INTRODUCTION

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

3 | DISCUSSION

SVCS obstructs blood flow leading to increased venous pressure along its tributaries. It can be of devastating consequence if associated with airway edema. Children are more susceptible than their adult counterparts due to narrower lumen, greater compressibility of the upper airway, and greater edema at onset. Management of airways in such a case presents a challenge, as even endotracheal intubation may not guarantee ventilation. In addition to this, the use of sedatives and anesthetics during the process may decrease the pharyngeal tone, which further compromises the upper airway. 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 the 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 immunohistochemical evaluation. Cervical mediastinoscopy and anterior mediastinoscopy may increase diagnostic
sensitivity but carries the added risks of bleeding and airway compromise. 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. 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. The response to chemotherapy was striking which further supported our working diagnosis. LDH which is commonly elevated in lymphoproliferative disorders has prognostic significance and can also be used to monitor treatment response and recurrence of the disease. 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 an excellent 5-year survival of 89.1% and 73.8%, respectively. 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. 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, that is, NHL is the commonest of the causes outside infancy. Emergency resuscitation and securing the airway in itself pose a challenge to reducing unfortunate outcomes of a near-curable pathology. Chemotherapy can lead to rapid resolution of symptoms. Hence, this further emphasizes the need for early diagnosis and treatment for better long-term outcome.

AUTHOR 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.

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CONFLICT OF INTEREST

The authors have no conflict of interest to declare.

DATA AVAILABILITY STATEMENT

All the required information is available in the manuscript itself.

CONSENT

Written informed consent was obtained from the patient for publication of the 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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REFERENCES

1. Nossair F, Schoettler P, Starr J, et al. Pediatric superior vena cava syndrome: an evidence-based systematic review of the literature. Pediatr Blood Cancer. 2018;65(9):e27225.

2. Wilson LD, Detterbeck FC, Yahalom J. Clinical practice. Superior vena cava syndrome with malignant causes. N Engl J Med. 2007;356(18):1862-1869.

3. Gupta V, Ambati SR, Pant P, Bhatia B. Superior vena cava syndrome in children. Indian J Hematol Blood Transfus. 2008;24(1):28-30.

4. Jeng MJ, Chang TK, Hwang B. Superior vena cava syndrome in children with malignancy: analysis of seven cases. Zhonghua Yi Xue Za Zhi (Taipei). 1992;50(3):214-218.

5. Ingrum L, Rivera GK, Shapiro DN. Superior vena cava syndrome associated with childhood malignancy: analysis of 24 cases. Med Pediatr Oncol. 1990;18(6):476-481.

6. Hon KL, Leung A, Chik KW, Chu CW, Cheung KL, Fok TF. Critical airway obstruction, superior vena cava syndrome, and spontaneous cardiac arrest in a child with acute leukemia. Pediatr Emerg Care. 2005;21(12):844-846.

7. King RM, Telander RL, Smithson WA, Banks PM, Han MT. Primary mediastinal tumors in children. J Pediatr Surg. 1982;17(5):512-520.

8. Northrip DR, Bohman BK, Tsueda K. Total airway occlusion and superior vena cava syndrome in a child with an anterior mediastinal tumor. Anesth Analg. 1986;65(10):1079-1082.

9. Lin SH, Su NY, Hseu SS, et al. Anesthetic managements of the patients with giant mediastinal tumors--a report of two cases. Acta Anaesthesiol Sin. 1999;37(3):133-139.

10. Maxwell SK, Mizubuti GB, McMullen M, Heffernan P, Duggan S. A tale of 2 tubes for emergency management of airway obstruction from an anterior mediastinal mass: a case report. A A Pract. 2020;14(10):e01257.

11. Carter BW, Marom EM, Detterbeck FC. Approaching the patient with an anterior mediastinal mass: a guide for clinicians. J Thorac Oncol. 2014;9(9 suppl 2):S102-S109.

12. Dosi T, Theakos N, Chatziantoniou C. Cervical mediastinoscopy and anterior mediastinotomy in superior vena cava obstruction. Chest. 2005;128(3):1551-1556.

13. Lokich JJ, Goodman R. Superior vena cava syndrome: clinical management. JAMA. 1975;231(1):58-61.

14. Piastra M, Ruggiero A, Caresta E, et al. Life-threatening presentation of mediastinal neoplasms: report on 7 consecutive pediatric patients. Am J Emerg Med. 2005;23(1):76-82.

15. Yadav C, Ahmad A, D’Souza B, et al. Serum lactate dehydrogenase in non-Hodgkin’s lymphoma: a prognostic indicator. Indian J Clin Biochem. 2016;31(2):240-242.

16. Endrizzi L, Fiorentino MV, Salvagno L, Segati R, Pappagallo GL, Fosser V. Serum lactate dehydrogenase (LDH) as a prognostic index for non-Hodgkin’s lymphoma. Eur J Cancer Clin Oncol. 1982;18(10):945-949.

17. Cancer Stat Facts: Non-Hodgkin lymphoma. 2022. Accessed 15 July, 2022. https://seer.cancer.gov/statfacts/html/nhl.html

18. Cancer Stat Facts: Hodgkin lymphoma 2022. Accessed 15 July, 2022. https://seer.cancer.gov/statfacts/html/hodg.html

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