Case Report

# Advanced Stage T-Cell Non-Hodgkin lymphoma in an 11-Month-Old Infant and Related Superior Vena Cava Syndrome: Importance of Transthoracic Echocardiography

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## Abstract

**Superior vena cava syndrome (SVCS) is rare in infants. Non-Hodgkin lymphoma is the most common cause of SVCS in children. Swelling in the face and neck are the most common clinical symptoms associated with this syndrome. However, these clinical findings are also observed in allergic diseases, which therefore often leads to misdiagnosis. Here, we reported the importance of echocardiography in diagnosing SVCS in an infant with advanced stage non-Hodgkin lymphoma.**

***Keywords:*** *superior vena cava syndrome, non-Hodgkin lymphoma, echocardiography, infant*

## Introduction

Superior vena cava syndrome (SVCS) occurs due to obstruction of the veins returning from the head, upper extremities, thorax, and right atrium. The superior vena cava (SVC) may be blocked by external compression, tumors, lymph nodes, aneurysms, or thrombosis caused by intravenous catheters (1). Non-Hodgkin lymphoma (NHL) is the most common cause of SVCS in children (2,3). The symptoms include facial edema, increased dyspnea in the supine position, coughing, abnormal vision, orthopnea, hoarseness, stridor, nausea, headaches, lightheadedness, and fainting (4).

## Case Report

An 11-month-old girl was admitted to the Pediatric Allergy Unit with facial and neck swelling, which persisted for three weeks. She also had a fever and cough for a few days. The patient was diagnosed with allergic angioedema and referred to our hospital because she did not improve after nine days of treatment with antihistaminic and corticosteroid therapy at another hospital. Upon physical examination, her body temperature was 38 °C, heart rate was 180 per min, and respiration rate was 60 per min. Facial and neck edema were also observed (Figure 1). Sonorous rhoncus was heard in both lungs using auscultation, heart sounds were rhythmic, and 1/6 systolic murmurs were heard on the left side at the 3rd–4th intercostal spaces. White blood cell count was 17000 mL. C-reactive protein (CRP) was positive and blood chemistry was in the normal range. There was no marked parenchymal infiltration

on chest radiography. However, pediatric cardiological evaluation was requested because a clinician suspected SVCS after observing a hyperdense image exceeding the heart contours in the mediastinum and a rightward shift of the mediastinal structures (Figure 2a). An external mass compressing the right atrium and SVC was demonstrated using transthoracic color Doppler echocardiography (Figure 2b). A flow rate of 2.5 $ms^{-1}$ was recorded in the SVC using venous Doppler ultrasound. Chest computed tomography (CT) detected a heterogenic mass of approximately 66 × 56 mm lightly compressing the right atrium and markedly compressing the SVC with hypodense areas compatible with necrosis. No compression of the trachea and principle bronchi was observed. Fluorine-18-fluorodeoxyglucose positron emission tomography was performed for primary staging. Dense hypermetabolic conglomerated lymphadenomegaly consistent with lymphoproliferative malignant pathology was found in supradiaphragmatic and infradiaphragmatic localizations (Figure 3a). Lymph node biopsy revealed T-cell NHL (Figure 3b).

**Figure 1:** Facial and neck edema in an infant.

## Discussion

NHL is the most common primary cause of SVCS (2,3), with SVCS developing in approximately 2% to 4% of NHL cases (5,6). The most common symptoms of presentation include facial and neck swelling, dyspnea, and coughing (4). In our case, facial and neck swelling were first observed by family members three weeks before admission to our clinic. Holme et al. reported a 7-year-old NHL patient with SVC syndrome who was treated for allergies due to neck swelling (7). Similarly, our patient was first misdiagnosed with allergies and was referred to our hospital due to a lack of response to the treatment. Diagnosis should be made quickly in the least invasive manner as SVCS is an oncological emergency. First, chest radiography and CT (if tolerated) images are evaluated. If there is no mediastinal mass image on radiography, transthoracic echocardiography is routinely performed to exclude the intrinsic causes (vascular thrombosis resulting from the intracardiac introduction of a catheter) (8). Ayala K et al. reported that transesophageal echocardiography is useful in the diagnosis of SVC syndrome (9). An infant with

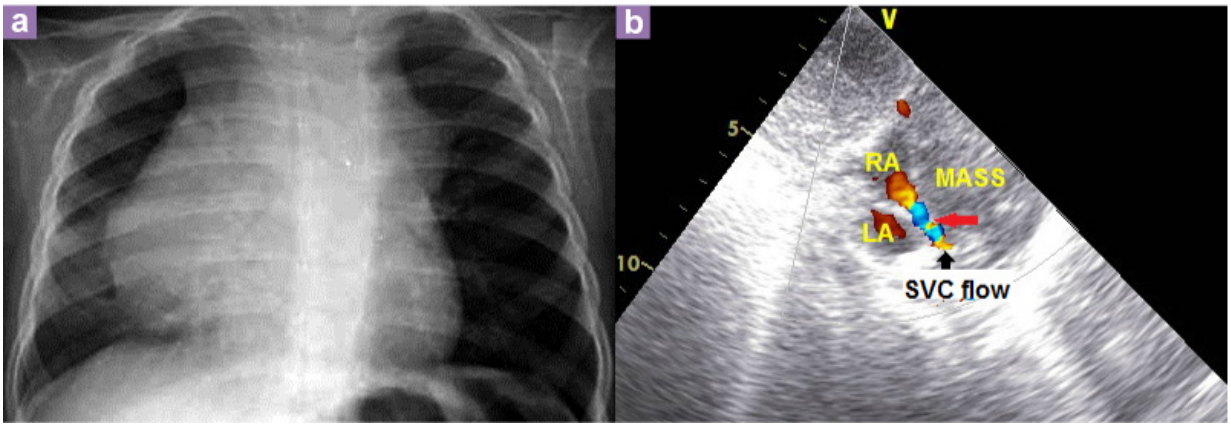


**Figure 2:** (a) Hyperdense image exceeding the heart contours in mediastinum and right shift of mediastinal structures in chest radiography. (b) Image of compression over the right atrium and SVC (red arrow) due to an external mass on transthoracic color Doppler echocardiographic imaging (RA: right atrium, LA: left atrium, SVC: superior vena cava).

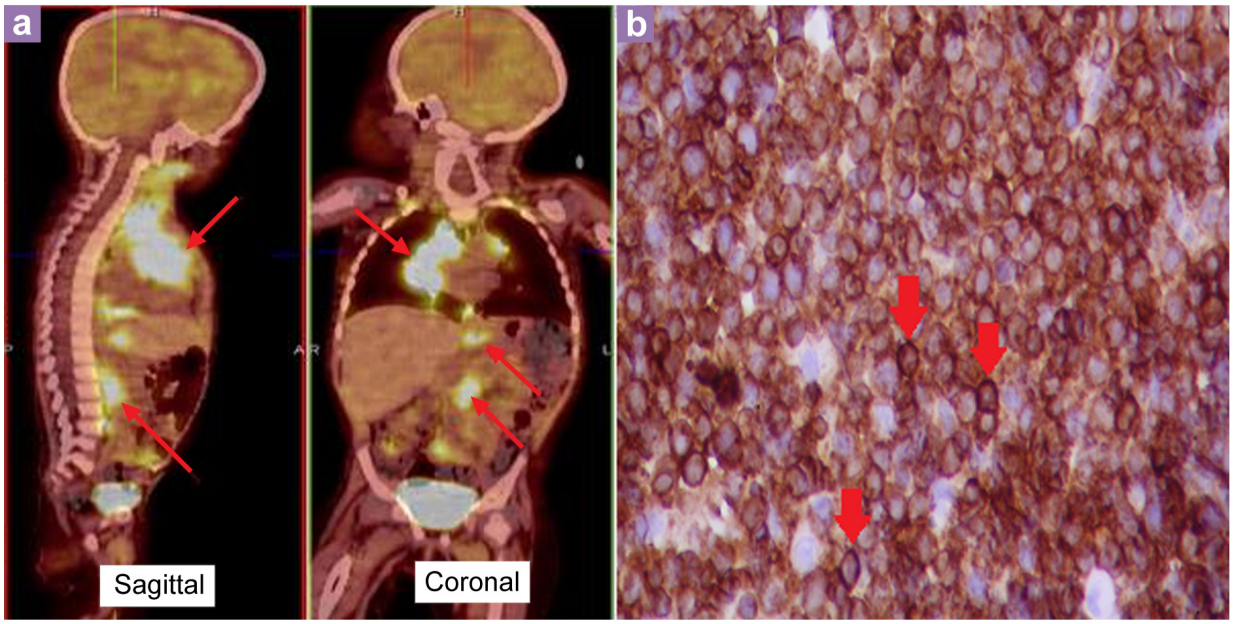


**Figure 3:** (a) Dense hypermetabolic conglomerated lymphadenomegaly consistent with lymphoproliferative malignant pathology was found in supradiaphragmatic and infradiaphragmatic localizations (arrows) in fluorine-18-fluorodeoxyglucose positron emission tomography. (b) Microscopic picture of the lymph node biopsy. CD3 immunohistochemistry staining on the blastic cells (arrows) 40× magnification.

SVCS may not tolerate CT as it is performed in the supine position and requires application of anesthetic. Therefore, when SVCS is detected in the clinic and if there is a mediastinal mass image on radiography, imaging of external compression to the right atrium and SVC with transthoracic echocardiography may help in quickly identifying the etiology and determining the appropriate treatment protocol. As our patient was 11 months old and required sedation prior to the CT scan, imaging of the external mass external compressing the right atrium and SVC with transthoracic echocardiography following direct radiography helped in identifying the etiology and sped up the diagnostic process. The presence of facial and neck edema in an infant may be due to allergies. However, SVCS should also be considered in such cases. Transthoracic echocardiography imaging methods are also crucial in identifying the external compression that may occur over the SVC and right atrium in a noninvasive manner without any additional discomfort as most patients have respiratory insufficiency.

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## Conflict of Interest

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Analysis and interpretation of the data: OY, MC
Drafting of the article and final approval of the article: OY, OK
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