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Primary leiomyosarcoma of inferior vena cava adjacent to hepatic veins: complete off-pump resection and inferior vena cava graft reconstruction with application of external skin surface cooling

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Abstract

OBJECTIVES: Resection of a leiomyosarcoma of the inferior vena cava (IVC) requires venovenous bypass, especially if IVC clamping above the hepatic veins is planned. This report describes the application of external skin surface cooling for off-pump resection of a primary IVC leiomyosarcoma adjacent to the hepatic veins with graft IVC reconstruction in conditions of suprahepatic caval clamping and uninterrupted Pringle's manoeuvre.

METHODS: A 62-year-old woman presented with IVC leiomyosarcoma adjacent to the hepatic veins. After anaesthesia induction, the patient's head, neck, abdomen, chest, thighs and shanks were covered by polyethylene bags containing granulated ice. The ice bag was also placed between the internal surfaces of the thighs. After 108 min, the target oesophageal temperature (29°C) was achieved, all bags were removed, except the bags that were used for temperature control during the operation located on head, neck and between internal surfaces of the thighs. Off-pump resection of the tumour was performed in conditions of suprahepatic IVC and portal triad clamping via median laparotomy.

RESULTS: IVC clamping was accompanied by arterial hypotension of 55–65 mmHg well-tolerating in the hypothermic background for 69 min. The simultaneous application of an uninterrupted Pringle's manoeuvre was accompanied by low levels of transaminases and bilirubin. There was no local relapse of the tumour or metastases over a 6-month follow-up.

CONCLUSIONS: External skin surface cooling in cases demanding IVC clamping above the hepatic veins and Pringle's manoeuvre allows avoiding venovenous bypass. This method is safe, cost effective, easily performed and may be used in clinical cases involving systemic arterial hypotension.

Keywords: Leiomyosarcoma of inferior vena cava • Skin surface cooling

INTRODUCTION

A primary inferior vena cava (IVC) leiomyosarcoma is a rarely encountered malignancy. A total of 322 cases have been described in the literature [1]. Resection remains the only option to prolong survival [2] but may present a challenge. The management of tumours that have extension towards the hepatic veins or have origin in the upper IVC segment may require suprahepatic IVC clamping resulting in systemic arterial hypotension with the necessity of venovenous bypass and Pringle's manoeuvre with risk of hepatic ischaemia. The use of external skin surface cooling (ESSC) for off-pump resection of an IVC leiomyosarcoma adjacent to the hepatic veins with graft IVC reconstruction in conditions of suprahepatic caval clamping and uninterrupted Pringle's manoeuvre is herein described.

CASE AND TECHNIQUE

A 62-year-old woman presented with permanent right-sided abdominal pain during the last 3 months. The patient's weight decreased from 75 to 64 kg during this time period. Physical examination revealed no oedema of lower extremities or enlargement of the liver. Computed tomography of the abdomen and retrograde inferior cavography demonstrated a right retroperitoneal tumour (8.0 × 9.0 × 7.5 cm), extending to the hepatic veins and into the partially patent IVC lumen (Fig. 1A and B). The inferior tumoural edge had invaded the right suprarenal gland and superior pole of the kidney. The Budd–Chiari syndrome was not presented. Caudally the tumour extended along the IVC until the level above the entrance of renal veins. The off-pump tumour resection combined with graft IVC

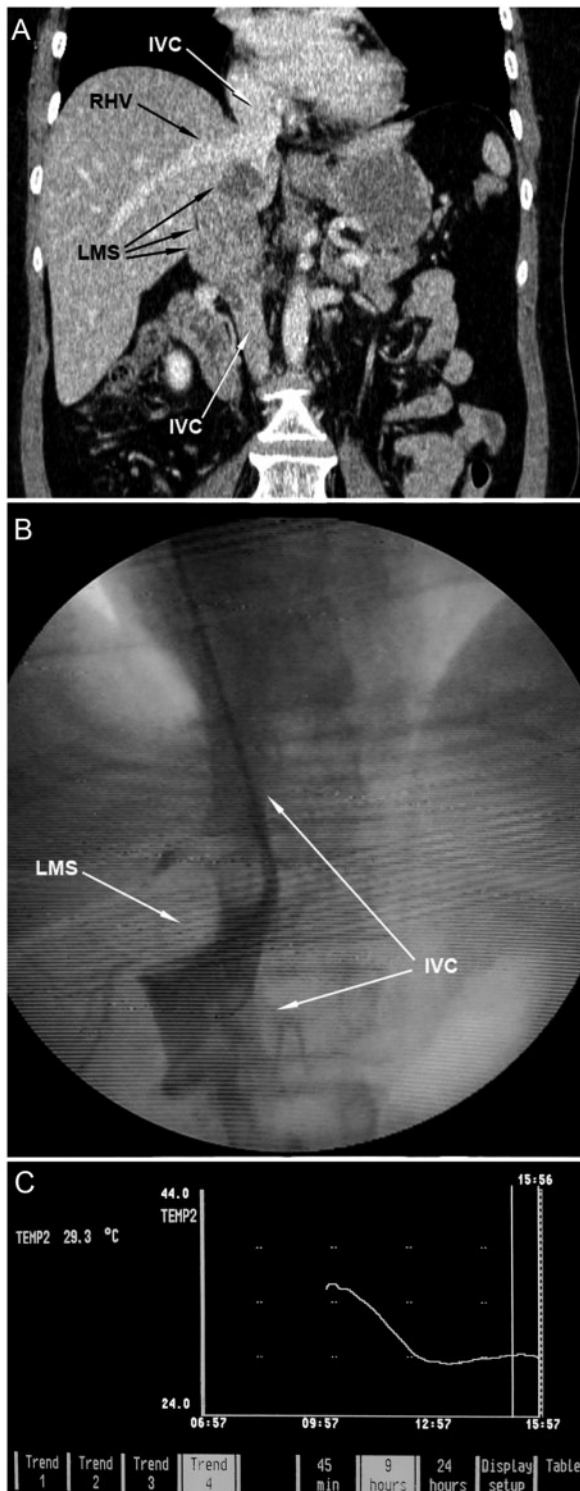


Figure 1: (A) Preoperative coronal computed tomography scan. (B) Retrograde cavography, showing patency of the IVC lumen. (C) The trends in the patient's body temperature, which decreased from 36.6°C to 29°C in 108 min. LMS: leiomyosarcoma; IVC: inferior vena cava; RHV: right hepatic vein.

reconstruction was undertaken by the technique described below.

The patient was placed on the operation table in a supine position. After anaesthesia induction, tracheal intubation, setting of arterial, venous, uric lines and the administration of double

doses of muscle relaxant, the patient's body was covered by polyethylene bags containing granulated ice. It was ensured that as much as possible of the skin's surface was in contact with the ice bags. A water ice generator was used to obtain a sufficient quantity of ice. The granules were put in polyethylene bags of approximately 30 × 20 cm, in 1 or 2 layers maximum to avoid excessive pressure on the skin. The front and sides of the abdomen, chest, thighs and shanks were covered by ice bags. Also, ice bags were placed on the patient's head, neck and between internal surfaces of the thighs. When the patient's temperature achieved the target level (29°C), all bags were removed, except the bags placed on the patient's head, neck and those between internal surfaces of the thighs. The bags that remained in place were used for further control of the patient's temperature during the operation. The patient's oesophageal temperature decreased from 36.6°C to 29°C after 108 min (rate of temperature decrease = 14–16 min/°C) (Fig. 1C). Sinus rhythm with the trend to bradycardia was present during cooling to 30°C and was then converted to atrial fibrillation. There were no ventricular arrhythmias. Antiarrhythmic drugs were not used during the operation.

After median laparotomy and tumour dissection, heparin was administered (3 mg/kg; activated clotting time, ≥ 600 s). The clamps were then placed above the hepatic veins, infrarenal segment of the IVC, left renal vein and hepatic triad. Systemic systolic arterial pressure decreased to 55–65 mmHg but was tolerated well in the hypothermic background. *En bloc* resection of the tumour, the affected IVC segment, right suprarenal gland and kidney was performed (Fig. 2A and B). Bleeding was collected with a blood sucker and returned completely via a catheter located in the superior vena cava. The linear polytetrafluoroethylene graft with external ring support was used for IVC reconstruction. Proximal anastomosis of the graft and upper IVC segment was done. The clamp, which was located at the cavoatrial junction, was relocated to the graft below the confluence of the hepatic veins, with subsequent cessation of Pringle's manoeuvre. Distal anastomosis was performed allowing drainage of the left renal vein. The duration of uninterrupted Pringle's manoeuvre and systolic arterial hypotension was 69 min. The total time of IVC clamping was 82 min. After restoration of the arterial pressure, the effects of heparin were reversed with protamine. After drainage, the wound was closed. The patient's rewarming was started in the operating room using a warming mattress and continued in the intensive care unit using a warming fan. The postoperative course was uneventful. The patient was extubated 12 h after operation. The levels of alanine aminotransferase, creatine phosphokinase and bilirubin were low (Fig. 2C). The patient was discharged from hospital 10 days later. The daily dose of warfarin was adjusted with target International normalized ratio 2.0–3.0 for permanent intake. Histopathologically and immunohistochemically, the tumour was consistent with the diagnosis of a high-grade leiomyosarcoma. A biopsy of the margin zones was negative for tumour cell content. The patient did not receive chemotherapy. The follow-up time was 6 months, without signs of local relapse of the tumour or metastases. Control abdominal ultrasound and computed tomographic examinations showed complete IVC graft patency (Fig. 3A–C).

DISCUSSION

Although development of collaterals and liberal administration of intravenous fluids may result in maintaining arterial pressure after suprahepatic IVC clamping, it is not possible to predict the

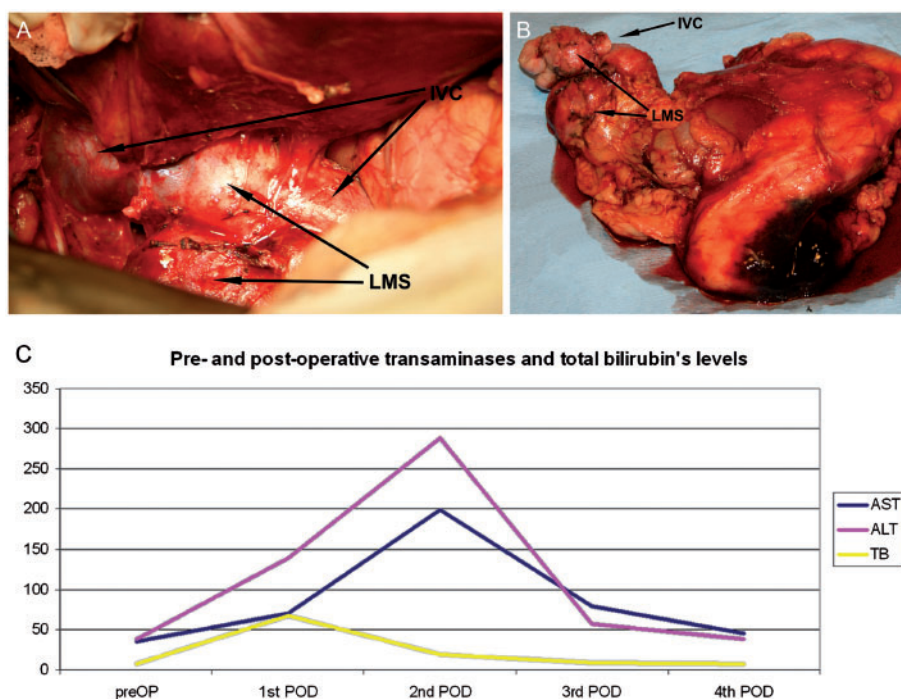


Figure 2: (A) Operative view of both the intra- and extraluminal parts of IVC leiomyosarcoma; (B) the tumour, removed *en bloc*; (C) pre- and postoperative transaminases (U/l) and bilirubin ($\mu\text{mol/l}$) levels. preOP: preoperative; POD: postoperative day; LMS: leiomyosarcoma; IVC: inferior vena cava; AST: aspartate transaminase; ALT: alanine transaminase; TB: total bilirubin.

development of hypotension, especially when the IVC lumen is patent. The placement of the clamp may demand venovenous bypass associated with a risk of severe bleeding and higher morbidity. If perfusion is inaccessible, the operation must be performed quickly, resulting in non-reconstructive surgery in the form of simple IVC ligation. Controversies exist in the literature regarding whether the IVC should be ligated or restored after resection [2]. It seems reasonable that patients with an open IVC lumen should be managed with IVC reconstruction for preventing legs oedema and reserve physiological left renal venous return [2], especially when a single kidney remains.

A second critical aspect of such cases is a Pringle's manoeuvre for preventing bleeding from the hepatic veins after IVC opening. Although evidence in support of the clinical application of intermittent portal triad clamping has been presented [3], it carries a risk of hepatic ischaemia. Additionally, the necessity to remove the clamp periodically prolongs the operation time, increasing blood loss.

ESSC with circulatory arrest was first introduced by John Lewis in 1952 before the era of the heart-lung machine, when he performed the world's first successful open-heart procedure, using surface cooling to close an atrial septal defect [4]. Initially used in open cardiac surgery, ESSC was later replaced by cardiopulmonary bypass. However, up to the late 1980s, a single cardiac surgical team used surface cooling of the body temperature from 26 to 24°C for the needs of cardiac surgery with time periods of circulatory arrest of 2 h with exclusively low perioperative complications [5–8].

In the case described here, ESSC application eliminated the need for perfusion, and tolerance to systemic arterial hypotension and hepatic ischaemia was observed for more than 1 h after IVC clamping and uninterrupted Pringle's manoeuvre.

During ESSC, a target level of temperature of 29°C was selected on the following theoretical basis. It was recognized that cooling from extreme climatic conditions brings life-threatening consequences if core temperature drops below 28.0°C [9]. Also,

the maximal antihypoxic effectiveness of cooling is achieved at the temperature level below 32°C [9]. It was supposed that the target temperature should lay in the range from 28.0°C to 32.0°C. Since the patients show a gradual trend in temperature growth over time, the target level of 29°C was chosen as optimal but might seem excessive.

Although it was recognized that a risk of clinically significant arrhythmias occurs only if the core temperature drops below 30°C [9, 10], the authors of the above-cited articles reported on patient's cooling to a temperature level of 24–26°C without life-threatening arrhythmias [5–7]. It should be mentioned that the intentional cooling in conditions of general anaesthesia, cardiac rhythm, haemodynamic, respiratory monitoring, acid-base and electrolytes laboratory control, deep muscle relaxation differs substantially from other conditions of unintended cooling. The main cause of the majority of cooling complications (skin necrosis, acidosis, arrhythmias, shivering, etc.) is a pronounced systemic pressor response, which has previously been reported to be associated with peripheral vasoconstriction [11].

Some technical features of ESSC providing its safety should be underlined. The maintenance of sufficient peripheral circulation is an important factor, not only in terms of cooling rate, but also in terms of avoiding ESSC complications [11]. A double dose of non-depolarizing muscle relaxant may facilitate heat loss by preventing thermogenesis from transversal striated muscle fibre contractions, decrease peripheral vascular resistance and minimize metabolic acidosis. A simultaneous intravenous infusion of dopamine and peripheral vascular dilators may also facilitate cooling by increasing the blood volume passing via superficial tissues and provide minimization of vasoconstriction. The patient's head and neck may serve as a highly effective heat exchanger, which may be used continuously for temperature control during operation.

The parameters of controlled ventilation should be maintained, taking into account the fact that as temperature

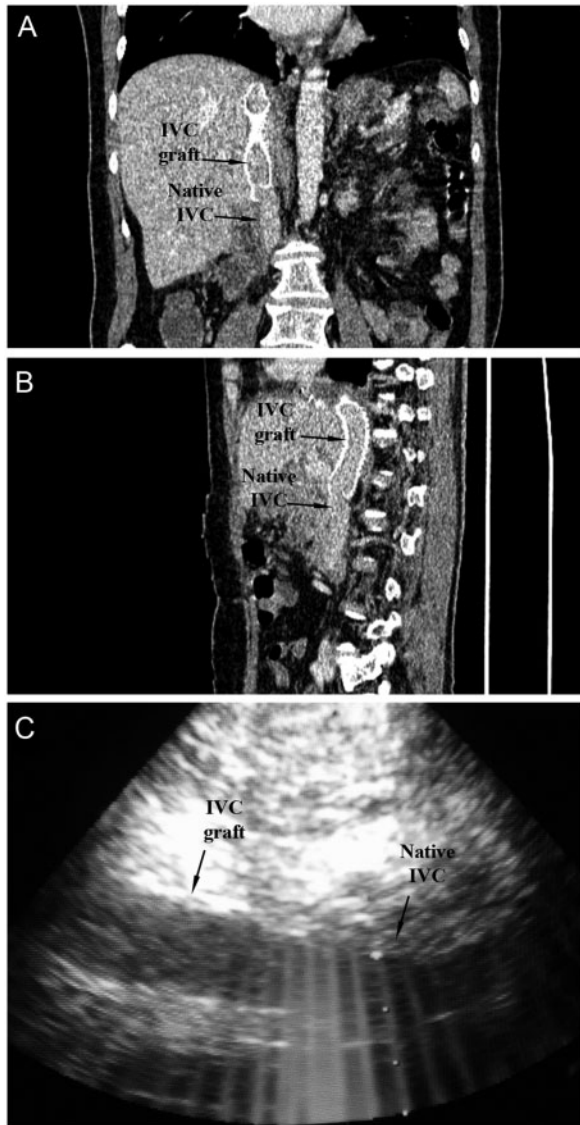


Figure 3: (A) Postoperative coronal computed tomography scan. (B) Postoperative sagittal CT scan. (C) Abdominal ultrasound image of the distal IVC anastomosis. IVC: inferior vena cava.

decreases, the blood solubility of carbon dioxide grows, and reduction in partial pressure of carbon dioxide occurs. As a result, the alkalosis develops with low blood level of potassium giving pro-arrhythmic effect [12]. The importance of respiratory and laboratory monitoring during ESSC should be underlined again. It seems appropriate to be ready to use anti-arrhythmic drug, e.g. amiodaron, for rhythm management.

The target level of anticoagulation in ESSC may also be a subject of discussion. Historically, ESSC with the goal of circulatory arrest for needs of cardiac surgery used deep anticoagulation [5–7]. The clamping of great veins demands less anticoagulation, and the doses of heparin may vary from surgeon to surgeon. The target level of anticoagulation used in this case (activating clotting time over 600 s) was justified to save all possible blood loss during the operation for reinfusion. Some volume of blood loss is unavoidable during this kind of operation. It is highly expedient to save autoblood and to minimize or remove donor transfusion. Bleeding complications resulting from administration of heparin only during an open and controlled surgical

field are not common because of the accessibility of quick heparin's neutralization by protamine.

Although there are no articles comparing ESSC and venovenous bypass in terms of complications rates, in the published literature, it seems that the rate of ESSC complication is minimal [5–7]. In contrast, complications of cardiopulmonary bypass (including fatal ones) have been published independently on surgical or percutaneous insertions of perfusion lines [13–16]. Interestingly, the use of venovenous bypasses instead of cardiopulmonary bypass in cases of tumours involving the IVC were not accompanied by a decreased rate of complications [15].

In conclusion, the application of ESSC in cases of suprahepatic caval and hepatic triad clamping seems to be a useful supplement to the surgical armamentarium. ESSC can easily be performed. It is not costly and may be used in other clinical cases when expected and stipulated systemic arterial hypotension develops.

Conflict of interest: none declared.

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