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Figure 1. Comparison between clinical domains in patients with and without ophthalmological involvement.

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AB0398

CASE SERIES OF PATIENTS WITH CHOLESTEROL CRYSTAL EMBOLISM SYNDROME THAT MIMICS SYSTEMIC VASCULITIS

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Background: Cholesterol crystal embolism (CCE) syndrome is a multisystemic disorder caused by small arteries cholesterol crystal emboli subsequent to small pieces of atheromatous plaques from the aorta or other major arteries break off. CCE is often overlooked because it mimics symptoms of systemic vasculitis due to its clinical characteristics such as ulceration and gangrene of toes, livedo reticularis, renal insufficiency. Acute inflammatory reactants such as ESR, CRP may elevate in CCE patients since the cholesterol crystals trigger a foreign-body inflammatory reaction around the arterioles.

Objectives: This study aimed to explore the clinical characteristics of CCE patients, to make rheumatologists learn more about this disease.

Methods: Peer-reviewed articles in the electronic databases Medline, PubMed, Science Citation Index, China Biomedical Literature Database (CBM), China Journal Full Text Database (CNKI), and WANFANG Data were searched using the terms "cholesterol crystal embolism syndrome", "cholesterol embolism", "atherosclerotic embolism", "atherosclerotic nephropathy", or "CCE". Only articles or case reports containing detailed medical records of CCE patients were included. We also included CCE patients in our department.

Results: A 66-year-old male CCE patient presented with multiple ulceration and gangrene of toes and heels (Figure 1), subacute renal insufficiency, and elevated CRP and ESR. This patient had been considered as "suspected systemic vasculitis" and was referred to our rheumatology department. Another 39 Chinese CCE patients from the above databases were qualified for analysis. Among these 40 patients, 87.5% (35/40) were male and the mean age was 68±6 years. The most common involved was kidney and 90% (36/40) of patients presenting with renal insufficiency including the progressive increase of serum creatinine, hematuria, proteinuria, or sudden (or sharp) aggravation of hypertension. Next common involved was skin that occurred in 87.5% (35/40) of patients, especially in the toes and heels. For skin manifestations, blue toe syndrome occurred in 82.5% (33/40) of patients, ulceration or gangrene in 25% (10/40), and livedo reticularis in 15% (6/40). Additionally, 12.5% (5/40) showed ocular involvement such as visual impairment and visual field defect. In 2 patients, embolized cholesterol crystal in retinal arteries that is called Hollenhorst plaques was detected by fundoscopy. There were 62.5% (25/40) of patients having elevated CRP or ESR. The positive rate for skin or subcutaneous biopsies was 58% (11/19) and for kidney biopsies was 100% (6/6). The precipitating factors preceding the occurrence of classical symptoms such as blue toe syndrome, livedo reticularis and/or subacute renal insufficiency is important for CCE diagnosis especially for patients who had contraindications or were intolerant to biopsy. The precipitating factors include endovascular intervention (80%), vascular surgery (5%), and anticoagulant or thrombolytic therapy (2.5%). Only 12.5% (5/40) of patients were spontaneous and didn't have any predisposing factors. General interventions of CCE included statins (82.5%), antiplatelets (32.5%), and dialysis (32.5%). Twelve patients (30%) received glucocorticoids and 75% (9/12) of them renal function improved and ulceration healed (Figure 1). Among 36 patients who presented with renal insufficiency, the renal function returned to normal after treatment in 2 patients (5.6%), but 27 patients (75%) still showed abnormal renal function even though somewhat improved, and 7 patients (19.4%) needed renal replacement therapy or dialysis for maintenance.

Conclusion: This study reported CCE patients had high prevalence of renal insufficiency, blue toe syndrome, and ulceration or gangrene of toes, as well as elevated CRP or ESR, thus rheumatologists should be alert to this disease as one of the differential diagnosis of systemic vasculitis, especially for elderly patients with evidence of atherosclerosis who undergo a recent cardiovascular procedure.

Figure 1: A 66-year-old male CCE patient presenting with multiple ulceration and gangrene of toes and heels (A). Skin gangrene with or without ulceration scab was seen in bilateral heels, the third right toe, and the first and fourth left toes. The fifth right toe showed blue color (named as blue toe syndrome). After 6-month glucocorticoids treatment, the skin ulceration completely healed (B).

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AB0399

EPIDEMIOLOGY OF BEHÇET DISEASE IN A NORTHERN SPANISH HEALTH REGION

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Background: Behçet disease (BD) is a systemic and potentially severe disease. Its incidence varies widely worldwide. In Spain, published BD's epidemiological studies are scarce.

TABLE 1.

	Diagnostic criteria and study period	n cases / population size	Mean age at onset and sex (%female)	Prevalence (over 100000) / incidence	Oral / genital ulcers (%)	Skin lesions/ pathology test (%)	Ocular involvement (%)	Joint involvement (%)	Neurobehcet/ Vascular/ Gastrointestinal involvement (%)
Sánchez, L. et al. Southern Europe (Cantabria, Spain)	Expert opinion, ISG, ICBD / 1980- 2020	120 (expert opinion) / 96 (ICBD) / 59 (ISG) / 581641	37.6 ±13.8/ 48.3	20.6 (expert opinion), 16.5 (ICBD), 10.1 (ISG) / 0.022 (expert opinion), 0.018 (ICBD), 0.011 (ISG)	94.2 / 59.2	63.3 / 25.2	41.6	65	10.8 / 11.6 / 6.6
Calamia, K. T. et al. North America (Minnesota, USA)	ISG / 1960-2005	13 / NR	31 / 30	5.2 / 0.38	100 / 62	85 / NR	62	46	23 / 23 / NR
Altenburg, A. et al. Northern Europe (Berlin, Germany)	ISG and ABD classification tree / 1961-2005	590 / 3391344	26 / 58	4.9 / 1 (estimated)	98.5 / 63.7	62.5 / 33.7	58.1	53	10.9 / 22.7 / 11.6
Mohammad, A. et al. Northern Europe (Skane County, Sweden)	ISG / 1997-2010	40 / 809317	30.5 / 33	4.9 / 0.2	100 / 80	88 / NR	53	40	0 / 20 / NR
Mahr, A. et al. Southern Europe (SeineSaint-Denis County, France)	ISG / 2003	79 / 1094412	27.6 / 43	7.1 / NR	100 / 80	90 / 20	51	59	10 / NR / 10
Salvarani, C. et al. Southern Europe (Reggio Emilia, Italy)	ISG, 1988-2005	18 / 486961	33 / 50	3.7 / 0.24	100 / 78	100 / NR	56	50	11 / 6 / NR
Azizlerli, G. et al. Middle East (Istanbul, Turkey)	ISG / prevalence study	101 / 23986	NR / 48.5	42 / NR	100 / 70.2	Not globally reported / 69.3	27.7	Not globally reported	NR / Not globally reported / NR
Davatchi, F. et al. Middle East (Iran nationwide)	Expert opinion / 1975-2018	7641 / NR	25.6 / 44.2	80 / NR	97.5 / 64.4	62.2 / 50.4	55.6	38.1	3.9 / 8.9 / 6.8
Krause, I. et al. Middle East (Galilee, Israel)	ISG / 15 years (not specific years have been reported)	112 / 737000	30.6 / 47	15.2 / NR	NR / 68	41 / 44.4	58	70	11.6 / Not globally reported / NR
Nishiyama, M. et al. Asia (Japan nationwide)	1987 JCBD / 1991	3316 / NR	35.7 / 50.6	NR / NR	98.2 / 73.2	87.1 / 43.8	69.1	56.9	11 / 8.9 / 15.5

JCBD: Japanese diagnostic Criteria of Behçet's Disease; n: number of cases; NR: Not Reported

Objectives: In a well-defined Northern Spanish population-based cohort, the aim of this study was **a)** to estimate epidemiological variations, **b)** clinical domains and **c)** to compare our results with other regions.

Methods: Study of unselected all consecutive patients diagnosed with definitive or possible BD by expert rheumatologists between 1980 and 2020 in our health region. Two classification criteria were applied: **a)** International Study Group (ISG) for BD [1], and **b)** International Criteria for BD (ICBD) [2]. In addition, a literature review of Medline publications was carried out.

Results: In this study, from a total of 120 patients diagnosed with BD by expert opinion (58 women/62 men), 96 met ICBD and 59 ISG criteria. Mean age of the cohort at diagnosis was 37.6±13.8 years similar to other countries, as well as sex distribution.

Prevalence was higher than in most European populations regardless the diagnostic criteria applied: expert opinion (20.6), ICBD (16.5) or ISG (10.1) (TABLE 1). Incidence was lower (expert opinion: 0.022, ICBD: 0.018, ISG: 0.011). Clinical domains' frequency was in line with other regions except vascular and gastrointestinal involvement, which were lower.

Conclusion: BD's prevalence in Northern Spain is higher than in most European populations. These differences likely reflect a combination of true geographic variation, methodological artifacts as well as the easy access to Public Health System and its efficiency. In contrast, clinical phenotypes are similar to other regions.

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AB0400

URINARY ALBUMIN-TO-CREATININE RATIO INDICATES NECROTIZING AND CRESCENTIC GLOMERULONEPHRITIS IN ANCA-ASSOCIATED VASCULITIS

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Background: Renal involvement is a common and severe complication of anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) as it can cause acute kidney injury (AKI), end-stage renal disease (ESRD) or death.

Objectives: We have previously reported that elevated urinary albumin-to-creatinine ratio (uACR) correlates with rapid deterioration of kidney function in ANCA GN. Therefore, we here aimed to describe the association between proteinuric findings and histopathological diagnosis of necrotizing and crescentic ANCA GN in 50 urinary samples at admission and corresponding renal biopsies of patients with AAV.

Methods: A total number of 50 urinary samples at admission and corresponding renal biopsies with confirmed renal involvement of AAV were retrospectively included between 2015 till 2020 in a single-center observational study.

Results: Renal involvement of AAV revealed variable proteinuria ranging from low-range to nephrotic syndromes, however most patients presented with subnephrotic proteinuria. Severe deterioration of kidney function requiring RRT within 30 days after admission was associated with elevated levels of nonselective proteinuria, mostly attributed to albuminuria (uACR). Because we have previously shown that histologically confirmed ANCA GN with glomerular crescents and necrosis is associated with AKI and requirement of RRT during short-term disease course and elevated uACR levels were equally associated with AKI and requirement of RRT during the short-term course after disease onset, we next analyzed the association between uACR measurements at admission and histopathological findings within renal biopsies performed thereafter. Severely increased uACR levels >300mg/g correlated with reduction of normal glomeruli, attributed to increased glomerular crescents and necrosis. By contrast, no such association was observed for global sclerotic glomeruli, revealing that uACR reflects crescentic ANCA GN rather than adaptive glomerular hyperfiltration in chronic sclerosing stage. Since uACR levels could reflect both, either a specific renal involvement with necrotizing and crescentic ANCA GN or severity of systemic AAV disease, we next correlated uACR levels assessed at admission with extrarenal disease manifestation. We observed no association between uACR levels and extrarenal manifestation of AAV disease including pulmonary hemorrhage, skin involvement and BVAS assessment, suggesting that uACR levels reflected specific renal involvement in AAV. These observations were further confirmed by survival analysis for cumulative incidence of RRT during the short-term course of disease.

Conclusion: Early identification of patients who mostly benefit from aggressive immunosuppressive therapy is of clinical importance. Our observation that uACR levels at disease onset predict necrotizing and crescentic ANCA GN requires further investigation for therapeutic decision especially in patients with severe deterioration of kidney function.

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