| Case | 1 | 2 | 3 | 4 | 5 | 6 | 7 | 8 |
|---|---|---|---|---|---|---|---|---|
| Sex | Female | Female | Female | Female | Female | Female | Female | Female |
| Age (years old) | 73 | 62 | 34 | 64 | 64 | 59 | 58 | 75 |
| Disease Duration (years) | 6 | 30 | 9 | 3 | 17 | 26 | 6 | 25 |
| Disease Type | Diffuse | Diffuse | Diffuse | Diffuse | Diffuse | Limited | Limited | Diffuse |
| Autoantibody | ARA | ACA | ATA | ATA | ATA | ATA | ND | ND |
| Digital Ulcer | - | + | - | - | - | - | - | + |
| Interstitial Lung Disease | + | + | + | + | + | + | - | + |
| CKD (eGFR<60 mL/min/ 1.73 m2) | + | - | - | - | - | - | - | - |
| MPO-ANCA titer (IU/mL) | 14.2 | 10.0 | 63.8 | 38.7 | 9.1 | 11.5 | 20.1 | 13.9 |
| PSL administration | + | + | - | + | + | + | + | - |
| PSL Dosage (mg/day) | 1 | 5 | | 10 | 10 | 2 | 6 | |
| Previous History of IVCY | + | - | - | + | + | - | + | - |
| Follow-up Period (months) | 39 | 39 | 5 | 39 | 38 | 38 | 39 | 23 |
| Newly Onset of AAV | - | - | + | - | - | - | - | - |
| Newly Onset of Acute Renal Dysfunction | + | - | + | - | - | - | - | - |

ARA: anti RNA polymerase 3 antibody, ATA: anti topoisomerase 1 antibody, ACA: anti centromere antibody, ND: not detected, CKD: chronic kidney disease, PSL: prednisolone, IVCY: intravenous cyclophosphamide pulse therapy, ACEi: angiotensin converting enzyme inhibitor, ARB: angiotensin receptor blocker

antibody (ANCA) positivity. On the other hand, it is also reported that 7%–13% of patients with SSc revealed myeloperoxidase-ANCA (MPO-ANCA) positivity without vasculitis manifestation in 1990s, but their clinical characteristics were unclear. It is also unknown whether ANCA positivity leads to AAV or not in patients with SSc. It is important for physicians to clarify the characteristics of SSc patients with ANCA positivity, and answer the question whether they will shift ANCA-associated vasculitis (AAV).

**Objectives:** To assess the prevalence of ANCA positive patients with SSc, and clarify the characteristics of these patients.

**Methods:** We enrolled the 333 consecutive patients with SSc who visited our clinic during October 2014 to September 2015, all of who were checked MPO-ANCA using fluorescent-enzyme immune-assay. Clinical manifestation and laboratorial data were obtained from medical chart. The data were assessed by chai-square analysis and Welch's t test.

**Results:** Two patients were diagnosed AAV before October 2014. Eight patients (2.4%) revealed MPO-ANCA positivity without vasculitis manifestation. All of MPO-ANCA positive patients were female, and mean age and disease duration were 61.1 years old and 17.2 years, respectively, and there's no statistically significant differences comparing MPO-ANCA negative patients. As a result of evaluating clinical manifestations, we found that patients with MPO-ANCA positivity more frequently had interstitial lung disease than patients without MPO-ANCA positivity (87.5% vs. 36.7%, p<0.01). The clinical characteristics of 8 patients were shown in table 1. Only one patient out of 8 patients with MPO-ANCA positivity newly diagnosed AAV during mean of 33 months follow-up period.

**Conclusions:** The prevalence of MPO-ANCA positivity in SSc patients were lower than previous reports. MPO-ANCA positivity may be related to interstitial lung disease in SSc. MPO-ANCA positive patient may occasionally reveal AAV in the future, and careful observation are needed.

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## Vasculitis

FRI0474 **CLASSIFICATION OF SKIN INVOLVEMENT IN LEVAMISOLE-ADULTERATED COCAINE INDUCED VASCULOPATHY**

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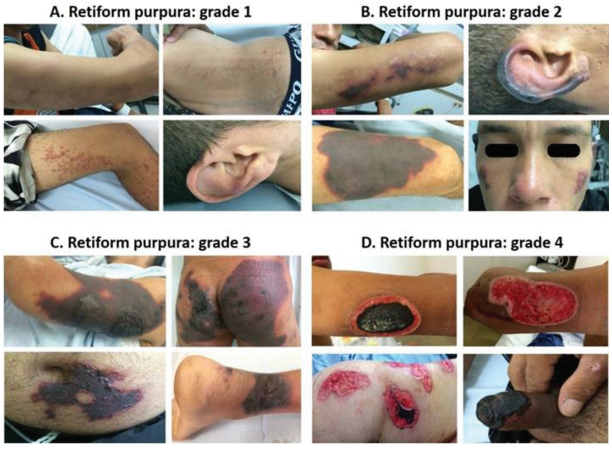
**Background:** Up to 88% of cocaine is tainted with levamisole, an anthelmintic withdrawn from the market due to toxicity. Since 2010 levamisole-adulterated cocaine induced vasculopathy (LACIV) patients, characterised by retiform purpura, ear necrosis, multisystemic compromise and positivity for multiple autoantibodies, have been reported. Knowing the pattern and the severity of skin involvement is essential in the approach of these patients.

**Objectives:** To describe the cutaneous manifestations of patients with LACIV and to propose a classification of skin involvement.

**Methods:** We describe the skin compromise of 30 patients with LACIV evaluated between December 2010 and May 2017. Based on this series and the review of the literature, we propose a classification according to the distribution and severity of the lesions.

**Results:** All patients were mestizo, median age of 31 (IQR 27–38), male:female ratio 5:1, time from symptoms to diagnosis 12 months (IQR 6–24). The most frequent clinical manifestations were skin lesions: ear necrosis (73%) and retiform purpura (83%) affecting the extensor part of the limbs, buttocks, face, and abdomen; sparing the scalp, palms and soles. Retiform purpura was classified in four grades according to distribution and severity (image). Skin biopsies revealed leukocytoclastic vasculitis (24%), pseudo-vasculitis (19%), thrombotic vasculopathy with leukocytoclastic vasculitis (19%), thrombotic vasculopathy with pseudo-vasculitis (19%), and pyoderma gangrenosum with vasculopathy (5%).

**Image: LACIV retiform purpura classification. A. Grade 1:** livedo reticularis or racemosa with incipient purpura (individual lesions≤1 cm)**. B. Grade 2:** More extended purpuric lesions which sometimes coalesce (individual lesions>1 cm)**. C. Grade 3:** Purpuric lesions with haemorrhagic blisters**. D. Grade 4:** Deep purpuric lesions with associated ulceration.




**Conclusions:** Given the higher consumption of cocaine and its contamination with levamisole, the report of LACIV patients is increasing. A classification of the skin involvement in LACIV is proposed, according to the frequency of affection and the stratification of purpuric lesions in four degrees of severity. Cutaneous involvement is one of the pillars for the diagnosis and properly treatment, therefore a detailed description of distribution and characteristics of the lesions are fundamental for these patients care.

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