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LETTER TO THE EDITOR
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Management of Venous Thrombosis in a T-Cell Acute Lymphoblastic Leukemia Case with Superior Vena Cava Syndrome

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Superior vena cava syndrome (SVCS) results from occlusion of the superior vena cava (SVC) and results in decreased venous return to the heart from the head and neck region, which accounts for 35% of total venous return. This can cause blood flow obstruction, respiratory distress, and acute and rapid development of neurological signs.[1]

In children, SVCS occurs due to oncological pathology (most often associated with leukemia or lymphoma), intrinsic blockade from thrombosis (most commonly associated with a central venous catheter), or external compression associated with the treatment of congenital heart defects.[2] Due to the relative rarity of this syndrome in children, few retrospective cohort studies are available, most of which have examined single subtypes of SVCS.[3] In addition, the ataxia-telangiectasia (AT) patients are 100 times more likely to develop cancer, and the reported lifetime incidence is between 10% and 40%.[4,5]

An 11-year-old male patient diagnosed with AT was admitted with complaints of sudden respiratory distress and neck swelling. On physical examination, his general condition was poor, and agitation, respiratory distress, and cyanosis were present. In the auscultation of the lungs; basal breath sounds were decreased in the right lung. Right side venous distention of the neck and hepatosplenomegaly were present. Posterior–anterior chest radiography showed a mass lesion of soft tissue density in the mediastinum, filling all its compartments and surrounding vascular structures, trachea, and main bronchus. In addition, there was a pleural effusion in the right lung (Figure 1). Flow oxygen support and prednisolone intravenous therapy were initiated for respiratory distress due to SVCS. In the chest computed tomography (CT), a mass lesion with soft tissue density filling all compartments in the mediastinum and surrounding vascular structures, trachea, and main bronchus was observed. In this mass lesion, the left brachiocephalic vein was interrupted and there was an appearance thought to belong to a contrast-stained tumor thrombus extending to the right subclavian vein and jugular vein in the SVC lumen. Azygos veins and collateral vascular structures in the upper hemithorax and neck were more prominent on the left due to the deterioration of venous drainage due to the compression effect of the mass. There was 47 mm deep pleural effusion on the right, irregularity, and thickening of the posterior leaves of the pleura, a mass lesion extending to the retrocrural area, and T9-10 neural foramen entrances without spinal canal extension in the lower part of the right hemithorax. In the right axilla, 16 × 8 mm diameters lymphadenopathies were observed (Figure 2). Tumor thrombus extending to the right subclavian vein and jugular vein eruption in the SVC lumen was detected (Figure 3). Thrombus was also demonstrated by echocardiography and color Doppler ultrasonography screening of the neck. Simultaneous values of blood count for white blood cell, hemoglobin, and platelet were 22 550 × 10^9/L, 7.8 g/dL, and 22 000 × 10^9/L, respectively.

Bone marrow aspiration (BMA) smear was done for the diagnostic procedure. As a result of flow cytometry and smear evaluation of BMA, the diagnosis of the patient was reported as T-cell acute lymphoblastic leukemia (ALL). Subcutaneous treatment of low molecular weight heparin enoxaparin sodium (Clexan) at a dose of 2 kg/day, together with T-cell ALL therapy,

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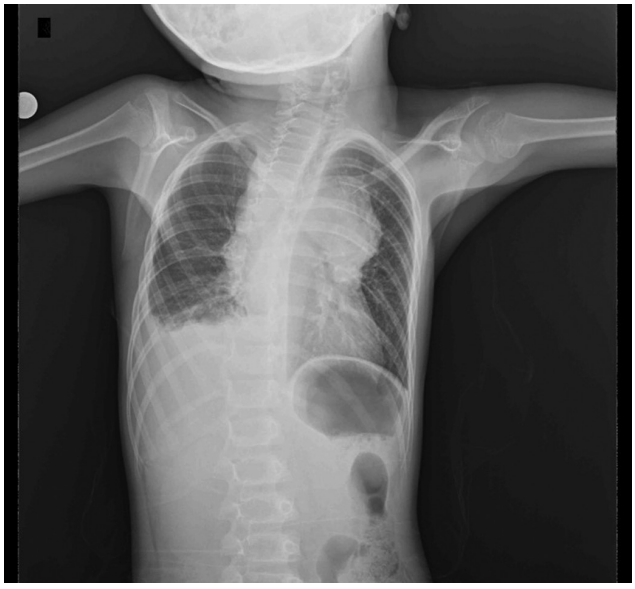

Figure 1. A mass lesion of soft tissue density in the mediastinum filling all its compartments and surrounding vascular structures, trachea, main bronchi, and right lung pleural effusion was observed in the chest radiography in first day of diagnosis.

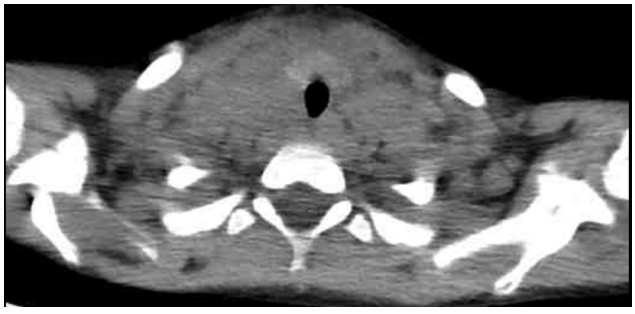

Figure 2. Tumor thrombus extending to the right subclavian vein and jugular vein spillage was detected within the vena cava superior lumen in computed tomography scan.

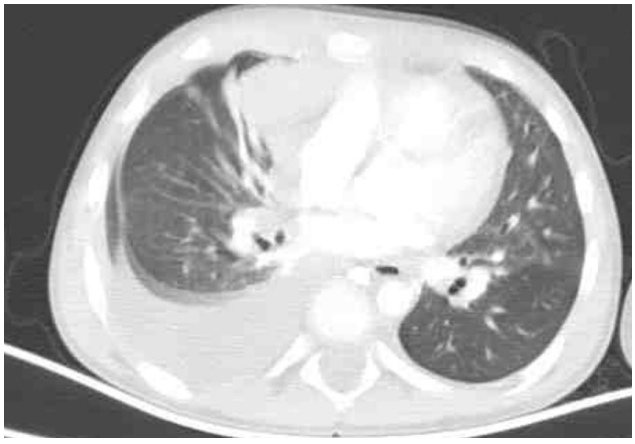

Figure 3. Right pleural effusion and a mass lesion filling all mediastinum and enveloping the trachea and bronchi in thoracic computed tomography scan.

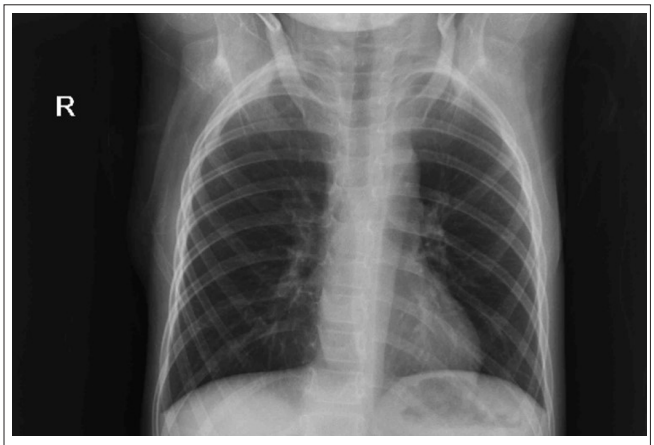

Figure 4. The total healing in the chest radiography after 3 months of treatment.

under the guidance of the ALLIC-BFM 2009 protocol was initiated. After weekly follow-up with echocardiography and Doppler ultrasonography, the resolution of the thrombus was detected on the 28th day of treatment. No additional complications are related to venous thrombosis developed during this period. Anticoagulant therapy was extended to 3 months. After 3 months of treatment, Doppler ultrasonography was performed and it was reported that venous flow continued in the SVC vein and there was no vein thrombosis. In addition, absolute improvement was observed in the chest x-ray (Figure 4). Anticoagulant treatment was discontinued and venous thrombosis was checked every 15 days for up to 3 months. No additional vein thrombosis was observed. Several pediatric-based cohort studies evaluated SVCS in pediatric populations in the literature. In Nosseir and et al study, 101 case reports/case series (142 patients) were analyzed. Morbidity (30%), mortality (18%), and acute complications (55%) were assessed as outcomes. Thrombosis was present in 36% with multi-modal anticoagulation showing improved outcome by >50%.They suggest that infant age , lack of collaterals, acute complications, and clinical presentation may have prognostic utility that could influence clinical decisions and surveillance practices in pediatric SVCS.[6]

In Ozcan and et al's study, 19 (5 were female) of 41 patients with mediastinal tumors had vena cava superior syndrome. Diagnosis included Hodgkin's lymphoma in 7 (37%), non-Hodgkin's lymphoma in 6 (32%), acute T-lymphoblastic leukemia in 4 (21%), neuroblastoma and anaplastic round cell sarcoma in 1 each, respectively. All of the 19 patients' showed facial swelling, venous distention, and mediastinal widening. All patients received intravenous corticosteroids (0.6 mg/kg dexamethasone). Furthermore, the patient with anaplastic round cell sarcoma received emergency radiotherapy. No patients died because of vena cava superior syndrome. In this retrospective study, they found that the most common cause of vena cava superior syndrome was Hodgkin's lymphoma, different from the literature.[7]

Thrombosis of the SVC may occur in the diagnosis of cancer-related SVCS. It is rare for AT to overlap with T-cell ALL and venous thrombosis may develop in these individuals. It may

cause an increase in mortality and morbidity of the disease. In these cases, anticoagulant therapy should be added to the primary disease treatment. In our case, no side effects were observed during enoxaparin subcutaneous injection and chemotherapy additional anticoagulant treatment. Future efforts should focus on establishing multicenter clinical trials to identify the most appropriate interventions needed to improve short- and long-term outcomes for children with SVCS.

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