

## CLINICAL REPORT

# Leg Ulceration in Chronic Venous Insufficiency Caused by an Absent Inferior Vena Cava

Hiroo AMANO, Yayoi NAGAI, Yukie ENDO, Tomoko IWASAKI and Osamu ISHIKAWA  
Department of Dermatology, Gunma University Graduate School of Medicine, Maebashi, Gunma, Japan

We report here the case of a 55-year-old Japanese man with a one-year history of multiple ulcers on the left crural region. He had had pronounced varicose veins on both legs and the abdominal region for 35 years. Computed tomography images of the thoracic and abdominal regions showed the absence of an inferior vena cava, with pronounced dilatation of paravertebral venous plexus, cutaneous and azygous veins. Conservative topical treatments led to complete healing of the ulcers in one month. An absent inferior vena cava is an uncommon abnormality, often complicated by cardiac and other visceral malformations. It is a rare cause of chronic leg ulcers. **Key words:** chronic venous insufficiency; leg ulcer; inferior vena cava; varicose veins.

(Accepted April 27, 2009.)

Acta Derm Venereol 2009; 89: 502–504.

Hiroo Amano, Department of Dermatology, Gunma University Graduate School of Medicine, 3-39-22 Showa-machi, Maebashi, Gunma 371-8511, Japan. E-mail: hamano@showa.gunma-u.ac.jp

An absent inferior vena cava (IVC) is a rare congenital disease (1). When accompanied by cardiac or visceral malformations, systemic symptoms, such as cyanosis,

dyspnoea, cardiac failure, cardiac growth and development delay, can occur (2). In rare cases, absent IVC may manifest itself symptomatically through chronic venous obstruction, with heaviness, cramps, oedema, or associated dermal changes, such as leg ulceration (3–5). We report here the case of a 55-year-old man with pronounced varicose veins and leg ulcerations caused by absent IVC.

## CASE REPORT

A 55-year-old man was referred to our department on 24 January 2005, with a one-year history of ulceration on the left crural region. He had had varicose veins for 35 years; however, he was otherwise healthy. In order to treat the varicose veins, the saphenous veins had been removed by stripping 30 years previously. This procedure was not effective, and in fact, it aggravated the symptoms. Physical examination revealed pronounced varicose veins and brownish indurated plaques on his lower legs, with multiple thumb finger-sized ulcers (Fig. 1a). Coalescent and vermiculated ulcers were covered with yellowish necrosis (Fig. 1b). Pronounced varicose veins were also noted on the back of the lower legs and on the abdominal wall (Fig. 2a and b). The results of routine laboratory tests including haematology, liver and renal functions, as well as CRP levels, were within normal limits. Anti-nuclear antibodies, anti-mitochondrial antibodies, anti-smooth muscle antibodies, peri-antinuclear cytoplasmic antibodies, anti  $\beta$ 2-GPI antibodies, and lupus anti-coagulant



Fig. 1. (a) Clinical features. Pronounced varicose veins and pigmentation on both legs and dorsal pedis. (b) Multiple thumb finger-sized ulcers with yellowish necrosis on the brownish induration. Multiple ulcers were coalescent and vermiculated.

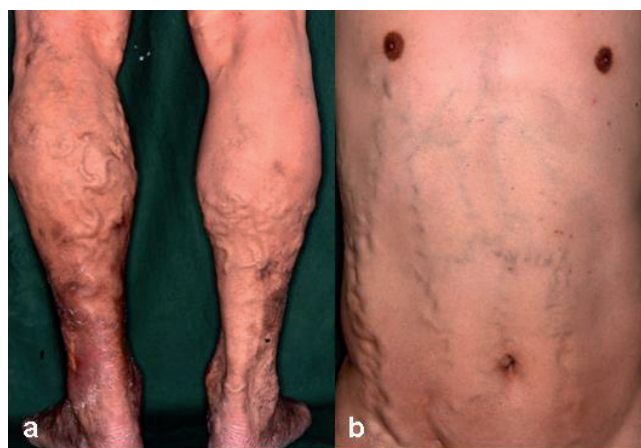


Fig. 2. (a and b) Clinical features. Pronounced varicose veins were also noted on the back of the lower legs and on the abdominal wall.

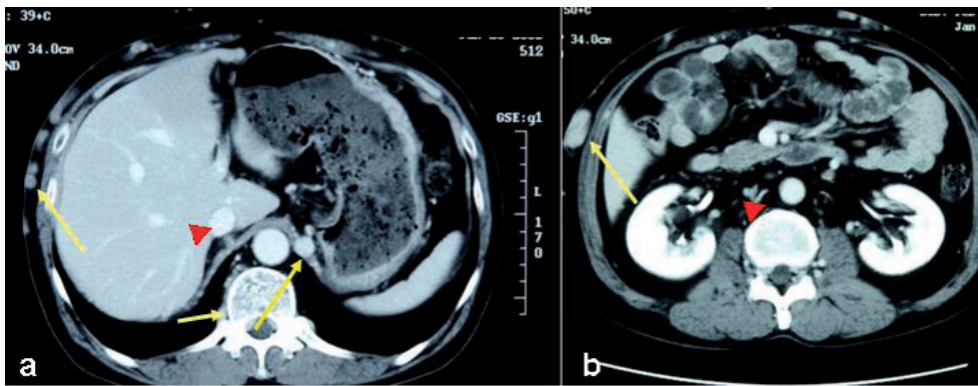


Fig. 3. Computerized tomography images of (a) thoracic and (b) abdominal regions. Pronounced dilation of the paravertebral venous plexus, cutaneous and azygos veins (yellow arrows) and inferior vena cava (red arrowheads) in (a) and the absence of inferior vena cava under the connection of hepatic veins with pronounced dilation of cutaneous veins in (b).

were all negative. Levels of protein C and protein S were within normal limits. Since prominent varicose veins were seen on the abdominal wall, we performed a thorough examination to detect underlying diseases. Computerized tomography (CT) images of the thoracic and abdominal regions showed the absence of the IVC with pronounced dilatation of the paravertebral venous plexus, cutaneous and azygos veins (Figs 3a and b, and 4). Magnetic resonance imaging angiography revealed the dilatation of collateral veins on the femoral and crural regions. Based on the above findings, we diagnosed leg ulceration due to absent IVC. Surgical treatment was not indicated because of the risk of venous thrombosis due to long bypass grafting. We used a conservative therapy consisting of 1% silver sulfadiazine and physical therapy, such as compression dressing. One month later, the ulcer healed leaving a slight hypertrophic scar. The patient has continued to use compression dressing. No relapse has been noted for the past 2 years. Since the absent inferior vena cava is congenital, compression dressing should be used to control the disease.

## DISCUSSION

Leg ulceration is a common inflammatory skin disease triggered by venous, arterial, and neuropathic factors

(6). Venous ulceration constitutes the majority of all leg ulcers, and is caused by increased pressure in the venous system. This sustained ambulatory pressure has been termed "venous hypertension", and is also referred to as "chronic venous insufficiency" (7, 8). Venous insufficiency occurs due to various conditions, such as: (i) dysfunction of valves in the superficial and/or communicating veins because of congenital or acquired incompetence; (ii) dysfunction of valves in the deep system because of congenital absence, inherent weakness, or thrombotic damage; (iii) deep venous outflow obstruction rather than valvular incompetence; and (iv) muscle dysfunction and calf muscle pump failure from inflammatory conditions of the joints or muscles, fibrosis, or neuropathies (7).

In older patients, the cause of venous insufficiency is often obesity (9). In contrast, younger patients with varicose veins tend to be female and may have a history of phlebitis and/or a family history of varicose veins. Patients with venous insufficiency are more likely to have heart disease.

Congenital absence of valves, previous surgery for varicose veins, primary valve or venous wall degeneration, arteriovenous shunts, and congenital vascular malformation can be the cause of venous insufficiency. Absent IVC can be one cause of venous insufficiency; however, it is very rare.

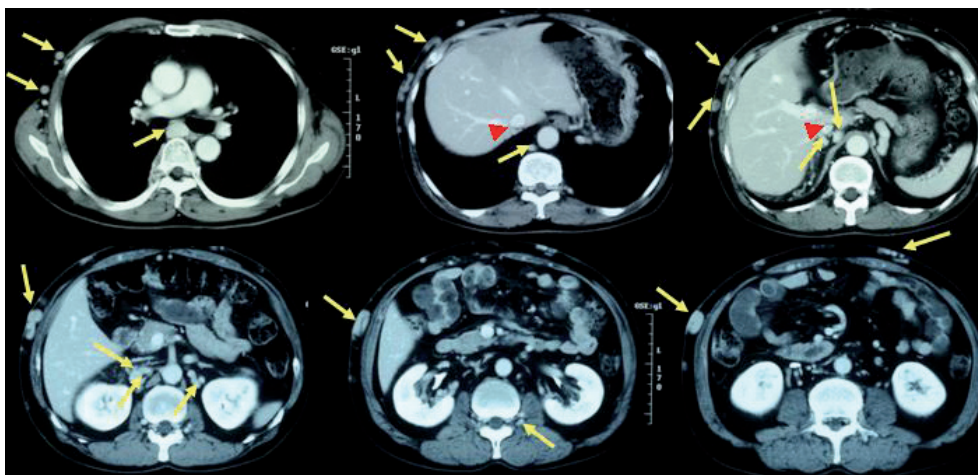


Fig. 4. From top left to bottom right: Multiple transaxial computerized tomography images extending from the level of the mid thorax to the level of the mid abdomen. Images showed the absence of inferior vena cava under the connection of the hepatic veins (red arrowheads). Pronounced dilation of the paravertebral venous plexus, cutaneous and azygos veins (yellow arrows).

The critical period for the fusion of the IVC is between days 33 and 40 of embryonic life, which is the same period for the organization of the heart. Developmental errors and a genetic predisposition may affect the processes responsible for the formation of abnormal conduction pathways during cardiogenesis. Therefore, absent IVC is often complicated by cardiac or other visceral malformations, including *situs inversus*, asplenia and polysplenicism. Initial symptoms may present as hypertension, cyanosis, dyspnoea, cardiac failure, cardiac growth and development delay in childhood (1, 10). Rarely, absent IVC may manifest itself symptomatically through chronic venous obstruction with heaviness, cramps, oedema, or associated dermal changes, such as leg ulcers (3, 4).

Absent IVC can also be diagnosed incidentally in medical examinations. It has been reported that the incidence of absent IVC was 1% in patients who underwent cardiac catheterization and 0.6% in patients investigated for congenital heart disease (3). Inama et al. (11) reported a case incidentally diagnosed during catheter ablation for Wolff-Parkinson-White syndrome. They also confirmed that a CT scan showed the absence of IVC and the presence of azygous continuation. To diagnose absent IVC, CT scans, venography and magnetic resonance imaging have proven useful.

Furthermore, absent IVC could be a potential independent risk factor for venous thromboembolism. Ruggeri et al. (12) estimated that absent IVC was present in approximately 5% of young patients under 30 years of age with deep vein thrombosis (DVT). Siragusa et al. (13) reported two patients with aplasia or agenesis of IVC out of 21 patients who had acute venous thromboembolism. They suggested that absent IVC might not be a rare finding in young patients with DVT or pulmonary embolism.

One of the treatment options for absent IVC may be venous bypass with a graft from the common iliac vein to the azygous vein in the chest via a retroperitoneal approach. However, when surgical treatment is not indicated, conservative therapy, such as local wound care and physical therapy, should be used. In our case, although the patient had undergone ambulatory phlebectomy (stripping of the saphenous veins) 30 years previously, the procedure was not effective, and it aggravated the symptoms. Surgical intervention should be evaluated carefully, as varicose veins caused by absent IVC are not candidates for phlebectomy.

As stated previously, DVT, lung embolization, arrhythmia and cardiac failure could lead to the diagnosis

of absent IVC. Additionally, pronounced varicose veins and leg ulcerations can be rare initial symptoms, as in our patient. Absent IVC is rare and might be difficult to diagnose solely from a clinical evaluation. The symptoms are variable and some patients are asymptomatic. However, when we encounter patients with prominent varicose veins located on the abdominal wall as well as the lower legs and/or refractory leg ulceration, absent IVC should be considered as a potential underlying disease. Furthermore, if the existence of this condition is recognized, detailed evaluation can lead to a correct diagnosis early in the course of the disease.

## REFERENCES

1. Arrazola L, Long A, Moss A, Sozen H, Najarian J. An absent inferior vena cava in a pediatric renal transplant recipient. *Clin Transplant* 2000; 14: 360–362.
2. Downing DF. Absence of the inferior vena cava. *Pediatrics* 1953; 12: 675–680.
3. Debing E, Tielemans Y, Jolie E, Van den Brande P. Congenital absence of inferior vena cava. *Eur J Vasc Surg* 1993; 7: 201–203.
4. Dougherty MJ, Calligaro KD, DeLaurentis DA. Congenitally absent inferior vena cava presenting in adulthood with venous stasis and ulceration: a surgically treated case. *J Vasc Surg* 1996; 23: 141–146.
5. Tofigh AM, Coscas R, Koskas F, Kieffer E. Surgical management of deep venous insufficiency caused by congenital absence of the infrarenal inferior vena cava. *Vasc Endovascular Surg* 2008; 42: 58–61.
6. Mekkes JR, Loots MA, Van Der Wal AC, Bos JD. Causes, investigation and treatment of leg ulceration. *Br J Dermatol* 2003; 148: 388–401.
7. Valencia IC, Falabella A, Kirsner RS, Eaglstein WH. Chronic venous insufficiency and venous leg ulceration. *J Am Acad Dermatol* 2001; 44: 401–421.
8. Alguire PC, Mathes BM. Chronic venous insufficiency and venous ulceration. *J Gen Intern Med* 1997; 12: 374–383.
9. Scott TE, LaMorte WW, Gorin DR, Menzoian JO. Risk factors for chronic venous insufficiency: a dual case-control study. *J Vasc Surg* 1995; 22: 622–628.
10. Lash C, Radhakrishnan J, McFadden JC. Renal vein thrombosis secondary to absent inferior vena cava. *Urology* 1998; 51: 829–830.
11. Inama G, Vergara G, Gramegna L, Rillo M, Fuochi C, Furlanello FJ. Catheter ablation of Wolff-Parkinson-White syndrome associated with congenital absence of inferior vena cava. *J Interv Card Electrophysiol* 1998; 2: 301–304.
12. Ruggeri M, Toretto A, Castaman G, Rodeghiero F. Congenital absence of the inferior vena cava: a rare risk factor for the idiopathic deep-vein thrombosis. *Lancet* 2001; 357: 441.
13. Siragusa S, Anastasio R, Falaschi F, Bonalumi G, Bressan MA. Congenital absence of inferior vena cava. *Lancet* 2001; 357: 1711–1712.