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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.