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AB0589 ANCA ASSOCIATED VASCULITIS: OUR EXPERIENCE FROM A TERTIARY CARE CENTER OVER 10 YEARS

Anjani Gummadi¹, Ankita Singh¹, Sandesh Guleria², Pandiarajan Vignesh², Anju Gupta¹, Deepti Suri², Ankur Jindal³, Biman Saikia⁴, Amit Rawat³, Surjit Singh². ¹Post Graduate Institute of Medical Education and Research, Chandigarh, Pediatrics, Advanced Pediatric centre, Chandigarh, India; ²Post Graduate Institute of Medical Education and Research, Chandigarh, Pediatrics, Advanced Pediatric centre, Chandigarh, India; ³Post Graduate Institute of Medical Education and Research, Chandigarh, Chandigarh, India; ⁴Post Graduate Institute of Medical Education and Research, Chandigarh, Department of Immunopathology, Chandigarh, India

Background: The anti-neutrophil cytoplasmic antibody (ANCA)- associated vasculitides (AAV) are a group of disorders characterized by necrotizing inflammation of the small to medium vessels in association with autoantibodies. Childhood ANCA vasculitides are rare but can cause organ or even life-threatening systemic vasculitis. Children most frequently present with rapidly evolving, severe disease.

Objectives: To describe the clinical spectrum of ANCA associated vasculitis, the treatment given and follow up

Methods: A single-centre retrospective analysis of ANCA associated vasculitis over a period of 10 years from 2008 to 2018

Results: Six children (2 boys; 4 girls) were diagnosed to have AAV during this period. Median age at diagnosis was 11.25 years (range 8-18 years). Median delay in diagnosis was 1.5 months (range 1-8 months). Presenting clinical features included lung disease- 4 children; arthritis- 3 (knee, right ankle-left 3rd MCP, left elbow-knee) hemoptysis- 2 children; ear discharge- 1; redness of eye-1; oliguria, gross hematuria-2; anasarca-2; 2 children had concomitant neurological symptoms (left foot drop, quadripareisis with seizures). All children had fever at presentation. Laboratory investigations showed elevated erythrocyte sedimentation rate(ESR) and C-reactive protein(CRP). ANCA testing was positive in 6 children (3 c-ANCA, +PR3, 3 p-ANCA, +MPO). Deranged renal function tests, proteinuria, microscopic hematuria) were seen in 4 out of 6 children. Renal biopsy showed pauci immune glomerulonephritis in 4 cases who presented with nephritis. All patients were treated with 5 pulses of intravenous methylprednisolone(30 mg/kg/day) with tapering doses of oral steroids. Intravenous cyclophosphamide pulses was given in 5 children while 1 received additional plasmapheresis (PLEX)- 8 cycles. Maintenance therapy included azathioprine(given in 5 children), low-dose prednisolone. Thrombocytopenia was noted in 1 child while on azathioprine; this was subsequently substituted with mycophenolate mofetil. Two children have been lost to follow-up. Median duration of follow-up is 35.5 months (range 8-124 months). All 4 children have attained remission with good compliance and there are no relapses.

Conclusion: While an evidence base treatment of AAV in children is limited, our data suggests that prolonged remission can be achieved with aggressive immunosuppressive therapy, however prognosis of childhood AAV remains guarded.

Table: Characteristics of children enrolled in study

Patient/ features	Age (years)/ year at diagnosis	Lower airway (lung)	Upper airway	Renal	CNS	Arthritis	ANCA	Therapy	Follow up (months)	Mortality
Patient 1	8/M (2007)	+	-	++	-	-	P ANCA	Steroids	Lost to follow-up	-
Patient 2	11.5/M (2008)	+	-	+	-	-	C ANCA	Steroids, cyclophosphamide, azathioprine, MMF	124	-
Patient 3	9/F (2013)	+	-	++	Seizures + quadripareisis	-	P ANCA	Steroids, cyclophosphamide, PLEX	Lost to follow-up	-
Patient 4	11/F (2015)	-	+	++	-	+	C ANCA	Steroids, cyclophosphamide, azathioprine	46	-
Patient 5	12/F (2016)	+	-	+	-	Left foot drop	P ANCA	Steroids, cyclophosphamide, azathioprine	25	-
Patient 6	18/F (2018)	+	-	++	-	+	C ANCA	Steroids, cyclophosphamide, azathioprine	8	-

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AB0590 A SINGLE CENTER RETROSPECTIVE ANALYSIS OF EFFICACY AND SAFETY BETWEEN LOW-DOSE VERSUS HIGH-DOSE RITUXIMAB AS REMISSION INDUCTION THERAPY IN JAPANESE PATIENTS WITH ANCA-ASSOCIATED VASCULITIS

Kei Hirose, Manami Hirata, Yuriko Yamamura, Akiko Ueno, Masahiro Yamamura. Okayama Saiseikai General Hospital, Center for Rheumatology, Okayama city, Japan

Background: Administration of four once-weekly doses of 375 mg/m² rituximab (RTX) has been indicated for ANCA-associated vasculitis (AAV) as remission induction therapy. However, randomized controlled trial for Japanese AAV patients have never been conducted, although Japanese AAV patients are characterized by the predominance of elderly patients with microscopic polyangiitis (MPA).

Objectives: To compare the efficacy and safety between low-dose versus high-dose RTX therapy as remission induction therapy in Japanese patients with AAV.

Methods: A single center retrospective analysis of 27 consecutive AAV patients with RTX therapy was performed. Clinical and laboratory variables at diagnosis, rates of complete remission (CR), defined as Birmingham Vasculitis Activity Score (BVAS)=0 and prednisone <7.5 mg/day, adverse effects, and vasculitis relapses following RTX use.

Results: Twenty-five MPA patients and 2 GPA patients (14 males and 13 females) were included in the present study. Twenty-six patients were positive for MPO-ANCA. Their median age was 77 years (range: 40-85 years). Treatments were determined according to the discretion of the attending physician. As remission induction therapy, 18 patients were treated with once or twice (1/2) RTX infusions (375 mg/m²), while 9 patients with 3 or 4 times (3/4) RTX infusions. At 6 months, 55.6% of the 1/2 infusion group (10/18) and 44.4% of the 3/4 infusion group (4/9) reached CR. At 6 months, mean PSL levels were 7.8 mg/day in the 1/2 infusion group and 6.6 mg/day in the 3/4 infusion group. At 18 months, 88.9% of the 1/2 infusion group (16/18) and 77.8% of the 3/4 infusion group (7/9) were survived. 0% of the 1/2 infusion group (0/18) and 33.3% of the 3/4 infusion group (3/9) were relapsed. Severe adverse effects occurred in 38.9% of the 1/2 infusion group (7/18) and in 22.2% of the 3/4 infusion group (2/9).

Conclusion: Our retrospective analysis indicated that cumulative CR rates and PSL tapering did not significantly differ between low-dose versus high-dose RTX as remission induction therapy in Japanese AAV patients, mostly elderly MPA patients, although there was no significant difference in severe adverse effects such as opportunistic infections between them.

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AB0591 POSITIVE PREDICTIVE VALUE OF THE GIANT CELL ARTERITIS DIAGNOSIS IN THE DANISH NATIONAL PATIENT REGISTRY: A VALIDATION STUDY

Peter Hjort¹, Philip Therkildsen¹, Berit Dalsgaard Nielsen¹, Ib Hansen¹, Annette de Thurah², Ellen-Margrethe Hauge¹. ¹Aarhus University Hospital, Rheumatology, Aarhus, Denmark; ²Aarhus University Hospital, Aarhus, Denmark

Background: Giant cell arteritis (GCA) is the most frequent systemic vasculitis[1]. The diagnosis is clinical and based on symptoms, histopathology, biochemistry and imaging. In Denmark, diagnostic codes for all in- and out-patient hospital diagnoses are registered in the Danish National Patient Registry (DNPR) [2]. Since GCA can be difficult to diagnose and treatment is initiated on suspicion, we hypothesized that the overall positive predictive value (PPV) of the GCA diagnosis code in the DNPR is low. High data quality is important for future epidemiological research in GCA.

Objectives: To establish PPV of the diagnostic code of GCA in the DNPR. Furthermore, to identify characteristics associated with a high PPV of the diagnostic code.

Methods: 293 patients aged ≥50 years with a first-time register-based GCA diagnosis were included from the DNPR in the period January 2012-January 2018. Patients were sampled based on the ICD-10 codes (M31.5 and M31.6) from two regional hospitals and one university hospital in the Central Region of Denmark. As gold standard we used the medical records (including biochemistry, histopathology and imaging results) and categorized each patient as true GCA or non-GCA. Based on the data from the prescription database, patients were divided into four categories depending on the number of prednisolone prescriptions they received. Two independent investigators (PH and PT) reviewed the medical records. In case of disagreement the final diagnosis was reached by consensus or by expert opinion (ITH). To test how the PPV varied,

sub-analyses were performed for number of prescriptions, specific symptoms, temporal artery biopsies (TAB), number of visits and number of prescriptions.

Results: We confirmed 183 of 293 diagnoses resulting in a PPV of 62.8% (95% CI: 57.1-68.2). In patients having at least three admissions, 95 of 110 diagnoses were confirmed (PPV: 86.4%; 95% CI: 78.5-91.7). We confirmed 82 of 95 patients with at least three visits and a TAB. This resulted in a PPV of 86.3% (95% CI: 77.7-91.9). The highest PPV was observed in patients with at least 3 visits and ≥ 3 prescriptions of prednisolone. In this sub-analysis we confirmed 88 of 95 diagnoses resulting in a PPV of 92.6% (95% CI: 85.2-96.5).

Conclusion: This is the first study to validate the diagnostic code of GCA in the DNPR. The overall PPV of a first-time diagnosis of GCA in the DNPR is low. The probability of identifying true cases of GCA increases substantially when diagnostic codes are combined with 3 visits and ≥ 3 prescriptions of prednisolone.

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AB0592 SYSTEMIC

¹Peking University Third Hospital, Department of Rheumatology and Immunology, Beijing, China, ²School of Basic Medical Sciences, Peking University, Department of Pathology, Beijing, China

Background: Systemic vasculitis involving the breast is a rare clinical condition and may mimic breast cancer or mastitis clinically or radiographically.

Objectives: In this article, we reported a case of polyarteritis nodosa (PAN) with breast involvement and made a summary on the published cases of systemic vasculitis affecting breasts in order to better understand this disorder.

Methods: We reported a case of PAN affecting breast and conducted a retrospective review of literatures reporting patients with the diagnosis of systemic vasculitis involving breasts by using MEDLINE, EMBASE, Web of Science, the Cochrane Library, and Scopus database in any language up to June 1st, 2018.

Results: A case of a 27-year-old female patient presented with a painful mass of the right breast was diagnosed as PAN by the biopsy. After six months of treatment of full dose prednisone and methotrexate, her condition kept stable and inflammatory markers remained normal. A total of 67 cases were included, with granulomatosis with polyangiitis (GPA), giant cell arteritis (GCA) and PAN as the main types. The typical manifestation was mass (79.2%, 53/67) in the breast. All the diagnoses were made according to the pathology of the breast biopsy. Glucocorticoid and immunosuppressant were the main therapies. 74.6% (50/67) patients achieved remission during follow-up.

Conclusion: In total, we reviewed the clinical features and outcome of 67 cases of the systemic vasculitis involving the breast including the case we reported. Definite diagnosis depends on tissue biopsy, which is important, as the prognosis of diseases is upon to the types of vasculitis.

Figure 1

Figure 2

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AB0593 INTERSTITIAL LUNG DISEASE ASSOCIATED WITH ANCA POSITIVITY: A RETROSPECTIVE ANALYSIS

Sabrina Juman¹, Alexander Oldroyd², Nazia Chaudhuri³, Sahena Haque⁴. ¹The University of Manchester, Faculty of Biology, Medicine and Health, MANCHESTER, United Kingdom; ²University Of Manchester, Centre for Musculoskeletal Research, Manchester, United Kingdom; ³Manchester University NHS FT, Respiratory Medicine, Wythenshawe, United Kingdom; ⁴Manchester University NHS FT, Rheumatology, Wythenshawe, United Kingdom

Background: The association between interstitial lung disease (ILD), anti-neutrophil cytoplasmic autoantibodies (ANCA) and vasculitis has been described in case reports and small series. Treatment approaches have been diverse and include the use of various immunosuppressive agents. To date, little is known about the clinical phenotype and outcomes of ANCA associated ILD.

Objectives: The objectives of this study were to ascertain the prevalence of ANCA in ILD patients at a tertiary respiratory centre; describe the clinical phenotype; describe lung function outcome and examine the progression of computed tomography (CT) patterns in patients with ANCA-associated ILD.

Methods: This retrospective observational study was conducted at a tertiary ILD centre. Patients with ILD and coexistent ANCA were identified from review of a clinical database. All patients had a multidisciplinary team diagnosis of ILD confirmed on CT scan and had no other underlying cause of ILD identified. Data regarding clinical characteristics, CT scan pattern of ILD, pulmonary function tests (PFTs) and treatment were collected retrospectively.

Results: Sixty-nine patients that fulfilled the criteria of coexistent ILD and ANCA were identified. Thirty-one patients were cANCA positive and 38 patients pANCA positive with a median age of 67 years (interquartile range 15 years). Sixteen patients were positive for MPO-ANCA and eight for PR3-ANCA. Only 26% of the cANCA and 24% of the pANCA population had coexistent ANCA-associated vasculitis diagnosed by a MDT clinic comprising of rheumatology, respiratory and renal physicians.

Figures 1 and 2 show the treatment distribution in c and pANCA populations respectively. Approximately 23% of the entire study population did not require treatment. Of the patients that required treatment, steroids were used in 81% of cANCA and 61% of pANCA patients. The most commonly used disease modifying anti-rheumatic drug was mycophenolate mofetil followed by azathioprine and/or methotrexate. Only a small proportion of patients required cyclophosphamide or rituximab.

At 24-months follow-up, 68% were alive, 22% deceased and data unavailable for 10% of the cANCA group compared to 61% alive, 2% end-of-life, 16% deceased and 21% unavailable data in the pANCA group. Diffusing capacity of the lungs for carbon monoxide (DLCO) declined in both p and cANCA groups over the first 12 months before improving. This improvement in PFTs is likely due to treatment escalation over time. Overall the pANCA group showed a mixture of progressive fibrotic and inflammatory CT features while the cANCA and MPO-ANCA groups showed predominantly progressive fibrotic CT findings.

Kruskal-Wallis test was used to determine whether there was an association between the PFTs of the p and cANCA population at three to six months, 12 months and 24 months. This test was then repeated for comparison of the MPO-ANCA and PR3-ANCA study population. There was no significant difference in lung function over time between the pANCA and cANCA group or MPO-ANCA and PR3-ANCA group.