Superior Vena cava Syndrome in a Child with Mediastinal Mass, a Case Report

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Abstract

Facial puffiness as a consequence of superior vena cava syndrome (SVCS), can be a presentation from which the physician would have to conjecture a diagnosis of a mediastinal mass including lymphomas. Pediatric SVCS can rapidly progress and pose a greater challenge for airway protection as was in our case.

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Keywords

Superior Vena Cava Syndrome, Mediastinal mass, Pediatric lymphomas

Abstract

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Key Clinical Message

Pediatric lymphomas can rapidly progress and present with superior vena cava syndrome which needs timely diagnosis and treatment for better clinical outcomes.

Introduction

Superior Vena cava Syndrome (SVCS) results from the compression of Superior Vena cava (SVC) which drains the head, neck, upper limbs and upper park of the chest and torso, including the viscera above the diaphragm. Malignancy, hematological causes (inc. thrombosis), and congenital heart diseases are the majority etiological factors in the pediatric population (1). The former two being of similar etiology to their adult counterparts(2). Increasing use of intravascular devices such as catheters and pacemakers has increased the risk of thrombosis as a cause(2). Lymphoma (most commonly non-Hodgkin lymphoma (NHL)) remains the commonest of the oncological etiologies(1, 3-5). The median age of presentation was 4.75 years with bimodal distribution; first peak at infancy and later in adolescence(1). Here we present a case of an 8 year old girl who presented with superior vena cava syndrome secondary to a mediastinal mass.

Case Presentation

A six year old child presented to our outpatient department with complaints of facial puffiness and feeling of lethargy. She additionally reported shortness of breath and cough for the last 10 days. On examination, there was a marked facial edema, and prominent veins were detected on her upper chest (figures 1 and 2). Breath sounds were decreased on the right supra mammary and mammary areas, but the heart sounds were normal. There was no distention of her abdomen nor presence of edema in her lower limbs. Her oxygen saturation was also recorded to be normal. On further inquiry, it was found that she was managed as a case of nephrotic syndrome for a month before she was referred to our center. However, the report of a urine analysis in our center was normal and without proteinuria. Suspecting a diagnosis of a superior vena cava syndrome, a chest X-ray was ordered which showed a widened mediastinum (figure 3). A diagnosis of a mediastinal pathology compressing the Superior vena cava was made and a prompt referral to a tertiary care center was done. Her steroids which she had been prescribed as a part of management for suspected nephrotic syndrome were also discontinued.

By the time she reached the tertiary care center, her shortness of breath had increased (respiratory rate: 34) and her blood pressure had decreased. After an initial fluid resuscitation and stabilization she was admitted for further investigations and management. Her laboratory results showed a hemoglobin level of 11.6 gm% and an LDH level of 1580 U/L. A subsequent Computed Tomography scan of her chest, abdomen and pelvis revealed the presence of a large mediastinal mass with infiltration into the right pleura with encasement of mediastinal vessels with multiple renal space occupying lesions and peripancreatic nodules suggestive of a diagnosis of lymphoma. Additionally, moderate right-sided pleural effusion and partial collapse of the right middle lobe and the right upper lobe along with a mild pericardial effusion were also seen. Given the urgency of the situation with risk of imminent respiratory collapse, chemotherapy with cyclophosphamide, vincristine and prednisolone was started without a definitive diagnosis via. a biopsy. An X-ray after 8 days of the initial chemotherapy regimen showed a striking shrinkage in her mediastinal mass (figure 4). This shrinkage did not allow a biopsy to be taken from the mass itself. However, a bone marrow aspiration and biopsy was done which reported as negative for malignancy. She showed marked improvement in her symptoms and was finally discharged after 2 weeks of hospitalization. She was asked to come for a monthly follow up to complete her 4 cycles of chemotherapy.

Discussion

SVCS causes obstruction of blood flow leading to increased venous pressure along its tributaries. It can be of devastating consequence if associated with airway edema(6). Children are more susceptible than their adult counterparts due to narrower lumen, greater compressibility of upper airway, and greater edema at onset(7). Management of airways in such a case presents a challenge, as even endotracheal intubation may not guarantee ventilation(8-10). In addition to this, use of sedatives and anesthetics during the process may decrease the pharyngeal tone, which further compromises the upper airway(9). One should be aware of such

a consequence to avoid a fatal catastrophe. Similarly, distal airway compression can also lead to absorption atelectasis leading to collapse of a segment of the lungs, as was in our case.

In the case of lymphoma, a tissue diagnosis is required to characterize the tumor and define the optimal treatment. For this, a core needle biopsy is usually done. Multiple attempts may be needed to define specific characteristics based on architectural and immuno-histochemical evaluation(11). Cervical mediastinoscopy and anterior mediastinoscopy may increase diagnostic sensitivity but carries the added risks of bleeding and airway compromise(12). A pathological diagnosis could not be made in our case due to the urgency of the situation, but whenever possible, it is desirable to obtain an adequate tissue specimen to formulate appropriate and effective therapy. It is also important to note that children with SVCS often tolerate the necessary procedures poorly(13). Because of the risks of anesthesia in a patient with airway compromise and embarrassed venous return, as exemplified in our case, empiric treatment may be necessary before a definitive diagnosis is established, to avoid hemodynamic and ventilator destabilization(14). The response to chemotherapy was striking which further supported our working diagnosis. LDH which is commonly elevated in lympho-proliferative disorders has prognostic significance and can also be used to monitor treatment response and recurrence of the disease (15, 16). If the mass were to reoccur, a consensus was made to take a biopsy at that time in the future.

Both hodgkins lymphoma and non hodgkins lymphoma have excellent 5 year survival of 89.1% and 73.8% respectively(17, 18). Treatment should be approached as a near curable disease. Literature suggests that in cases of SVCS caused by lymphoma, chemotherapy can be as effective as radiotherapy(13). In accordance with this, our patient received Cyclophosphamide, Prednisolone and Vincristine with rapid clinical improvement. Radiotherapy was not used in our case.

Conclusion

In summary, a diagnosis of SVC requires vigilance on the part of the physician, as symptoms may be as trivial as facial swelling alone. It demands high clinical suspicion and conscientious examination. Malignancy, i.e., most commonly NHL is the commonest of the causes outside infancy. Emergency resuscitation and securing the airway in itself poses a challenge to reduce unfortunate outcomes of a near-curable pathology. Chemotherapy can lead to rapid resolution of symptoms. Hence, further emphasizing the need for early diagnosis and treatment for better long term outcome.

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Conflict of Interest

Authors' have no conflict of interest to declare

Authors Contributions

Ashes Rijal: wrote the original manuscript, reviewed and edited the manuscript

Anish Kumar Shrestha: wrote the original manuscript, reviewed and edited the manuscript

Sharmila Chaudhary: obtained the information, reviewed and edited the manuscript

Anisha Shrestha: reviewed and edited the manuscript

Data availability statement

All the required information is available in the manuscript itself.

Ethical consent statement

Written informed consent was obtained from the patient for publication of case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Figures

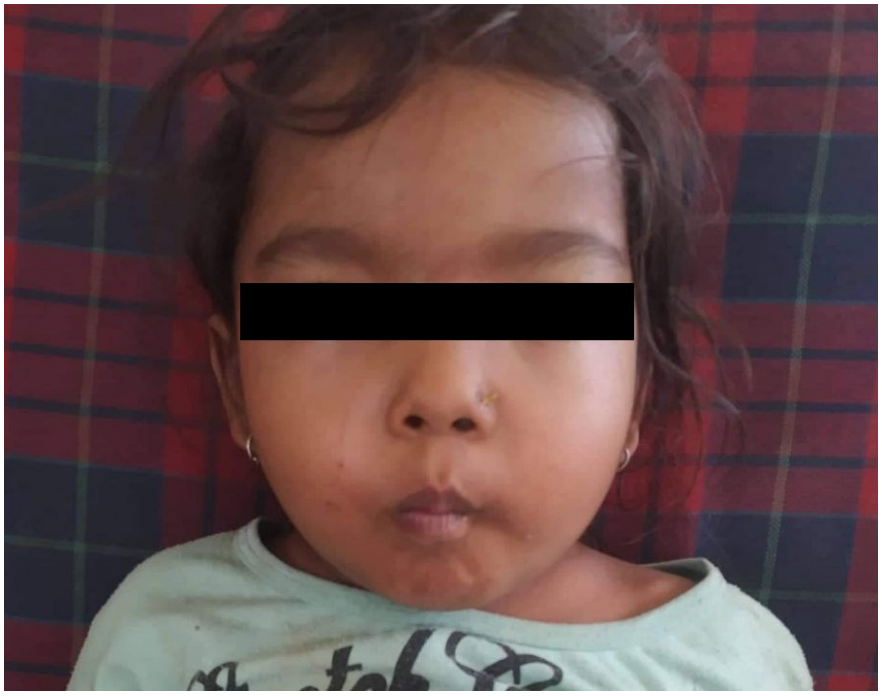

Figure 1 Photo showing prominent facial puffiness and edema

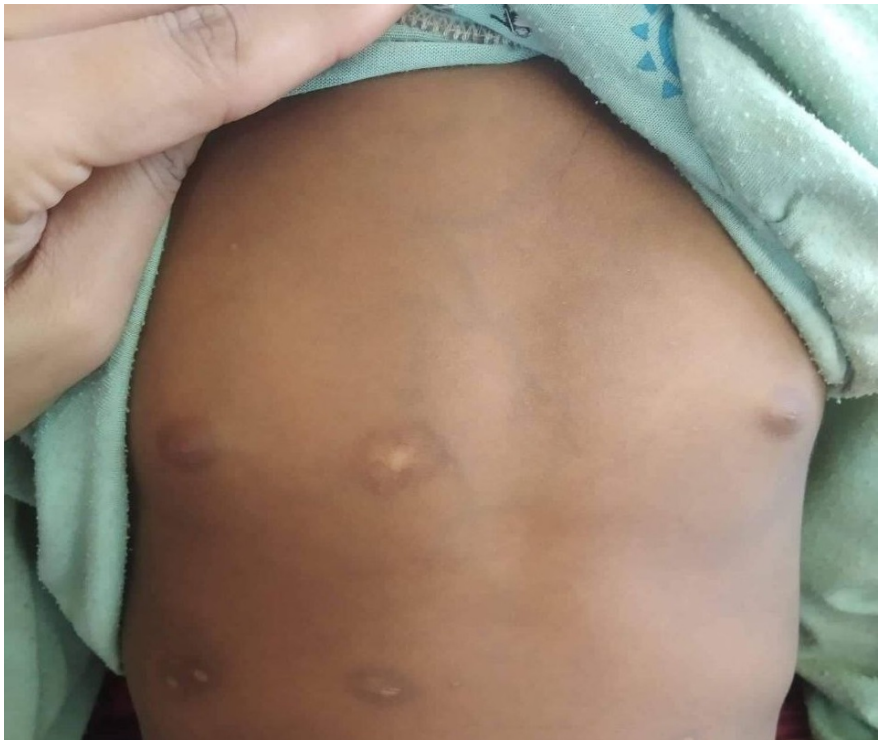

Figure 2 Photo showing prominent chest veins

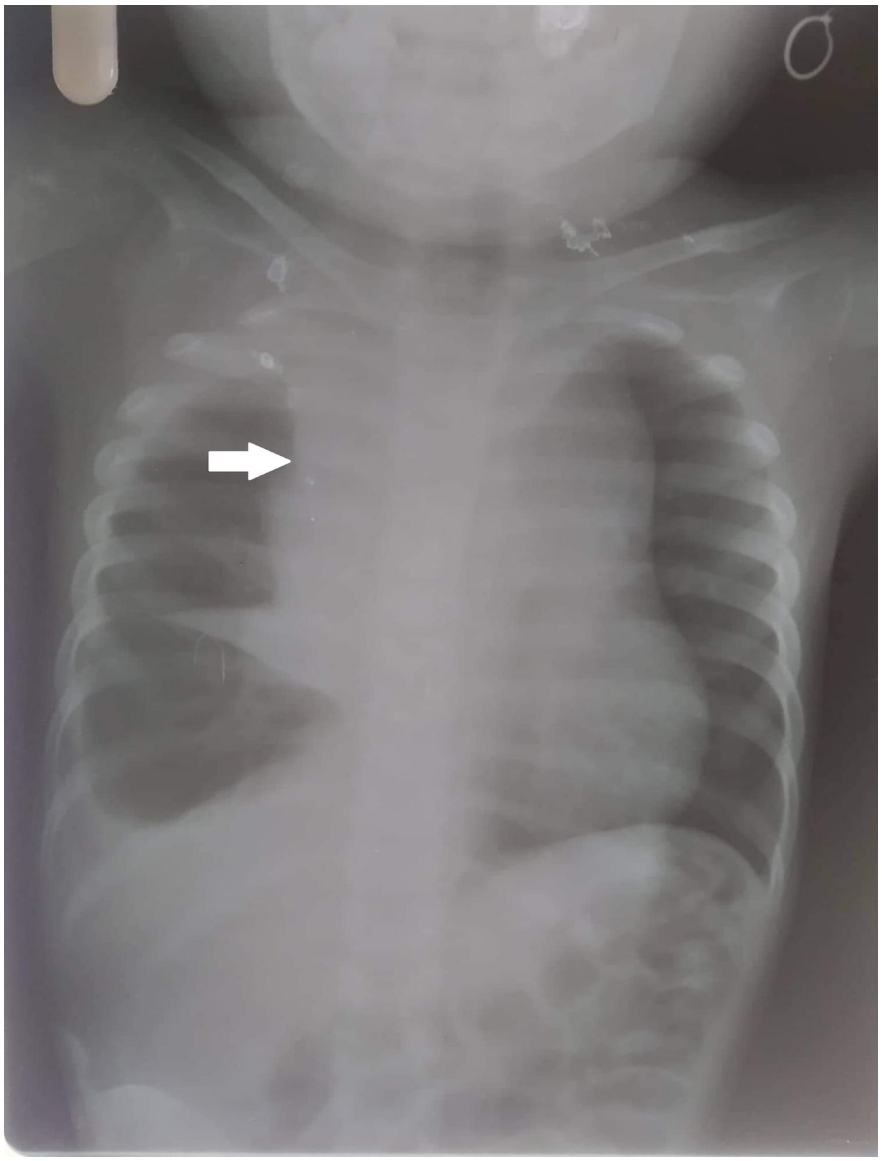

Figure 3 Initial X ray showing widened mediastinum (solid white arrow)

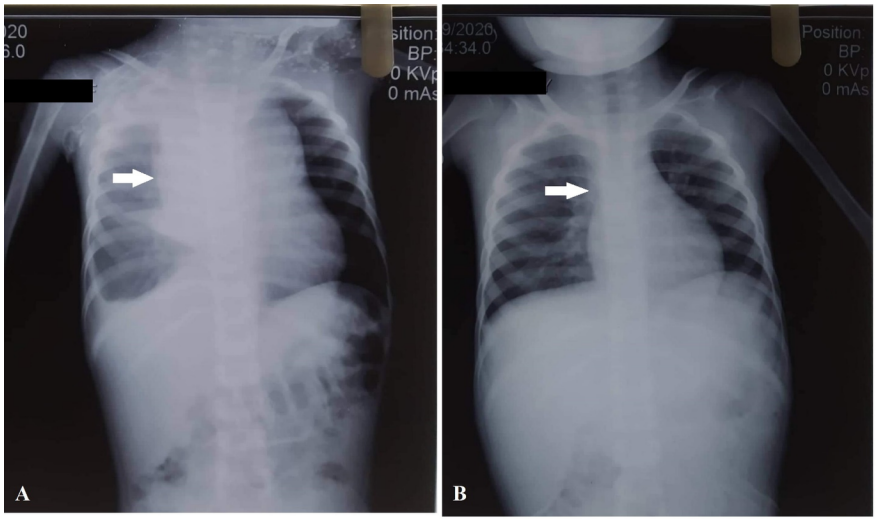

Figure 4 Change in the mediastinal shadow (solid white arrows) following chemotherapy as shown in the two X rays taken 11 days apart (A: before chemotherpay. B: after chemotherapy).