

# The Sleight of Hand: Paraneoplastic Acral Vascular Syndrome

Elizabeth Sargent, DO<sup>1,4</sup>; Hima Doppalapudi, MD<sup>2,4</sup>; Kellie Zawisa, DO<sup>1,4</sup>; Elizabeth Brooks, DO<sup>3,4</sup>

1. McLaren Macomb Family Medicine Residency Program, Mt. Clemens, MI

2. McLaren Macomb Internal Medicine Residency Program, Mt. Clemens, MI

3. McLaren Macomb Internal Medicine Residency Program, Assistant Program Director, Mt. Clemens, MI

4. Michigan State University College of Osteopathic Medicine, East Lansing, MI



## Introduction

- Paraneoplastic acral vascular syndrome is a condition that may range from Raynaud's phenomenon to gangrene.
- It can change throughout the course of a patient's malignancy.
- A suspicion for this condition should be considered when a patient presents with new onset digital ischemia without cardiovascular or rheumatologic risk factors.
- Traditionally, Raynaud's phenomena is an intense vasoconstriction most often due to cold or stress.
- Primary Raynaud's phenomenon occurs in the absence of another disorder.
- Secondary Raynaud's phenomenon has multiple causes, including malignancy, as will be presented in this case.

## Case Presentation

- A 58-year-old man with a history of newly diagnosed metastatic rectosigmoid adenocarcinoma presented to the hospital with generalized anasarca and pain. He was also found to have discoloration of multiple fingers and small areas of dry gangrene on two fingers.
- PMH:** colorectal cancer
- PSH:** L2-L4 fusion
- Allergies:** Demerol
- Medications:** HCTZ 25mg once daily, Aspirin prn
- FM:** Heart disease; No known cancer
- SH:** Quit smoking 25 years ago, drinks 3-7 beers a day
- Vitals:** Temp 98.1, BP 162/93, RR 18, HF 101, O2 sat 97% RA
- Physical exam:** erythematous discoloration of multiple fingers and two small non-tender areas of dry gangrene on his right third and left fourth digits. Radial pulses were palpable bilaterally and there was no discoloration of his toes. (Figures 1,2,3)



FIGURE 1

## Diagnostics

WBC	10.31 <sup>3</sup> /uL
Hg	11.7 g/dL
Hct	34.6%
Plt	444 <sup>3</sup> /uL
ANA	Positive
Scl 70	Negative
Complement C3	Normal
Complement C4	Normal
Fibrinogen	582.1 mg/dl
CEA	CEA 18.9 mg/ml

CTA Thorax	Bilateral distal pulmonary emboli
Lower Extremity Dopplers	Acute popliteal gastrocnemius DVT
Bilateral upper extremity arterial doppler	Small vessel disease and possible vasospasm



FIGURE 2

## Hospital Course

- He was diagnosed with colon cancer three weeks prior to this admission and noticed discoloration of his fingers the week of his diagnosis of cancer.
- He had no history of Reynaud's phenomenon. He was seen by his primary care physician for the discoloration and was given an antibiotic. However, his symptoms did not improve even after completing the course of antibiotics.
- The tumor burden was significant causing near obstruction of the distal colon and deemed non-resectable due to adherence to the pelvic wall.
- Although not given to treat his digital ischemia, the patient received several medications during his hospitalizations that have been used in the treatment of digital ischemia associated with malignancy including anticoagulation, aspirin, amlodipine and prednisone. Unfortunately, none of these medications improved his digital ischemia.
- He progressively declined and decided to transition to hospice care.
- The patient's digital lesions continued to progress with acrocyanosis of all digits and three lesions of gangrene.

## Discussion

- Paraneoplastic acral vascular syndrome may present before, at the time of, or shortly after the diagnosis of malignancy.
- Different theories have been suggested to explain the mechanism behind paraneoplastic acral vascular syndrome:
  - Vasospasm is the main mechanism suggested due to increased sympathetic activation from either vasoconstrictive substances produced by the tumor or by malignant invasion of the sympathetic nerves
  - Other possible mechanisms include fragments of tumor, emboli from nonbacterial thrombotic endocarditis, myeloproliferative disorders, hyper-viscosity of paraproteinemia, or immunologic mechanism
- Malignancy may cause both venous and arterial hypercoagulability.
- Medications that may provide some improvement in symptoms include calcium channel blockers, vasodilators, aspirin, prostacyclin analogs, anticoagulation, antiplatelets, steroids, and topical nitroglycerin.
- Even when clinical remission is obtained the complete disappearance of digital symptoms is only obtained in less than half of patients.
- Some findings suggestive of paraneoplastic acral vascular syndrome in a patient with new onset digital ischemia include presentation over the age of 50, lower hemoglobin levels, elevated platelet count, positive ANA and positive cryoglobulin.
- The greatest value from knowledge of this condition comes in these situations where a patient exhibits new onset digital ischemia without obvious cause.



FIGURE 3

## References

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