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Title

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Journal

Proceedings of UCLA Health, 27(1)

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Publication Date

2023-06-15

CLINICAL VIGNETTE

An Elderly Man with Cancer and Acrocyanosis

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A 71-year-old man with an unknown prior past medical history was admitted to the hospital with generalized weakness after a syncopal event with a fall. His only presenting symptom in addition to syncope was generalized weakness over several months. During his hospitalization however, he had multiple complications. He was found to have decompensated heart failure as well as a non-ST-elevation myocardial infarction, and was admitted to the intensive care unit (ICU). In the ICU, he was also noted to be in septic shock, with *E. coli* bacteremia. The patient also sustained right-sided rib fractures with pneumothorax, acute kidney injury, and required pressor support. CT of his abdomen and pelvis showed a colon mass.

The admitting medical team successfully treated his complications. Septic shock resolved with treatment of *E. coli* infection, heart failure resolved, and he was able to oxygenate well on room air. His renal function also normalized. Colonoscopy showed a fungating obstructing mass in his proximal ascending colon as well as an ulcerated non-obstructing mass in the rectosigmoid with biopsies consistent with adenocarcinoma.

The patient was discharged to a skilled nursing facility (SNF) to undergo rehabilitation. His physical exam at the time of SNF admission was remarkable for obesity, and generally appeared of chronically ill. Cardiopulmonary exam was unremarkable, but abdominal exam was notable for a firm, irregularly shaped right lower quadrant mass.

Another prominent finding was painful acral cyanosis in all ten fingers was not reversible with warming temperature. Only one of toe also showed acral cyanosis. The patient reported that prior to the recent hospitalization, he had never had reversible or irreversible cyanosis of his fingers. At the SNF, he received oxygen and amlodipine but the only action that improved his pain slightly was to put his hands under the bed covers to keep them warm. The differential diagnosis of his cyanosis included Raynaud's syndrome, thromboembolism, cold agglutinin disease, or a paraneoplastic syndrome.

Laboratory tests were notable for an unremarkable complete blood count, a normal reticulocyte count, elevated lactate dehydrogenase, and a normal haptoglobin.

Before his anticipated discharge from the SNF, some of his fingertips were starting to become necrotic and more painful. His physician referred him to rheumatology for a more exten-

sive evaluation of his acrocyanosis and digital necrosis, but the patient opted for a home discharge with hospice care and no further testing.

Paraneoplastic Acral Vascular Syndrome

Paraneoplastic acral vascular syndrome is a rare disorder presenting with digital cyanosis and ischemia including possible gangrene, in the context of a malignancy. In 50% of cases, it is associated with adenocarcinoma equal incidence in males and females.^{1,2} It presents with manifestations of digital ischemia, including Raynaud's phenomenon, acrocyanosis, and digital gangrene.³ It is different from primary Raynaud's phenomenon, in being always associated with a malignancy, rather than idiopathic or associated with systemic vasculitis or rheumatologic disorders.

The pathophysiologic mechanisms of paraneoplastic acral vascular syndrome are not well understood. According to several reports, ischemia may be caused by a vasoconstrictive substance produced by the tumor cells.¹ It also may be caused by a thromboembolic mechanism, with embolization of tumor microfragments directly in the bloodstream. The role of smoking in neoplastic ischemia is not known but may be a contributing factor. Other mechanisms include tumor invasion of the sympathetic nervous system, hyperviscosity, and an overproduction of vasoconstrictive, thrombogenic and immunologic mediators by tumor cells.⁴

Administration of intravenous prostacyclin has decreased the frequency and severity of attacks in Raynaud's phenomenon associated with scleroderma. A few patients with Raynaud's phenomenon associated with malignancy treated with prostacyclin also noted improvement in symptoms.⁵ More definitive improvement in symptoms may result from treatment of the underlying malignancy.

Discussion

This patient presented with painful acrocyanosis of all fingers and one toe, with ischemic findings and progression to necrosis, in the context of a new diagnosis of adenocarcinoma of the colon. The patient's presentation was partially consistent with Raynaud's phenomenon in that his acrocyanosis worsened with cold and included ischemia. Raynaud's phenomenon can be primary, or secondary to other disorders, frequently vasculitides or rheumatologic syndromes. In this patient, there were no signs

or history of vasculitis or rheumatologic disorders. However, we were not able to obtain a more extensive evaluation for these conditions, as the patient chose to receive comfort care.

Acrocyanosis may be present in cold agglutinin disease, where patients have autoantibodies that induce red blood cell agglutination in cooler parts of the body. Our patient laboratory testing, including normal haptoglobin and reticulocyte counts pointed made hemolysis a less likely cause of his acrocyanosis.

Thromboembolic phenomena can also lead to digital ischemia consistent with the patient's presentation. The largely symmetric pattern as well as the absence of other areas of thromboembolism lessen the likelihood that thromboembolism was the cause, though it does not rule it out.

The patient's presentation with digital ischemia in the context of an adenocarcinoma is most consistent with the rare diagnosis of paraneoplastic acral vascular syndrome. His adenocarcinoma was too advanced for cancer treatment to impact his digital ischemia. Comfort care with hospice enrollment was the most appropriate patient-centered care at the time of his discharge from the SNF.

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