

# Cutaneous Manifestations of Abdominal Arteriovenous Fistulas

Jessica Scruggs, MD; Daniel D. Bennett, MD

*Abdominal arteriovenous (A-V) fistulas may be spontaneous or secondary to trauma. The clinical manifestations of abdominal A-V fistulas are variable, but cutaneous findings are common and may be suggestive of the diagnosis. Cutaneous physical examination findings consistent with abdominal A-V fistula include lower extremity edema with cyanosis, pulsatile varicose veins, and scrotal edema.*

*We present a patient admitted to the hospital with lower extremity swelling, discoloration, and pain, as well as renal insufficiency. During a prior hospitalization she was diagnosed with venous stasis dermatitis; however, her physical examination findings were not consistent with that diagnosis. Imaging studies identified and characterized an abdominal aortocaval fistula. We propose that dermatologists add abdominal A-V fistula to the differential diagnosis of patients presenting with lower extremity edema with cyanosis, and we summarize other physical examination and laboratory findings that may suggest the diagnosis.*

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**A**bdominal arteriovenous (A-V) fistulas often present with cutaneous findings in the lower extremities that may come to the attention of a dermatologist. Although the clinical presentation of abdominal A-V fistulas varies, cutaneous findings are common and can include lower extremity edema with cyanosis, pulsating varicose veins, and scrotal

edema.<sup>1-3</sup> We report a case of abdominal aortocaval fistula presenting with lower extremity edema, erythema, and cyanosis that had been previously diagnosed as venous stasis dermatitis.

## Case Report

A 51-year-old woman presented to the emergency department with worsening lower extremity swelling, redness, and pain. Her medical history included a diagnosis of congestive heart failure, chronic obstructive pulmonary disease, hepatitis C virus, tobacco abuse, and polysubstance dependence. Swelling, redness, and pain of her legs developed several years prior, and during a prior hospitalization she had been diagnosed with chronic venous stasis dermatitis as well as neurodermatitis.

On admission, the patient had cool lower extremities associated with discoloration and many crusted ulcerations. Aside from obesity, her abdominal examination was unremarkable and no bruits were noted. Her creatinine level was 2.9 mg/dL (reference range, 0.6–1.2 mg/dL) and her blood urea nitrogen level was 63 mg/dL (reference range, 8–23 mg/dL). She had no leukocytosis, and her erythrocyte sedimentation rate and C-reactive protein level were within reference range. Her alkaline phosphatase level and transaminases were minimally elevated. A toxicology screen was positive for cocaine. Deep vein thrombosis was ruled out via negative lower extremity ultrasonography. Doppler evaluation of her lower extremities revealed normal pulses, but she did not tolerate an attempt to obtain ankle blood pressures secondary to pain. Cardiac echocardiography demonstrated an ejection fraction of 65%, bilateral atrial enlargement, high right-sided filling pressure, and a normal-appearing left ventricle. Abdominal ultrasonography revealed hepatomegaly as well as a prominent inferior vena cava (IVC) and hepatic veins. Renal ultrasonography was normal.

The dermatology department was consulted to evaluate her skin findings. The patient reported lower extremity skin pain and denied picking or scratching

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Dr. Scruggs is from and Dr. Bennett was from the Department of Dermatology, Texas A&M College of Medicine, Temple, and Scott & White Memorial Hospital, Temple. Dr. Bennett currently is from the Department of Dermatology, Hospital of the University of Pennsylvania, Philadelphia.

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Correspondence: Daniel D. Bennett, MD, Department of Dermatology, Hospital of the University of Pennsylvania, 3600 Spruce St, 2 Maloney Bldg, Philadelphia, PA 19104.

the lesions on the legs. On examination of the lower extremities, she had livedoid change and striking purplish erythema with sharp demarcation on the upper thighs (Figure 1). Her legs were cool to the touch. Slow capillary refill was noted on examination of her toes. In addition, she had multiple punched out ulcers with yellowish crusts and scattered small pustules. The patient also had excoriated crusted papules on the arms and abdomen. Some livedoid change and coolness of the skin was noted on the elbows and hands. No purpura was noted anywhere on examination. Based on the physical examination findings, a vasculopathic process was suspected, but vasculitis was deemed to be unlikely. Secondary infection and colonization of her wounds was confirmed by swab cultures that grew *Staphylococcus aureus*, coagulase-negative staphylococci, *Acinetobacter lwoffii*, and *Fusarium* species. A skin biopsy revealed focal ulceration with acute inflammation and epidermal hyperplasia with hyperkeratosis suggestive of excoriation and/or trauma. Tissue cultures for acid-fast and fungal organisms were negative. Testing for cryoglobulins, classical and perinuclear antineutrophil cytoplasmic antibodies, histoplasma antigen, and antinuclear antibody was negative.

Magnetic resonance angiogram revealed bilaterally enlarged common iliac veins with megacava, and there was venous contamination of the lower extremities suggesting a fistulous communication. A subsequent abdominal aortogram (Figure 2) showed contrast that immediately filled the IVC consistent with a fistula between the distal aorta and the IVC as well as a fistula between the right common iliac artery and right common iliac vein.

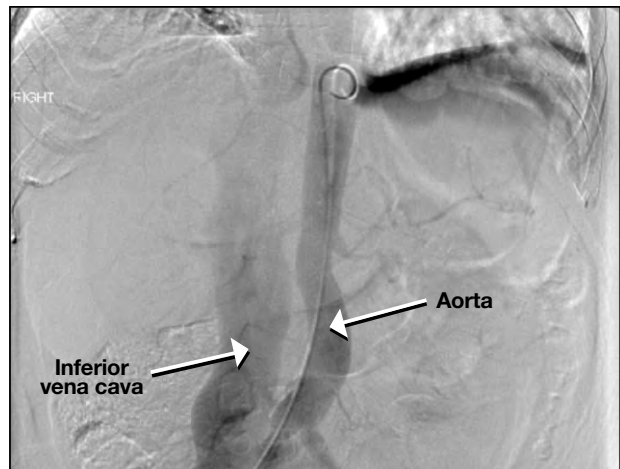
Surgical intervention was delayed because of multiple comorbid conditions including *Stenotrophomonas* bacteremia, endocarditis, and an episode of septic shock necessitating mechanical ventilation and aggressive resuscitation. With worsening of her renal function, she was deemed to be a poor surgical candidate. The patient died shortly after hospice placement.

## Comment

Most abdominal aortocaval fistulas are spontaneous, resulting from the erosion of an atherosclerotic, aortic, or iliac aneurysm into an adjacent vein. Spontaneous abdominal aortocaval fistulas also have been reported to result from syphilitic and mycotic aneurysms, as well as aneurysms from Marfan syndrome, Ehlers-Danlos syndrome, and Takayasu arteritis.<sup>2</sup> Traumatic abdominal A-V fistulas may occur following penetrating and blunt trauma in addition to iatrogenic injury during diagnostic or surgical procedures.<sup>1</sup> Although our patient was found to have an aortocaval



**Figure 1.** Erythema, edema, and cyanotic livedoid patches involved the legs. Note the abrupt color change on the upper thighs. Multiple punched out and crusted ulcers also were present.



**Figure 2.** An abdominal aortogram showed contrast that immediately filled the inferior vena cava.

fistula, similar clinical findings also may be expected with large fistulas involving the abdominal iliac vessels. Abdominal A-V fistulas between the iliac artery and iliac vein are rare and more frequently result from trauma.<sup>4</sup>

The clinical presentation of abdominal A-V fistula is variable; therefore, the diagnosis may be overlooked. Abdominal pain with a pulsatile abdominal mass, a machinelike abdominal bruit, and acute dyspnea are believed to be pathognomonic for abdominal A-V fistulas.<sup>2</sup> Some cases present without typical findings. Other signs and symptoms include back pain; high-output cardiac failure; renal failure; systemic hypotension; and signs of venous hypertension including scrotal edema, pulsating varicosities, priapism, hematuria, and rectal bleeding.<sup>1,3</sup> In a discussion of the diagnosis of abdominal A-V fistulas, Gilling-Smith and Mansfield<sup>3</sup> referred to “the triad of machinery murmur, high output cardiac failure and

regional venous hypertension” and noted that it was present in only 25 of 148 cases they reviewed. In the same review, leg edema was noted in 53 patients and cyanosis in 34.<sup>3</sup> Another review found lower extremity edema with mottling and cyanosis to be the primary manifestation in 8 of 20 patients. These authors also described frequent findings of cyanotic congested lower extremities that contrasted with cool pale upper extremities and trunk.<sup>2</sup>

Prompt diagnosis is desirable and treatment is prompt surgical repair. Unfortunately, the diagnosis often is missed or delayed. In one review, the diagnosis was not made prior to surgery in 38% of cases.<sup>1</sup> In another review, the diagnosis was made intraoperatively in 25% of cases.<sup>2</sup> Multiple radiographic studies may be helpful in the diagnosis and localization of abdominal A-V fistulas. Abdominal color Doppler ultrasonography, computed tomography, and magnetic resonance imaging are noninvasive and often sufficient.<sup>5,6</sup> Angiography may be useful if the diagnosis is unclear or if more detail is necessary prior to surgery.<sup>2,6</sup> Multidetector-row computed tomography and virtual angiography also have been suggested to delineate the anatomy of a fistula prior to surgery.<sup>7</sup>

Our patient’s chronic course, obesity, and history of substance abuse all likely contributed to the delayed diagnosis of abdominal aortocaval fistula. The lack of eczematous changes or evidence of hemosiderin deposition militated against a diagnosis of stasis dermatitis. We found the purplish color of her skin to be suggestive of vascular compromise, and we hypothesized that the sharp demarcation noted on the upper thigh was suggestive of involvement of the deep vessels of the leg. Our patient’s echocardiographic findings of high right-sided filling pressure and abdominal ultrasonography findings of enlarged abdominal veins

also were clues to the diagnosis. Her renal insufficiency also was likely due to her A-V fistula.

Given the prevalence of cutaneous findings in abdominal A-V fistula, dermatologists are likely to encounter similar patients, both as inpatients and outpatients. We propose that dermatologists add abdominal A-V fistula to the differential diagnosis of patients presenting with edematous lower extremities, especially when associated with cyanosis and symptoms of heart failure. Physical examination and laboratory findings are highly variable; therefore, a high index of suspicion must be maintained and appropriate studies and consultations must be obtained urgently.

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