Catheter-Related Superior Vena Cava Syndrome Complicated by Chylothorax in a Premature Infant

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A premature infant experienced respiratory distress due to chylothorax. Contrast-enhanced chest computed tomography showed superior vena cava (SVC) obstruction with collaterals, and surgery confirmed an obstruction of the SVC by intravascular fibrotic tissue and thrombi caused by placement of a central venous catheter. The respiratory distress improved after surgical intervention. In the differential diagnosis of acute respiratory distress, it is necessary to consider catheter-related chylothorax and SVC syndrome when a previously stable patient has an acute change in cardiopulmonary status, regardless of the duration of catheter use. Although often considered safe and easy, central venous catheter placement may result in complications. Using appropriate catheters and choosing proper insertion sites can minimize these events. Early diagnosis and treatment can be life-saving should complications occur. *(Chang Gung Med J 2003;26:782-6)*

Key words: **superior vena cava syndrome, chylothorax, central venous catheter.**

Use of central venous catheters (CVCs) is often necessary for treating critically ill patients and those who require total parenteral nutrition. Complications associated with catheter insertion and maintenance, however, occur in as many as 20% of patients with CVCs. Chylothorax, one of the several rare serious complications of CVC placement, occurs as a result of increased thoracic duct pressure, which is secondary to obstruction or increased superior vena cava (SVC) pressure.(1) Chylothorax may lead to high morbidity and may even threaten survival, because of the compromised respiratory space and large losses of lymphocytes, proteins, and immunoglobulins. We present an unusual case of respiratory distress in a 6-month-old premature infant with chylothorax. The chylothorax resulted from SVC obstruction, which was presumably caused by left basilic percutaneous central venous catheter (PCVC) line insertion in the early neonatal period. The chylothorax and SVC syndrome in this baby were both relieved after surgical intervention.

CASE REPORT

This 6-month-old (with a corrected age of 2 months) baby boy was born at a gestational age of 23 weeks and weighed 650 g. His Apgar scores were 3, 5, and 6 at 1, 5, and 10 min after birth, respectively. Respiratory distress was noted in the delivery room, and he was immediately intubated and sent to the neonatal intensive care unit for further treatment.

He had respiratory distress syndrome (grade II) and received conventional mechanical ventilator support without surfactant administration due to subsequent improvement in his respiratory condition. A PCVC (a silicone radiopaque catheter, 30 cm in length, 0.3×0.6 mm in diameter, VYGON GmbH CokG, Germany) was inserted into the left basilic vein on day 7 with the tip located at the junction of the SVC and right atrium to infuse hyperalimentation

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fluid. During hospitalization, apnea of prematurity, a patent ductus arteriosus (PDA), retinopathy of prematurity, and several episodes of infection occurred. Surgical ligation of the PDA was performed via a left thoracotomy on day 26.

Orogastric tube feeding was begun with premature formula on day 10. The left basilic PCVC was removed on day 48 due to leakage of the catheter with concomitant discontinuation of parenteral nutrition. A 14-day course of dexamethasone therapy for treatment of bronchopulmonary dysplasia (BPD) was introduced on day 47. Weaning and extubation were attempted several times but failed. The chest radiography showed early signs of BPD changes.

Frequent apnea, bradycardia, and desaturation were noted from day 77 on, and necessitated a higher level of ventilatory support. Follow-up chest radiography showed patchy infiltration over the right lower lung field on day 80. Respiratory distress persisted, and follow-up radiography revealed pleural effusion on the right side. Pleurocentesis was performed, and a milky fluid was drained out. The fluid composition was 6.7×10^3/l white blood cells (WBCs) (neutrophils: lymphocytes, 1:97), 0.55×10^3/l red blood cells (RBCs), 13.32 mmol/l glucose, 38 g/l total protein, 1.036 mmol/l cholesterol, and 37.5273 mmol/l triglyceride. A chest tube was inserted on the right side on day 81. His enteral feeding was changed to a medium-chain triglyceride (MCT)-predominant semi-elemental formula, (Alfare, Nestle; Laan, 110, 8070 JC, Nunspeet Amsterdam, the Netherlands). He also received a concurrent parenteral nutrition (PN) infusion from the PCVC line, which was inserted into the right femoral vein with the tip at the junction of the inferior vena cava and right atrium. After drainage of the pleural fluid, the shortness of breath and desaturation improved.

Six days later, his respiratory symptoms recurred, with edematous changes of the head and neck. Chest radiography showed pleural effusion on the left side. Another left pleurocentesis was performed, and milky fluid was again drained. The fluid composition of the left side drainage was 4.836×10^3/l WBCs (neutrophils: lymphocytes, 2:89), 0.864×10^3/l RBCs, 6.771 mmol/l glucose, 46 g/l total protein, 1.1137 mmol/l cholesterol, and 6.6783 mmol/l triglyceride. A chest tube was inserted on the left side. For the following days, the 2 chest tubes had daily drainage of 50-150 ml of milky fluid. Culture of the pleural effusion showed no bacterial growth.

Unfortunately, chylothorax with respiratory distress progressed under higher ventilator settings (FiO_2, 70%; rate, 46/min; PIP, 29 cmH_2O; PEEP, 8 cmH_2O). Head and neck swelling also progressed to both upper limbs and the chest wall. From day 86 to 100, his body weight markedly increased (100-150 g/day). Under the impression of SVC syndrome, an echocardiogram was performed, which revealed decreased blood flow in the SVC. Contrast-enhanced chest computed tomography (CT) showed total obstruction of the SVC adjacent to the right atrium, with greatly dilated collaterals which drained the left subclavian vein to the spinal canal; and contrast extravasation from the right subclavian vein due to high pressure of the venous return (Figs. 1, 2).

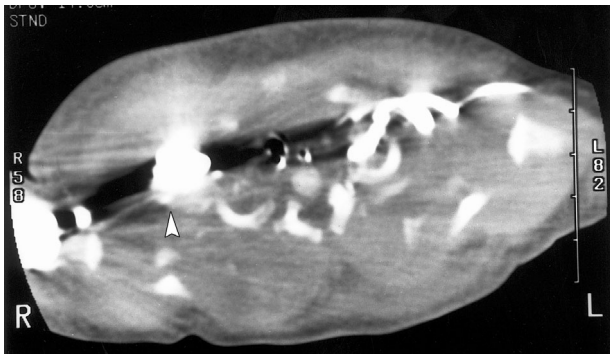

Fig. 1 Contrast-enhanced computed tomography at the level of the thoracic vertebrae (T2 level) showing contrast extravasation (arrowhead) and a dilated left subclavian vein.

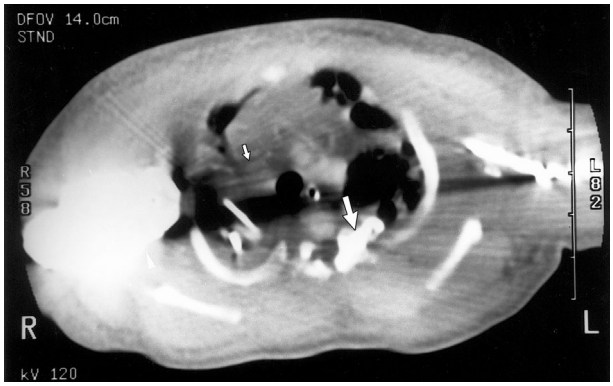

Fig. 2 Contrast-enhanced computed tomography at the level of the thoracic vertebrae (T4 level) showing poor opacification of the superior vena cava (small arrow) and dilated collaterals (large arrow) into the spinal canal over the left side.

Surgical intervention for the SVC syndrome was performed on day 101. Surgical findings included marked (70%-80%) stenosis at the junction of the SVC and right atrium as demonstrated by contrast-enhanced chest CT. Angioplasty with incision of the SVC and pericardial patch closure, and a thrombectomy of the brachiocephalic and innominate veins were performed.

Follow-up chest radiographic examinations revealed no pleural effusion after surgery, but persistent right-side diaphragmatic paralysis was observed. The neck and head swelling disappeared within 2 weeks. Oral feeding was restarted with Alfare on day 129, and PN was discontinued 3 weeks later. At 7 months postnatal age, he was thriving on enteral feeding and required continuous nasal positive airway pressure for ventilatory support. Recurrent head and neck swelling with mild respiratory distress but without significant chylothorax was noted in later infancy. Diuretics were helpful in reversing the recurrent SVC syndrome. There were several episodes of extravasation or leakage of infusate from subsequently inserted central lines, and idiopathic fibrosis of the vascular wall after central venous catheterization was found during the operation. He was discharged at 10 months of postnatal age and has received regular outpatient follow-up.

DISCUSSION

Chylothorax is the most common cause of pleural fluid in a newborn. Beghetti et al.(1) categorized chylothorax into 3 groups: (1) chylothorax which occurs after direct trauma to the thoracic duct (often after cardiovascular surgery); (2) chylothorax which occurs due to increased thoracic duct pressure secondary to obstruction and/or high pressure in the SVC after CVC insertion;[2,3] and (3) congenital chylothorax associated with various syndromes, such as Turner syndrome and Down syndrome. The present case appears to belong to the second group.

From the clinical course and chest CT findings, SVC syndrome due to CVC insertion was the most likely cause. Chylothorax developed at a postnatal age of 80 days, whereas a thoracotomy for PDA ligation was performed in early neonatal life. The chyle initially accumulated on the right side, and thus the earlier operation for PDA was unlikely the cause of the chylothorax. The PCVC catheter was inserted on day 7, with the tip extended to the junction of the SVC and right atrium, thereby allowing PN and drugs to be given. The catheter was removed due to leakage on day 48, and this might have veiled something else such as catheter obstruction from thrombosis or increased SVC pressure at that time. Afterwards, an operation confirmed obstruction of the SVC by fibrotic tissue and thrombi.

SVC obstruction should be considered in a patient presenting with refractory chylothorax. A cardiac echocardiogram can be used to detect thrombi in the great veins and right atrium in such cases. Berman et al. reported that the duration of venous catheterization prior to recognition of the thrombus ranged from 2 to 90 days.[2] Further invasive procedures for the diagnosis of SVC syndrome include phlebography and cardiac catheterization.[1] In contrast to the ordinary course of SVC syndrome reported by Vain et al.,[3] our case ran an unusual course with the chylothorax preceding the ensuing head and neck swelling.

Catheter-related complications can occur in up to 20% of patients with central venous catheter placement.[4] These complications include pneumothorax, cardiac tamponade, sepsis, thromboembolization, hemothorax, hydrothorax, hydromediastinum, and SVC syndrome. The catheter itself and the hyperosmolar fluid can synergistically destroy the vascular wall by mechanical and chemical irritation to the vascular intima,[4] resulting in increased vascular permeability and subsequent inflammation, with development of vascular erosion and perforation,[5] or regeneration leading to fibrotic tissue. Furthermore, catheters in the left subclavian or jugular vein have higher potential to injure vessels due to the right angle of the SVC and the left brachiocephalic vein.[6,7] Catheter displacement due to neck and limb movement,[4] respiration, and cardiac rhythm also increases the incidence of vascular destruction. Although the high flow rate in the central veins should minimize the risk of vascular destruction from hyperosmolar fluid,[7] if the catheter tip is not in the proper position and abuts the vascular wall due to migration,[8] the mixing of infusate is not optimal.

Venous thrombosis is a known complication of CVC with an incidence of 5%-10%.[2] Prolonged catheter use, administration of PN (especially fat and calcium),[2,9] and the nature of the catheter (larger-

diameter, inflexible, polyethylene catheter) are risk factors. A patient with venous occlusion might present with recurrent chylothorax, sepsis, thrombocytopenia, and SVC syndrome. Thrombosis and stenosis of the SVC can be treated with surgery or antithrombotic therapy. Thrombolytic agents have doubtful effects on catheter thrombi,[1,2] and heparin infusion can reduce some local catheter-related problems.[10,11] Shulman et al. suggested that 0.1 N HCl was effective in clearing insolubility-induced precipitation in catheters.[12] Cardiac catheterization for balloon or stent dilatation of vascular stenosis has also been reported.[13,14] In this case, both the SVC syndrome and chylothorax improved after surgical angioplasty.

Use of a flexible CVC of an appropriate gauge and length, and avoidance of left jugular or subclavian vein insertion[6] are recommended to decrease complications. The catheter must be kept in an alignment in the vessel parallel to the long axis of the vein without migration.[7] If the catheter tip is in a suboptimal position, its removal is recommended in order to prevent disastrous complications. Whenever signs of obstruction, infection, or migration of the catheter tip are noted, or parenteral medication or alimentation is discontinued, the catheter should be removed as soon as possible.

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REFERENCES

1. Beghetti M, La Scala G, Belli D, Bugmann P, Kalangos A, Le Coultre C. Etiology and management of pediatric chylothorax. J Pediatr 2000;136:653-8.
2. Berman W Jr, Fripp RR, Yabek SM, Wernly J, Corlew S. Great vein and right atrial thrombosis in critically ill infants and children with central venous lines. Chest 1991;99:963-7.
3. Vain NE, Swarner OW, Cha CC. Neonatal chylothorax: A report and discussion of nine consecutive cases. J Pediatr Surg 1980;15:261-5.
4. Ellis LM, Vogel SB, Copeland EM 3rd. Central venous catheter vascular erosions. Diagnosis and clinical course. Ann Surg 1989;209:475-8.
5. Madhavi P, Jameson R, Robinson MJ. Unilateral pleural effusion complicating central venous catheterization. Arch Dis Child Fetal Neonatal Ed 2000;82:248-9.
6. Ross P Jr, Seashore JH. Bilateral hydrothorax complicating central venous catheterization in a child: case report. J Pediatr Surg 1989;24:263-4.
7. Flatley ME, Schapira RM. Hydropneumomediastinum and bilateral hydropneumothorax as delayed complications of central venous catheterization. Chest 1993;103: 1914-6.
8. Thomas CJ, Butler CS. Delayed pneumothorax and hydrothorax with central venous catheter migration. Anaesthesia 1999;54:987-90.
9. Rubin M, Bilik R, Aserin A, Ziv Y, Sinai Y, Dintsman M. Catheter obstruction: analysis of filter content of total nutrient admixture. JPEN 1989;13:641-3.
10. Nieto-Rodriguez JA, Garcia-Martin MA, Barreda-Hernandez MD, Hervas MJ, Cano-Real O. Heparin and infusion phlebitis: a prospective study. Ann Pharmacother 1992;26:1211-4.
11. Hentschel R. Wiescholek U, von Lengerke J, Harms E, Jorch G. Coagulation-associated complications of indwelling arterial and central venous catheters during heparin prophylaxis- a prospective study. Eur J Pediatr 1999; 158[suppl 3]: S126-9.
12. Shulman RJ, Reed T, Pitre D, Laine L. Use of hydrochloric acid to clear obstructed central venous catheters. JPEN 1988;12:509-10.
13. Adiotomre PN, Burns JE, McIntosh N. Hydrops foetalis and chylothorax associated with superior caval vein obstruction and resolution following balloon dilatation. Acta Paediatr 1994;83:983-5.
14. Ing FF, Mullins CE, Grifka RG, Nihill MR, Fenrich AL, Collins EL, Friedman RA. Stent dilation of superior vena cava and innominate vein obstructions permits transvenous pacing lead implantation. Pacing Clin Electrophysiol 1998;21:1517-30.

早產兒中央靜脈導管使用併發上腔靜脈症候群及乳糜胸

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本文描述一早產兒因患有後天形成之乳糜胸而導致呼吸窘迫。我們從打入顯影劑後的斷層掃瞄中可以發現到上腔靜脈受到阻塞並有多條側枝循環，而從手術中發現到其乃因使用中央靜脈導管而導致血管中產生纖維組織及多處栓塞，因而造成上腔靜脈症候群併乳糜胸。病人呼吸窘迫的症狀於開刀後改善。由此得知，在一個情況穩定的病人身上突然出現呼吸窘迫的症狀，不管其中央靜脈導管使用多久，其所引發的併發症是必須被列入鑑別診斷的。儘管放置中央靜脈導管被認爲是很簡單、很安全的程序，它仍會造成某些併發症。適當的靜脈導管材質及適當的放置位置可減少這些併發症，倘若併發症已發生，及早診斷及治療將會使傷害減至最低。(長庚醫誌 2003;26:782-6)

關鍵字：上腔靜脈症候群，乳糜胸，中央靜脈導管。

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