Lower limb ulcers due to an absent inferior vena cava

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Key words: chronic venous obstruction; compression dressings; inferior vena cava; leg ulcer; varicose vein.

INTRODUCTION

Chronic obstruction of the inferior vena cava (IVC) is a relatively rare condition that may be caused by a congenital caval abnormality or previous IVC thrombosis.[1,2] It is usually asymptomatic and mostly an incidental finding.[3] In some cases, however, the absence of the IVC can produce symptoms from chronic venous obstruction, such as heaviness, edema, dermatitis, and ulcers.[4] Here we report a male patient suffering from leg ulceration due to absence of the IVC.

Abbreviations used:

DVT: deep vein thrombosis
IVC: inferior vena cava

CASE REPORT

A 43-year−old man presented at our hospital with a 2-year history of lower limb ulcers. He had had varicose veins for many years but was otherwise healthy. Initial examination demonstrated varicose veins and mild edema in both the lower limbs. In addition to the varicose veins, there were brownish indurated plaques and hardened plaques with multiple walnut-sized ulcers. The ulcers were partially covered with fibrin and were painful (Fig 1).

The results of blood tests were within normal limits. The ankle brachial pressure index was normal, and Doppler examination of the lower limbs revealed no abnormality and no stenosis or occlusion of the lower limb arteries. Deep vein thrombosis (DVT) was ruled out because D-dimer was normal and ultrasound images of the lower extremities showed no thrombosis. Computed tomography imaging revealed absence of the IVC as well as marked dilation of the paraspinal venous plexus and the cutaneous and azygos veins (Fig 2). There were well-developed collaterals over the anterior abdominal wall and in the bilateral lower limbs. On the basis of these findings, we diagnosed the patient as having leg ulceration because of the absence of the IVC and administered conservative therapy with 1% silver sulfadiazine and application of compression dressings. After 4 months, the ulcers healed, leaving pigmentation. Since then, the patient has continued to use compression bandages.

DISCUSSION

Chronic obstruction of the IVC is a relatively rare disorder that may be caused by a congenital caval abnormality or previous IVC thrombosis.[1,2] Patients with chronic obstruction of the IVC usually present with DVT or recurrent nonhealing venous leg ulcers.[2,5,6] The age at presentation is variable. The cause of ulceration due to chronic obstruction of the IVC is increased venous pressure, leading to the development of collateral circulation in the lower limbs and abdominal wall. This collateral circulation can cause valvular dysfunction, and venous hyperpressure may induce valvular damage, leading to skin changes such as pigmentation and sclerosis, and finally leg ulcers. Because computed tomography imaging and history revealed no cause of IVC stenosis in this case, the leg ulcers were considered due to IVC deficiency.

There is disagreement in the literature regarding the cause of the absence of the infrarenal IVC. Although it has been suggested that the absence of the IVC may be a developmental defect, it is difficult to determine what might satisfactorily account for

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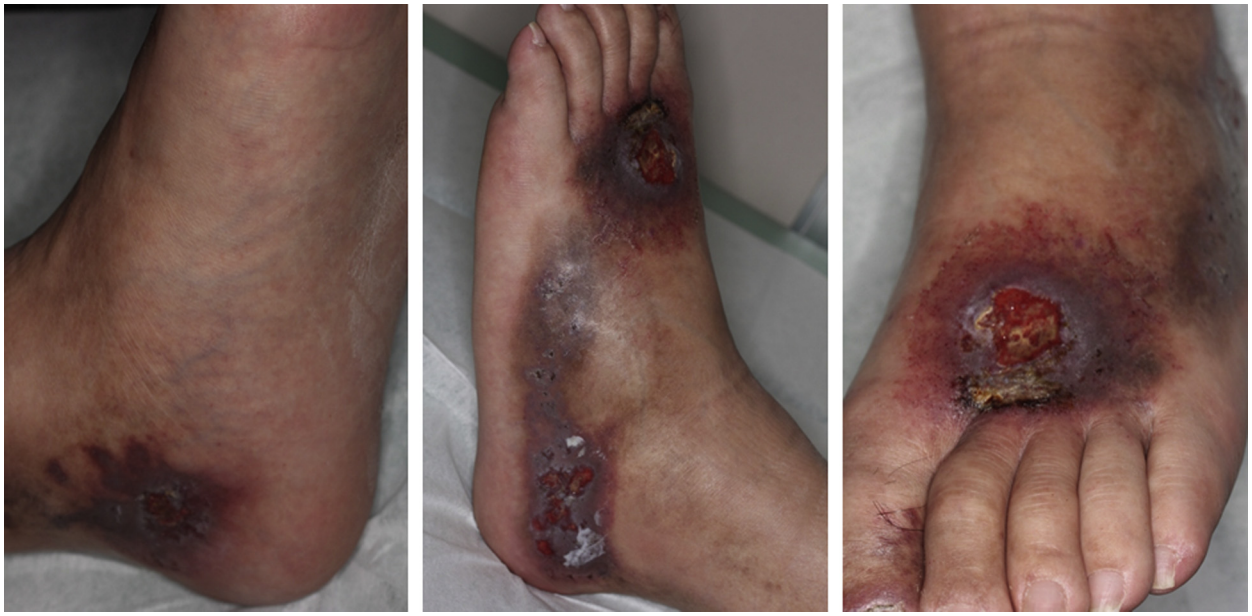

Fig 1. Clinical features. Nonhealing venous ulcer and pigmentation on the left leg.

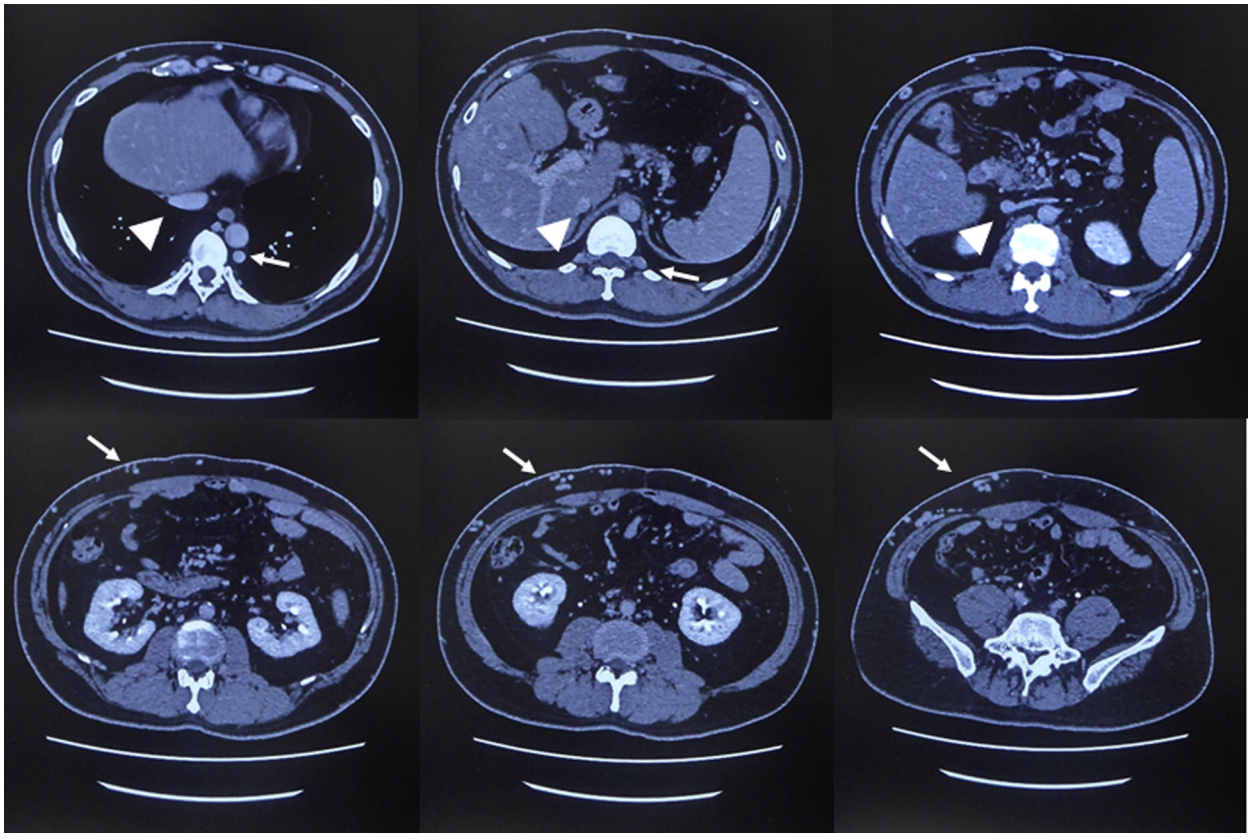

Fig 2. *From top left to bottom right:* Multiple transaxial computed tomography images extending from the level of the mid-thorax to the level of the mid-abdomen. Images show the absence of the inferior vena cava under the connection of the hepatic veins (*arrowheads*). Pronounced dilation of the paravertebral venous plexus and the cutaneous and azygos veins (*arrows*).

this anomaly.[6] It has also been suggested that the absence of the subrenal IVC may result from intrauterine or perinatal IVC thrombosis and may not be a developmental disorder.[7] Some adolescents with DVT and absent IVC have a history of perinatal IVC thrombosis.[8] Existing data suggest that such perinatal thrombosis may underlie cases of absent IVC detected later in life. In the absence of early symptoms, absence of the IVC can manifest as hypertension, cyanosis, dyspnea, heart failure, cardiac enlargement, and childhood developmental delay.[9] In other cases, it may be manifested symptomatically by chronic venous occlusion with associated skin changes, such as severe edema or leg ulcers,[2] or it can be a potential independent risk factor for venous thromboembolism.

As therapeutic options for absent IVC, surgical or conservative treatment can be considered. Anticoagulation in combination with compression dressings has been the traditional treatment option for patients with IVC obstruction. Conservative treatment may relieve symptoms and prevent recurrent thrombosis.[6] A venous bypass graft from the common iliac vein to the azygos vein can be considered for prevention of DVT, or if conservative treatment fails to heal the ulcers.[6] In our patient, the ulcer disappeared after treatment with topical ointment and compression dressings. However, we will need to perform long-term follow-up and consider treatment choices and additions over time.

In conclusion, we have reported a patient with lower limb ulcers associated with the absence of the IVC. When encountering young patients with unexplained venous thrombosis, recurrent refractory lower extremity ulcers, varicose veins, and enlarged abdominal veins, IVC deficiency should be considered. When the presence of this condition is recognized, detailed evaluation can lead to a correct diagnosis early in the course of the disease. For treatment, patient care needs to be individualized until strong objective data become available.

Conflicts of interest

None disclosed.

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