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AB0538

PREGNANCY OUTCOMES IN PATIENTS WITH TAKAYASU'S ARTERITIS: CASE SERIES

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Background: Takayasu's arteritis (TA) is most prevalent in women of childbearing age. Although its activity and risk of relapse are low during pregnancy, up to 40% of patients may have unfavorable obstetric outcomes and therefore it is important to know their clinical behavior.

Objectives: To describe the clinical features and obstetric outcomes of pregnant women with TA treated in a tertiary center.

Methods: Retrospective evaluation of medical records of 6 pregnancies in 6 women with TAs treated in a tertiary center in Medellín, Colombia between 2011-2018.

Results: Six women who were 17.5 (RI 9.25) years old at diagnosis and 24 (RI 8.25) years old at delivery, their disease duration were 5.5 (RI 10.5) years. Three patients had extensive aortic involvement classified as Numano type V, two as type IIB and one as type I. At delivery, three patients were active and required immunosuppressants, five had high blood pressure, one developed preeclampsia in the second trimester, one had severe mitral and tricuspid insufficiency with decreased ejection fraction of the left ventricle; two had aneurysms (left subclavian artery and ascending aorta). There were two fetal deaths, one due to intrauterine growth restriction and placental insufficiency and another of unknown etiology; both patients with disease activity, extensive aortic condition and arterial hypertension; no pregnancy resulted in abortion or preterm birth. Five deliveries were by caesarean section by maternal indication; there was no aortic dissection, aneurysmal rupture or cerebral hemorrhage (table).

Table. Patient's characteristics

Patient	Age at diag-	Age at delivery	Clinical features	Hata-Numano classification	Maternal outcome	Fetal outcome
1	6	17	HT, absence of left brachial and radial pulses, L carotid and subclavian murmurs, LVEF 47%	V	HT, C-section	Term delivery, SGA
2	22	26	HT, absence of L brachial and radial pulses, L carotid and subclavian murmurs, dyspnea, angina	V (plus pulmonary)	HT, vaginal birth	Placental insufficiency, IUGR, fetal death
3	14	38	HT, absence of L brachial and radial pulses, L carotid and subclavian murmurs, intermittent claudication, dyspnea, aortic regurgitation	IIB	HT, C-section	Term delivery, SGA
4	18	24	HT, malaise, absence of L brachial and radial pulses, intermittent claudication, arthralgia	IIB	Preeclampsia, C-section	Term delivery, SGA
5	17	22	Fever, malaise, intermittent claudication, arthralgia	I	C-section	Term delivery, SGA
6	21	24	HT, absence of bilateral femoral, popliteal, tibial and pedal pulses, headache, dizziness, amaurosis	V	HT, C-section	Fetal death

HT: hypertension; L: left; LVEF: left ventricular ejection fraction; C-section: cesarean section; SGA: small for gestational age; IUGR: intrauterine growth restriction

Conclusion: Pregnant women with active disease and extensive aortic condition presented unfavorable obstetric results, suggesting that an inadequate control of vasculitis may lead to greater maternal-fetal complications.

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AB0539

URINARY INFLAMMATORY CELL ANALYSIS REFLECTS THE RENAL HISTOPATHOLOGY IN ANTI-NEUTROPHIL CYTOPLASMIC ANTIBODY-ASSOCIATED VASCULITIS

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Background: The anti-neutrophil cytoplasmic autoantibody (ANCA)-associated vasculitides (AAVs) include microscopic polyangiitis (MPA), granulomatosis with polyangiitis (GPA), and eosinophilic granulomatosis with polyangiitis (EGPA). These small-vessel vasculitides are characterized by necrotizing inflammation of the vessel wall, particularly affecting small arteries, arterioles, and capillaries in systemic organs, and the kidney is one of the most frequently involved organs. Although kidney biopsy is necessary for deciding the therapeutic protocol, it is invasive and is sometimes hard to perform biopsy because of patient's severe general condition. We have already reported that T cells and macrophages appear in the urine of patients with glomerulonephritis, accompanied by active cellular infiltration such as cellular crescent formation and diffuse interstitial cell infiltration, but not in the urine of patients with glomerulonephritis without the active inflammatory lesions.

Objectives: In this study, we examined the utility of urinary inflammatory cell analysis for accessing kidney histopathological findings in AAVs.

Methods: This was a cross-sectional, retrospective chart study. Thirty-six AAV patients who had been referred to Niigata University Hospital between 2002 and 2018, and performed percutaneous kidney biopsy and urinary inflammatory cell analysis, were participated in this study. Thirty-two patients had MPA, and 4 had GPA. The kidney biopsy findings were classified into Berden's classification (a method to categorize glomerular lesions into four classes) and Neumann's classification (a method to evaluate glomerular, tubulo-interstitial, and vascular lesions by using activity indexes and chronicity indexes). Flow-cytometric analysis of urinary inflammatory cells was performed for each subject. Numbers of urinary T cells or macrophages were determined by multiplying the number of viable cells in the gated mononuclear cell region in each sample by the percentage of urinary CD3-positive or CD14-positive cells in the population, respectively. The correlations between the results of both methods and the numbers of urinary inflammatory cells were examined using Kruskal-Wallis test and Spearman's rank correlation coefficient. A p-value of <0.05 was taken to denote statistical significance.

Results: The numbers of urinary inflammatory cells showed a trend of increase in crescentic category without statistical significance in Berden's classification. Meanwhile, activity indexes had significant positive correlations with the number of urinary CD3-positive cells ($r = 0.541$, $p = <0.001$), CD14-positive cells ($r = 0.354$, $p = 0.034$), and total inflammatory cells ($r = 0.449$, $p = 0.006$) in Neumann's classification.

Conclusion: The numbers of urinary inflammatory cells reflect the active lesions of kidney histopathological findings, and these results indicate the usefulness of urinary inflammatory cell analysis for assessment of kidney biopsy findings in patients with AAVs.

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AB0540

CLINICAL ANALYSIS OF AORTA INVOLVEMENT IN PATIENTS OF ANCA-ASSOCIATED VASCULITIS

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Background: ANCA-associated vasculitis (AAV) is an autoimmune disease that involves abnormal death of neutrophils and leads to necrotic inflammatory reactions in blood vessels, including microscopic polyangiitis (MPA), Granulomatous polyangiitis (GPA) and Eosinophilic granulomatous polyangiitis (EGPA). AAV is mainly involved in small blood vessels, and intermediate arterial lesions can also occur, but the large arteries and their primary branches are rarely involved.

Objectives: To summarize the clinical characteristics of aortic involvement in patients with ANCA-associated vasculitis (AAV).

Methods: The clinical manifestations, systemic involvement, laboratory examination, imaging characteristics and treatment of aortic involvement in AAV patients admitted to Peking Union Medical College Hospital from January 2013 to December 2018 were retrospectively analyzed.

Results: Nine patients were enrolled in our study. The ratio of male to female was 2:1 and the median age was 47 years old. Of the 9 patients, 4 were GPA (44%), 4 were MPA (44%) and 1 was EGPA (11%). The aorta is involved in an average of 3 locations per case, mainly in 7 locations: 3 ascending aorta and aortic arch, 4 in the head and arm trunk (including carotid and subclavian artery), 2 in the abdominal aorta, and 1 in the abdominal cavity. There were 2 cases of renal artery, 1 case involving brachial radial artery, 2 cases of iliac artery and lower limb artery, and 1 case involving left main coronary artery, anterior descending branch, circumflex branch, and right coronary artery. Aortic lesions: 3 cases had aneurysms, arterial dilatation and / or dissection, 6 cases had arterial stenosis or occlusion and 3 cases had periarteritis. When major arterial involvement was found, the AAV of the patients were mostly active, with an average of 19 points for BVAS vasculitis activity and 1 for FFS score. 6 cases had lung involvement (67%), 6 cases had kidney involvement (67%), 4 cases had ENT involvement (44%), 3 cases had nervous system involvement (33%), and 1 case had gastrointestinal involvement (11%). All patients were treated by steroid and immunosuppressant, while 1 case received the operation of ascending aorta and aortic arch replacement.

Conclusion: Mainly involved in small blood vessel inflammation, AAV may also have aorta involvement, which was more common in patients who had active disease and need more positive treatment. The affected aorta areas of these patients were mainly ascending aorta, aortic arch, and head and arm trunk, which can be manifested as aneurysms, dissections, and arterial stenosis Periarthritis, etc. If necessary, surgically treatment of the affected aorta could be considered when the situation of AAV was stable enough.

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AB0541

IMPROVED SURVIVAL IN PATIENTS WITH GIANT CELL ARTERITIS: A POPULATION-BASED COHORT STUDY

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Background: In previous studies patients with giant cell arteritis (GCA) have had survival rates that are similar to the general population.

Objectives: To investigate survival trends and cause-specific mortality in patients diagnosed with GCA over a 60-year period.

Methods: We assembled a population-based incidence cohort of patients with GCA diagnosed between 1950 and 2009. All patients were included if they met the American College of Rheumatology (ACR) 1990 Criteria for the

Classification of GCA. Patients diagnosed between 2000 and 2009 could also be included if they met the following criteria: age greater than or equal to 50 years, elevated inflammatory markers, and radiographic evidence of large-vessel vasculitis attributed to GCA. A non-GCA comparison cohort was assembled from the same underlying population for each patient with GCA. Patients were followed until death, last contact, or December 31st, 2018. Survival trends were analyzed by grouping patients into the following categories according to year of GCA diagnosis: Group A 1950-1979; Group B 1980-1989; Group C 1990-1999; and Group D 2000-2009. Mortality rates were estimated using the Kaplan-Meier method and were compared with expected mortality rates for persons of the same age, sex, and calendar year, as estimated by regional population life tables. Cause-specific mortality was obtained from death certificates for patients in both cohorts. The causes were grouped according to ICD-9 chapters and hazard ratios were estimated against the non-GCA comparators.

Results: The study population included 245 incident cases of GCA: 194 (79%) women and 51 (21%) men with mean age ($\pm$ SD) of 76.2 ($\pm$ 8.3) years and median follow-up of 10.6 years. There was no overall difference in survival between the GCA cohort and the general population. The 2-, 5-, and 10-year survival rates (95% CI) were 89% (86, 93), 76% (70, 81), and 56% (50, 63) respectively with a standardized mortality ratio of 0.99 (0.86, 1.14). The standardized mortality ratios for Groups A, B, C, and D were 0.83 (0.57, 1.17), 0.92 (0.63, 1.3), 1.21 (0.85, 1.69), 0.70 (0.50, 0.95), respectively. The overall all-cause mortality adjusted for age, sex, and calendar-year was similar between the GCA patients and their comparators with a hazard ratio of 1.03 (0.84, 1.24). Mortality due to neoplasms was significantly lower in the GCA cohort with a hazard ratio of 0.53 (0.3, 0.92). Other cause-specific mortalities were not significantly different between the groups.

Conclusion: In this population-based cohort of patients with GCA diagnosed over a 60-year period, the survival of patients diagnosed in recent years was significantly better than that of the general population. The explanation for this novel finding is unclear, but likely to be multifactorial. In this study the number of deaths due to neoplasm in the GCA group was significantly lower.

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AB0542

REACTIONS TO PNEUMOCOCCAL 13-VALENT VACCINE IN PATIENTS WITH BEHCET SYNDROME

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Background: The European League Against Rheumatism (EULAR) recommends pneumococcal 13-valent (PCV13) and 23-valent vaccines in patients with rheumatic diseases (1). Adverse reactions to 23-valent pneumococcal vaccine were previously reported in patients with Behçet Syndrome (BS) (2). These were proposed to be associated with the pathergy phenomenon which may be observed in patients with BS.