Superior Mediastinal Syndrome

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Abstract
Superior mediastinal syndrome is a rare condition in the pediatric age group and more common in adults. In children, the most common cause is non-Hodgkin's Lymphoma followed by T cell lymphoblastic leukemia. It is a life-threatening pediatric oncologic emergency requiring immediate diagnostic evaluation and management. We report a case of 6-year-old boy with superior mediastinal syndrome secondary to acute T-cell lymphoblastic leukemia.

***Keywords**: airway; chemotherapy; life threatening; lymphoblastic leukemia; mediastinal; superior vena cava; T cell; venous access*

Introduction
Superior mediastinal syndrome (SMS) is a term that refers to a combination of superior vena cava syndrome (SVCS), which is compression of the superior vena cava with flow obstruction, along with compression of the trachea.[1,2] In children, superior mediastinal syndrome is most commonly caused by non-Hodgkin's lymphoma followed by acute T-cell lymphoblastic leukemia and other malignancies. Less common causes include infections like tuberculosis, histoplasmosis, syphilis and superior vena cava thrombosis as a complication of cardiac surgery or pressure of central line.[1,2] Diagnosis require high clinical suspicion based on thorough history and clinical finding leading to appropriate diagnostic evaluation and timely management.

Case Report

This is a case of a 6-year-old boy who presented at a local hospital with a 2-week history of stridor, difficulty in breathing, dysphagia, and low-grade intermittent fever. Findings on examination included stridor and decreased air entry on the right side of the chest. After initial work-up, the provisional diagnosis was foreign body aspiration. Bronchoscopy was planned; however, at the initiation of the procedure, the patient had an episode of bradycardia and desaturation that required brief CPR, and the procedure was deferred for another time. Subsequent attempt at bronchoscopy resulted to a similar episode; therefore, it was decided to abandon this procedure. The patient was kept intubated and shifted to the Pediatric Intensive Care Unit (PICU) of Sultan Qaboos University Hospital (SQUH) for further evaluation and appropriate management.

The patient was started on mechanical ventilation. On examination, he was critically sick, desaturating in supine position, tachycardic with a heart rate of 140 per minute, and fluctuating blood pressure. There was periorbital edema, facial congestion, neck fullness, upper chest swelling above the level of the clavicle, and cervical lymphadenopathy. On chest exam, there was a right-sided decreased air entry. The rest of the systemic examination was unremarkable. Complete blood count with peripheral smear was normal. Anteroposterior view of the chest showed widened mediastinum involving the paratracheal strip. Chest CT showed anterior and superior mediastinal mass compressing the superior vena cava, narrowing of lumen of the ascending aorta, transverse arch and descending aorta, and obliteration of the main bronchus and its branches. (Figure 1).

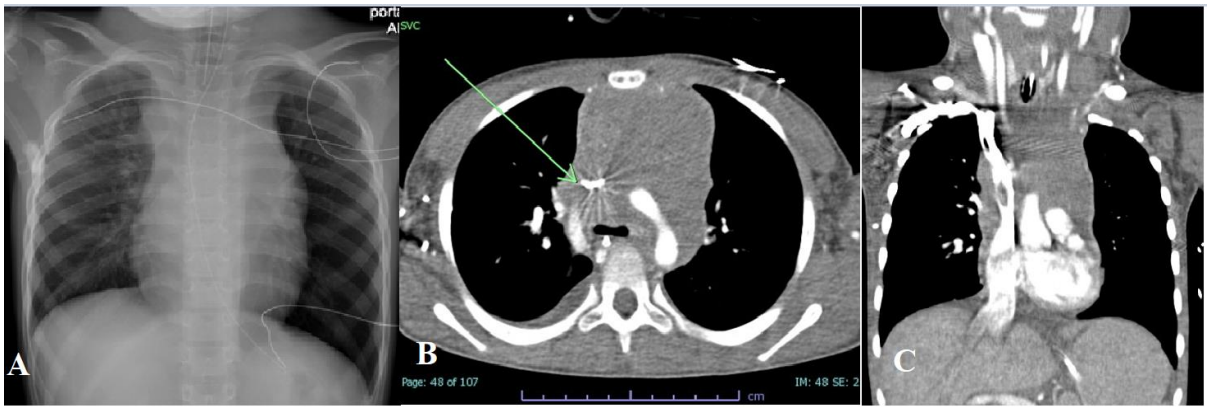

Figure 1: Imaging before treatment
1A. Chest X-ray show large mediastinal mass in the region of superior mediastinum and the patient is intubate
1B. CT show large soft tissue density mass lesion and is resulting in compression of superior vena cava (SVC) and is slit like on this axial image [marked by arrow]
1C. CT coronal reformation redemonstrates large soft tissue density mediastinal mass

Echocardiography was done which showed large soft tissue mass in the upper mediastinum encasing the superior vena cava and ascending aorta causing obstruction to the superior vena cava flow and flow acceleration in the aorta and transverse arch. He was kept intubated and ventilated on moderate setting. Central line was inserted in the right femoral vein and cannulation in upper extremities was avoided. The patient was sedated with midazolam and morphine infusion, avoiding bolus doses and muscle relaxant as it could be hazardous for the patient. He was started on high-dose methylprednisolone as a life-saving medication. Based on history, examination, and initial workup, the most likely diagnosis was superior mediastinal syndrome secondary to lymphoma or leukemia. After taking high-risk informed consent, the patient was taken to operating theater for excisional biopsy; however, due to severe desaturation, the procedure was not done. Based on the result of the bone marrow aspiration and trephine biopsy done in PICU, he was diagnosed as a case of acute T cell lymphoblastic leukemia. He was started on standard chemotherapy protocol. He was strictly monitored for tumor lysis syndrome during the induction phase of chemotherapy.

During the course of management, he showed dramatic improvement in symptoms including facial congestion and puffiness and was successfully extubated on day 8 of admission, and transferred out of PICU to the Hematology/Oncology ward. Repeat chest X-ray was completely normal and follow-up CT scan showed normal caliber superior vena cava, aorta, main bronchus

and its branches. (Figure 2). He was discharged home in a good condition with follow-up in Hematology.

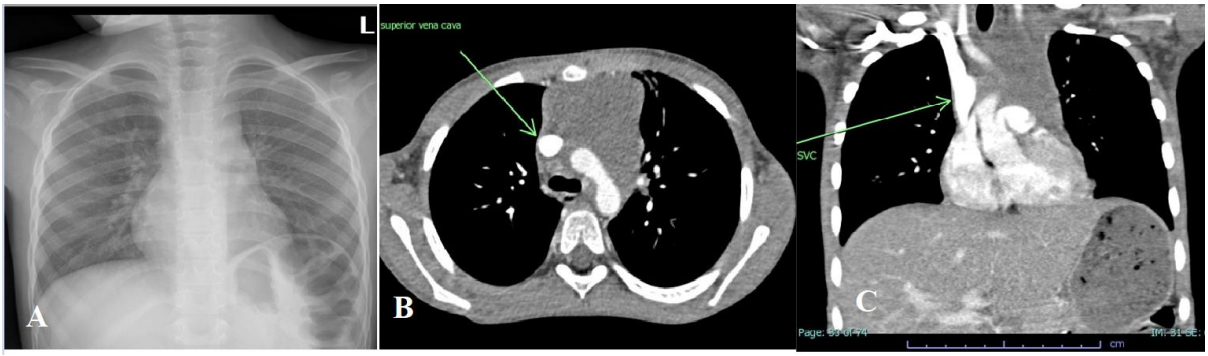

Figure 2: Imaging after treatment
2A. On this Chest X-ray reduced size of the superior mediastinal mass. Now ETT has been removed.
2B. CT show reduction is size of soft tissue density mass lesion in superior mediastinum. There is significant improvement is earlier seen compression of SVC and now show better caliber on this axial image [marked by arrow]
2C. CT coronal reformation show reduction is size of earlier seen mediastinal mass.

Discussion

Superior mediastinal syndrome is a life-threatening pediatric oncologic emergency caused by the coexistence of the signs and symptoms of superior vena cava obstruction and compression of the trachea leading to airway compromise.[1,2] The superior vena cava receives blood from the upper part of the body and accounts for 35% of venous drainage to heart.[2] Signs and symptoms of superior vena cava syndrome caused by its obstruction which impede venous return to the heart include swelling of face and upper body, facial plethora and cyanosis, jugular vein fullness, engorgement of collateral veins, conjunctival suffusions, and low cardiac output. Impeded venous drainage to the brain leads to cerebral edema manifested by visual changes, cognitive disturbances, syncope, seizures, and altered sensorium.[1,2,3] Respiratory symptoms include cough, dyspnea, orthopnea, stridor, and hoarseness.[1,2] It can also cause dysphagia secondary to compression of the esophagus.

The patient initially presented to a peripheral hospital with classical symptoms of superior mediastinal syndrome; however, misled with provisional diagnosis of airway foreign body aspiration and was taken for bronchoscopy twice without consideration of other possibilities and involvement of concerned subspecialty. In such cases, it is very important to consider multidisciplinary approach and subspecialty consults prior to proceeding for invasive procedures like bronchoscopy, as it can be life threatening and with devastated outcome.

Diagnosis is based on classical clinical finding while laboratory tests are helpful in identifying the underlying etiology. Diagnostic workup include complete blood counts (CBC) with peripheral smear, flow-cytometry, tumor markers, bone marrow biopsy, lymph node biopsy, imaging studies which includes chest radiograph, computer tomography (CT) or magnetic resonance imaging (MRI) and echocardiography.[2,4] During sedation, it is important to remember that anesthesia and particularly muscle relaxant can be life threatening and may lead to rapid airway compression that may be severe enough that endotracheal intubation may fail

to relieve obstruction. In addition, superior vena cava occlusion and pulmonary artery compression may lead to cardiovascular collapse with substantial risk for death. The diagnostic workup should be tailored according to clinical scenario and best performed under local anesthesia wherever possible. If necessary, use intravenous anesthetic agent without muscle relaxant to prevent sudden airway obstruction, the most common and feared complication. [5,6,7,8]

The goals of therapy are to relieve symptoms and treatment of underlying etiology. Emergency management is of crucial importance including proper positioning, elevation of the head and neck at a 45-degree angle, securing airway, intubation and mechanical ventilation if needed, supplemental oxygen, corticosteroid for debulking of mediastinal mass, and monitoring for tumor lysis syndrome.[4,9] Avoid venous access in the upper extremities as it will attribute to the augmentation of SVC obstruction. Venous access should be secured in the lower extremities to obviate this complication.[10] In this patient, venous access was established in the right femoral vein of the patient.

In our patient, there was a clinical observation of absence of peripheral blast cell with presence of superior mediastinal syndrome; however, these findings should not stop the clinician from doing further investigations to delineate etiology. Bone marrow aspiration and trephine biopsy were done and showed a T cell lymphoblastic leukemia after histopathological confirmation; therefore, chemotherapy was initiated which yielded an excellent outcome. As previously emphasized, definitive treatment of the underlying etiology is of paramount significance.

Conclusion

Superior mediastinal syndrome is an acute life-threatening emergency that requires prompt identification and timely management. High index of suspicion for underlying etiology is of prime importance. The most common cause identified in the pediatric age group is hematological malignancy. Prognosis depend upon the underlying disease and response to treatment, and is usually excellent for chemo-sensitive tumors, as in the featured case.

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