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Kyoto University
Title page

Title of paper
Acute obstruction of craniocervical venous drainage in a lung-transplanted patient

A list of authors
1. Satoshi Shimizu, M.D.
2. Tomohiro Koyama, M.D. Ph.D.
3. Toshiyuki Mizota, M.D.
4. Kazuhiko Fukuda, M.D. Ph.D.

Institution: Department of Anesthesia, Kyoto University Hospital.
54 Shogoinkawaharacho, Sakyo-ku, Kyoto, 606-8507, Japan

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Corresponding Author:
Satoshi Shimizu, M.D.
Department of Anesthesia, Kyoto University Hospital.
54 Shogoinkawaharacho, Sakyo-ku, Kyoto, 606-8507, Japan
Phone: 81-75-751-3433
FAX: 81-75-752-3259
Email: satosyl@gmail.com

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Summary

Acute obstruction of craniocervical venous drainage is potentially fatal. Our patient presented with severe craniofacial congestion during staged chest closure 4 days after bilateral lung transplantation. Vascular echography revealed a massive thrombosis of the right internal jugular vein and chronic obstruction of the innominate vein, possibly related to a central venous catheter. The severe craniofacial congestion was thought to be induced by acute obstruction of the collateral drainage system. The patient responded to anticoagulation therapy and recovered uneventfully. We discuss the importance of confirming the patency of craniocervical venous return upon central venous catheter insertion.
Central venous catheter (CVC)-related thrombosis is a well-described complication. Although the majority of patients with CVC-related thrombosis remain asymptomatic,¹ acute obstruction of craniocervical drainage is potentially life-threatening.²,³ Here we describe a rare case of a patient who presented with severe craniofacial congestion during staged chest closure 4 days after bilateral lung transplantation. Vascular echography revealed a massive thrombosis of the right internal jugular vein (IJV) and chronic obstruction of the innominate vein, possibly related to the CVC. Here we discuss the potential effect of acute obstruction of the IJV system on clinical outcome and the importance of confirming the patency of craniocervical venous drainage when selecting pulmonary artery catheter (PAC) and CVC insertion sites, especially in patients with a history of CVC placement.

Case report

We obtained written consent from the patient to report this case. A 24-year-old woman underwent emergent bilateral lung transplantation from a brain-dead donor for severe idiopathic pulmonary arterial hypertension refractory to medication. She had received continuous infusion of prostaglandin I₂ via a permanent CVC (Hickman catheter) for 13 years. Due to a CVC-related bloodstream infection, the Hickman catheter was frequently removed and alternately reinserted through the right and left subclavian veins. The catheter in the left subclavian vein had recently occluded and was reinserted into the right subclavian vein. Preoperative assessment revealed dyspnea with Hugh-Jones class IV. A pulmonary hemodynamic test revealed severe pulmonary hypertension with a pulmonary artery pressure of 122/55 mmHg. Results of the pulmonary function tests were within normal range. Arterial blood gas analysis
values were pH 7.436, PaO$_2$ 144.4 mmHg, PaCO$_2$ 44.6 mmHg, and HCO$_3^-$ 29.4 mmol/L with O$_2$ by nasal cannula at 1 L/minute. Chest radiography, electrocardiography, and transthoracic echocardiography indicated severe pressure overload of the right heart.

Standard monitors, including electrocardiogram, pulse oximeter, and arterial catheter were established, and a PAC and CVC were inserted through the right IJV using real-time ultrasound guidance. No thrombus was detected in the right IJV. The left IJV was not scanned with ultrasound. Cardiopulmonary bypass (CPB) was established during the lung transplantation, and the patient was successfully weaned from CPB with use of dopamine, norepinephrine, vasopressin, and inhaled nitric oxide after completion of the lung transplantation. Because of the larger graft size relative to the recipient, staged sternal closure was planned to avoid cardiopulmonary compression, and her chest wall was temporarily closed with a GoreTex patch. Two hours after admission to the intensive care unit (ICU), extracorporeal membranous oxygenation was initiated via the femoral veno-arterial route due to gradually deteriorating oxygenation. Unfractionated heparin was titrated to maintain an activated clotting time of over 150 seconds and an activated partial thromboplastin time of around 45 seconds. The extracorporeal membranous oxygenation support was discontinued on postoperative day (POD) 3 and anticoagulation therapy was withheld, considering the potential risk of increasing postoperative bleeding. On POD 4, the PAC and Hickman catheter were removed, and chest closure with sternal bone fixation wires was planned. The patient remained in a hemodynamically stable condition throughout the chest-closing procedure. Approximately 20 minutes after chest wiring, we observed severe facial swelling and congestion. Color and spectral Doppler ultrasound examination revealed a thrombotic occlusion of the right IJV (Fig.1A) and
the absence of flow within the left IJV (Fig.1B). We speculated that the craniocervical venous drainage was disrupted by a chest closure-induced increase in intrathoracic pressure, and the sternal wires were reopened to release the intrathoracic pressure. Direct inspection and vascular echography revealed chronic occlusion of the innominate vein, where a preoperative chest computed tomography (CT) scan showed severe calcification (Fig.2 A-C). The CVC was removed from the right IJV to prevent further progression of the thrombosis. The patient’s chest wall was closed again with a GoreTex patch and she returned to the ICU. The severe facial congestion was relieved after the chest reopening procedure. The head CT scan was negative for intracranial hemorrhage or cerebral edema associated with a probable increase in intracranial pressure. Anticoagulation with unfractionated heparin was resumed to maintain a target activated partial thromboplastin time of 45 to 60 seconds, followed by oral administration of warfarin (2-3 mg) for 8 weeks to maintain a prothrombin time-international normalized ratio of around 2. Color Doppler ultrasound examination revealed gradual improvement of the venous flow within the right IJV, and her facial swelling subsided during the ensuing days. Flow within the left IJV, however, remained absent. On POD 9, chest closure was uneventfully performed. The patient remained in the ICU until POD 21 and returned home on POD 118. At that time, the thrombus in the right IJV was completely dissolved.

Discussion

CVC-related thrombosis is diagnosed in 33% of patients in the ICU and is occlusive in 3% of the cases.\(^1\) Although CVC-related thrombosis is a relatively common complication, only a few cases of acute thrombotic obstruction of the IJV system have been reported.\(^2,3\) Due to its caliber, the IJV system is the most important
drainage channel in the craniocervical area,\(^4\) and acute obstruction of the IJV system leads to a significantly elevated intracranial pressure and impaired neurologic status.\(^2\) Iroh Tam et al. described a fatal case of acute bilateral IJV thrombosis.\(^3\)

In the present case, the craniocervical venous return was disrupted by both the gradual formation of an occlusive right IJV thrombosis and chronic occlusion of the innominate vein. Chronic obstruction of the innominate vein was assumed to result from the repeated and prolonged central venous catheterization used for continuous administration of prostaglandin I\(_2\) to control the patient’s pulmonary hypertension. Because the patient had not presented with notable craniofacial congestion before the delayed chest closing procedure on POD 4, it is likely that a collateral drainage pathway from the craniocervical area developed during the gradual formation of the occlusive right IJV thrombosis. The sternal closing procedure led to an elevated intrathoracic pressure and consequently compressed the collateral venous drainage, resulting in rapidly progressive craniofacial congestion.

Innominate vein stenosis was previously reported as a late complication of CVC placement.\(^5\) In fact, a preoperative chest CT scan of our patient showed severe calcification of the innominate vein, likely due to frequent placement of a Hickman catheter. Because the innominate vein has an important role in the craniocervical venous return from the left IJV, the patency of the innominate vein should have been carefully verified during the preoperative assessment. Chronic stenosis or obstruction of large veins might be rather common among potential lung transplant recipients with pulmonary arterial hypertension, because continuous infusion of prostaglandin I\(_2\), often via CVC, is strongly recommended as treatment for serious pulmonary arterial hypertension.\(^6\) Patients with vascular catheters placed for dialysis, plasmapheresis, or transjugular intrahepatic portosystemic shunt procedures might also be at risk.
Conkbayir et al. reported the successful use of color Doppler ultrasonography (CDUS) to detect innominate vein occlusion and demonstrate occlusion sites and collateral pathways.\textsuperscript{7} In a patient with innominate vein occlusion, CDUS might be useful for detecting reversed flow in the left IJV as well as engorged veins and some collateral veins in the axilla\textsuperscript{7}. Therefore, evaluation of the bilateral IJV and axillary veins of our patient with CDUS before catheterization could have led to detection of the abnormal flow pattern in the left IJV and suggested innominate vein occlusion.

Obstructive thrombus formation in the right IJV in our patient could be explained on the basis of Virchow’s triad of endothelial damage, altered blood flow, and hypercoagulability. The vessel endothelium was damaged during the PAC and CVC insertion process. Venous flow might have been disrupted in the superior vena cava by the three catheters: the PAC, CVC, and preexisting Hickman catheter. Previous studies have suggested that lung transplant recipients are in a hypercoagulable state.\textsuperscript{8}

In conclusion, we report a rare case of a patient who presented with acute obstruction of the IJV system after bilateral lung transplantation. Our case demonstrates the necessity of confirming the patency of craniocervical drainage with color Doppler sonography prior to selecting PAC and CVC insertion sites, especially in patients with a history of CVC placement.
References


Figure legends

Figure 1: Doppler ultrasonography of the neck approximately 20 minutes after chest wiring.

A. Color Doppler ultrasound examination revealed thrombotic occlusion of the right internal jugular vein (IJV). The mosaic pattern indicated a flow disturbance within the collateral vessels as a result of a chest closure-induced increase in intrathoracic pressure (arrowheads). CCA: Common carotid artery. B. Spectral Doppler ultrasonography showed the absence of flow within the left IJV.

Figure 2: Chest CT images for preoperative assessment.

A, B, C. Preoperative cephalad to caudal chest computed tomography (CT) images demonstrated severe calcification of the innominate vein (arrows). A Hickman catheter was introduced through the right subclavian vein (arrowheads).
Figure 1

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Figure 2

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