa (CPAN) is a small-to-medium vessel vasculitis limited to the skin. As it is a rare condition, there is little data available to guide treatment. Previous articles have suggested that colchicine and dapsone be used in mild disease and glucocorticoids in acute flares, with DMARDs such as methotrexate or azathioprine being reserved for moderate to severe disease, or steroid-refractory disease.

Objectives: Our objective was to review therapeutic regimens used to treat CPAN and to evaluate patient response.

Methods: A retrospective chart review including records from January 2003 to June 2017 was performed to identify patients with both histologic and clinical features of CPAN. We extracted information regarding patient age, clinical presentation, biopsy results, and response to treatment.

Results: Thirteen patients were identified who had clinical features of CPAN that were supported by histologic findings on biopsy. The majority of patients were female (92.3%). The average age at diagnosis was 44.9 years and mean duration of follow up was 53 months. All patients had involvement of the lower limbs with the most common cutaneous manifestation being subcutaneous nodules (92.3%), followed by papules (46.2%) and livedo reticularis (30.8%). Eight out of thirteen patients (61.5%) received steroids and three did not experience clinical benefit. Another patient did not respond to prednisone and hydroxychloroquine, but had significant improvement of her symptoms on prednisone and methotrexate. Seven out of nine patients (77.8%) who were treated with methotrexate had a favourable response. Three of those patients responded to methotrexate after failing other therapies such as dapsone, azathioprine, and prednisone.

Conclusion: In this case series, we found that most patients had a favorable response to methotrexate and thus the role of methotrexate in treating CPAN merits further investigation. As CPAN often has a chronic relapsing and remitting course, initiating methotrexate at the same time as steroid therapy may lower overall lifetime exposure to steroids.

REFERENCES

- [1] Daoud MS, Hutton KP, Gibson LE. Cutaneous periarteritis nodosa: a clinicopathological study of 79 cases. *Br J Dermatol*. 1997 May;136(5):706-13.
- [2] Ross K, Contreras J, Aung-Din D, Lien M. Asymptomatic cutaneous polyarteritis nodosa: treatment options and therapeutic guidelines. *Cutis*. 2017 Aug;100(2):125-8.
- [3] Maillard H, Szczesniak S, Martin L, Garot D, Machet MC, Machet L, et al. [Cutaneous periarteritis nodosa: diagnostic and therapeutic aspects of 9 cases]. *Ann Dermatol Venerol*. 1999 Feb;126(2):125-9.
- [4] Khoo BP, Ng SK. Cutaneous polyarteritis nodosa: a case report and literature review. *Ann Acad Med Singapore*. 1998 Nov;27(6):868-72.
- [5] Scharf NEC, Alaoui S, Vignon-Pennamen MD, Cordoliani F, Fermand JP, Morel P, et al. Successful treatment in two cases of steroid-dependent cutaneous polyarteritis nodosa with low-dose methotrexate. *Dermatology*. 2001;203(4):336-8.

Disclosure of Interests: Mohan Stewart: None declared, Ada Lo: None declared, Kamran Shojania: None declared, Sheila Au: None declared, Jan Dutz: None declared, Michael Seidman: None declared, Jon Chan Grant/research support from: Janssen, UCB, Novartis, Pfizer, Celgene, Consultant for: Amgen, Celgene, Eli Lilly, Janssen, Amgen, Abbvie, Novartis, Pfizer, UCB, Sandoz, Merck,

DOI: 10.1136/annrheumdis-2019-eular.6353

AB0618

A SNAPSHOT OF DISEASE ACTIVITY IN PATIENTS WITH BEHCET'S SYNDROME: PREDICTIVE FACTORS AND RELATIONSHIP WITH QUALITY OF LIFE

Rosaria Talarico, Alice Parma, Eralda Zera, Emanuele Calabresi, Chiara Tani, Chiara Baldini, Marta Mosca. *Rheumatology Unit, Clinical and Experimental Medicine, Pisa, Italy*

Background: Behçet's Syndrome (BS) is globally characterized by a variable spectrum of disease profile: while prevalent muco-cutaneous lesions and arthritis represent the only clinical features in patients with a benign disease subset, there are other patients who develop potentially sight or life-threatening manifestations, due to ocular, neurological or major vascular involvement. Beside the organ involvement, demographic factors could considerably influence the long-term and short-term outcomes of BD

Objectives: The primary aim of the study was to evaluate disease activity in a cohort of BS patients consecutively followed in a BS clinic of a tertiary centre; the secondary aims were to identify potential predictive factors of disease activity and to compare disease activity with quality of life.

Methods: One-hundred and thirty patients (71 males and 59 females; mean age 42 ± 8 years, mean disease duration 11 ± 4) with a diagnosis of BS according to the ISG criteria were studied. Disease activity has been evaluated by BDCAF and quality of life by the Short Form (SF) 36. Patients were also categorized in major or minor involvement of BS according or not to the presence of ocular, neurological and vascular involvement in the course of disease Predictors of long-term outcome were identified by univariate analysis using the log-rank test and by multivariate analysis using Cox proportional hazards regression models.