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Role of EBUS in lymphoma presenting as superior vena cava syndrome: bronchoscopic and sonographic findings: a case report

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Abstract

Background The clinical description of superior vena cava syndrome has been widely studied; however, there is limited information on bronchoscopic findings in clinical practice.

Case presentation A 57-year-old man presented with facial and neck swelling and pedal edema of 6 months duration. Computed tomography showed mediastinal lesions in the right paratracheal stations with thrombosis of the right internal jugular vein and superior vena cava (SVC). Without establishing a diagnosis, he was started on oral steroids elsewhere and his symptoms progressed. He did not receive anticoagulation therapy. Bronchoscopy showed edematous supraglottic and glottic regions with hyperemia of the airway mucosa. Endobronchial Ultrasonography revealed a mediastinal mass of heterogeneous echotexture in the lower right paratracheal region, with mediastinal collateral blood vessels. He underwent EBUS-guided aspiration cytology and intranodal forceps biopsy, which confirmed the diagnosis of non-Hodgkin's.

Conclusion In cases with an unconfirmed diagnosis of lymphoma, it is prudent to refrain from administering glucocorticoids, as these medications can exhibit lympholytic properties and may hinder the diagnostic process. Due to extensive collateral formation in superior vena cava syndrome, utilizing Doppler during endobronchial ultrasound transbronchial needle aspiration (EBUS TBNA) can identify numerous mediastinal collateral vessels, thus minimizing the risk of bleeding.

Keywords Superior vena cava syndrome, Bronchoscopy, Lymphoma

Background

The obstruction of the superior vena cava can result in SVC syndrome (SVCS), which can lead to significant health issues and even death. Malignancy accounts for around 70% of cases of SVC syndrome, making it the most frequent cause. Clinically, it can manifest as headache, stridor, dyspnea, orthopnea, and swelling of the face, neck, and upper limbs [1, 2]. This case report describes SVC syndrome in a patient with non-Hodgkin's lymphoma whose presentation with its unique bronchoscopic and EBUS pictures.

Case presentation

A 57-year-old man presented to our department with facial and arm swelling of 6 months duration. He had been evaluated elsewhere for these complaints 6 months prior and was found to have an anterior mediastinal mass with enlarged mediastinal lymph nodes. He was prescribed oral dexamethasone 10 mg once daily, which he continued to take. He was not given anticoagulants at that time. Over the last 2 months, he developed a cough

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and shortness of breath, prompting a referral to our hospital.

Clinical examination revealed facial puffiness, diffuse neck swelling, distended neck veins, and swelling of the right arm. There were no peripheral lymph nodes. Routine blood investigations were normal, and chest radiography showed a right para-hilar opacity. Computed tomography (CT) of the neck and thorax with contrast revealed a mediastinal mass in the right paratracheal region, circumscribing the superior vena cava (SVC) and measuring up to 31 mm. There was thrombosis of the SVC extending into the right internal jugular vein, causing distension of the lumen. The presence of multiple mediastinal and chest wall collaterals suggested a subacute to chronic onset of SVC thrombosis (Fig. 1).

Compared to scans taken 6 months earlier, there was a reduction in the size of the mediastinal lesion. However, there was an increase in the extent of SVC thrombosis, with the interval development of mediastinal and chest wall collaterals.

A multidisciplinary team (MDT) discussion was held, which suggested that endobronchial ultrasound (EBUS) and transbronchial needle aspiration (TBNA) would be the preferred methods for obtaining histology. The patient was initially started on low molecular weight heparin, which was stopped one day before the procedure. Bronchoscopy and endobronchial ultrasound (EBUS) were performed under spontaneous breathing with conscious sedation using intravenous midazolam and fentanyl. Oxygen was supplemented via nasal prongs. The lesion, as seen by EBUS, measured 3.5 cm.

TBNA was conducted using a 22-G needle under EBUS guidance until rapid on-site evaluation (ROSE) confirmed an adequate sample. Intra-nodal biopsies were taken 4–5 times under real-time guidance with 1.4 mm forceps. Bronchoscopy revealed an edematous supraglottic and glottic region. The tracheobronchial mucosa was hyperemic with extensive mucosal vascularity (Fig. 2).

Endobronchial ultrasound revealed a mediastinal mass of heterogeneous echotexture in the lower right paratracheal region, with increased vascularity at the periphery of some lesions, suggesting mediastinal collateral vessels (Fig. 3). Five samples were obtained. Since ROSE indicated atypical lymphoid cells, we required more tissue for immunohistochemistry to accurately classify the lymphoma. There was minimal bleeding during the procedure. Post-procedure period was uneventful. The patient was initially started on enoxaparin, which was stopped one day prior to the procedure. It was restarted 12 h after the procedure, once it was confirmed that there was no bleeding.

Discussion

Historically, SVCS has often been managed by administering immediate radiation therapy without considering a specific tissue type diagnosis. Ahmann's analysis in1986 showed that diagnostic procedures can be performed safely and that histological diagnosis should always be performed before starting treatment [3]. Conditions such as lymphoproliferative disorders can greatly benefit from upfront combination chemotherapy. However, obtaining a histological diagnosis after radiation therapy can be challenging. The recommended course of action for dealing with SVCS is to provide symptomatic relief and address the underlying cause through the use of treatments including radiation and chemotherapy [3].

The presentation of SVC narrowing can vary based on the rate and severity, which may be acute, subacute, or chronic. Acute cases typically occur within a few days and have less prominent collateral formation than subacute and chronic cases, resulting in more severe symptoms. In contrast, the subacute and chronic cases

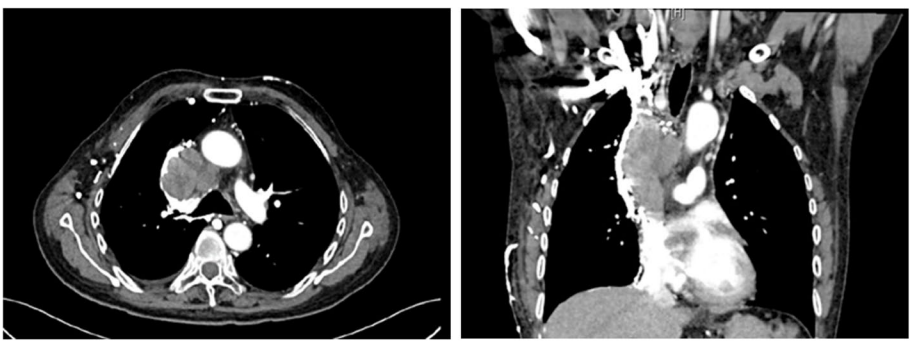

Fig. 1 Contrast-enhanced computer tomography images of the thorax in the axial (**A**) and coronal (**B**) planes. **A** The scan shows a mediastinal mass in the right paratracheal station. **B** The coronal view of the superior vena cava shows a severely narrowed lumen, more by the thrombus within. Multiple mediastinal and anterior chest wall collaterals have opened up secondary to the SVC obstruction

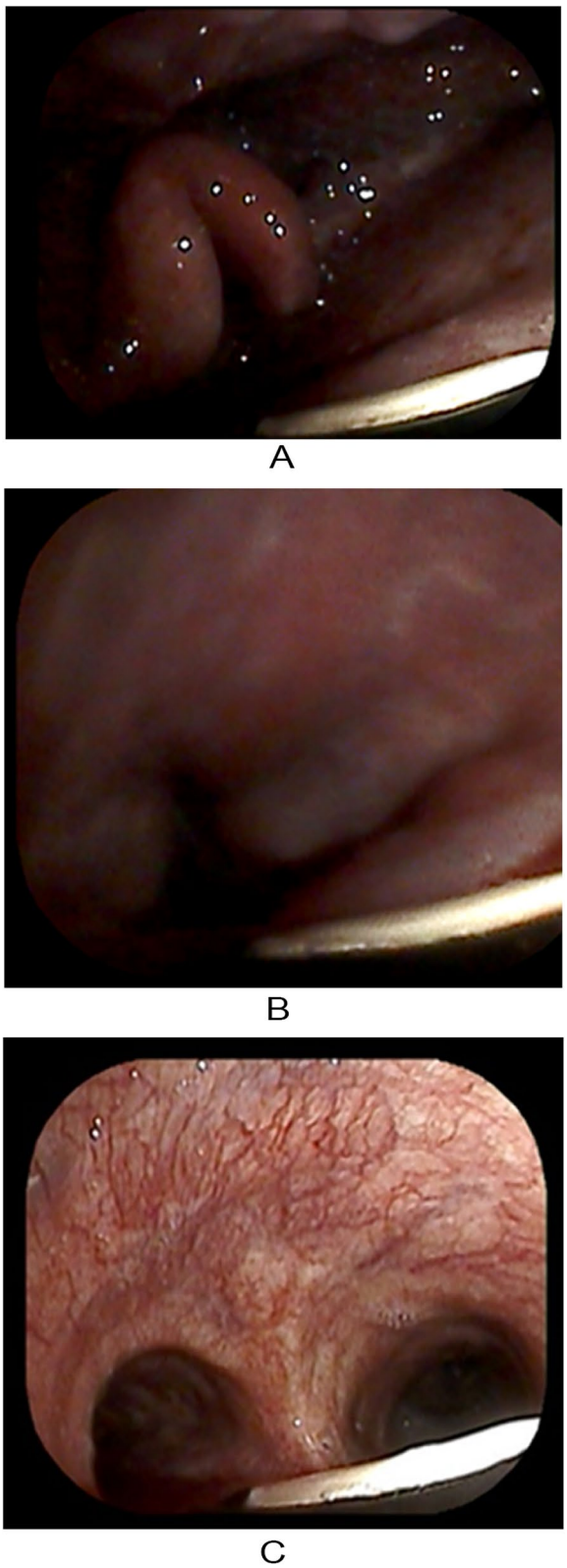

Fig. 2 Edematous epiglottis (**A**), glottic region (**B**) and carina with multiple mucosal collaterals (**C**)

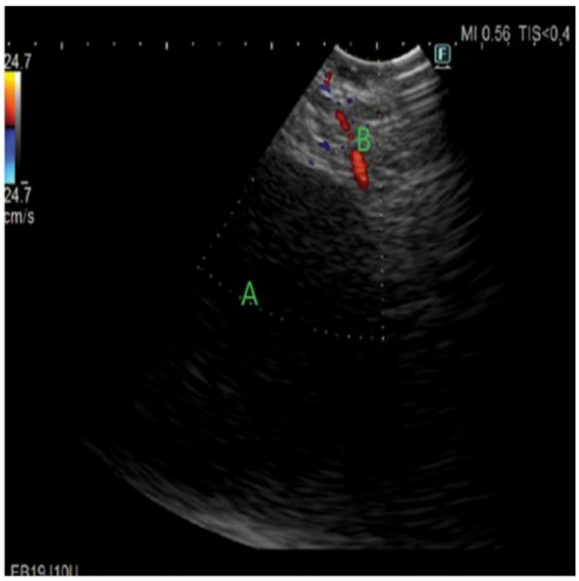

Fig. 3 Doppler EBUS view of the mediastinal mass. **A** Heterogenous echotexture mass. **B** Mediastinal collateral vessels in the periphery of the lesion

have sufficient time for collateral formation and growth. Therefore, severe or long-lasting SVC obstruction can occasionally be observed in slow-growing illnesses without any obvious accompanying symptoms [4, 5].

However, the bronchoscopic appearance of SVC syndrome has not been well explained in the literature. In a case report, a 64-year-old patient presented with SVC syndrome presented with acute dyspnoea, and an otolaryngologic examination revealed a supraglottic edema, mimicking angioedema [6].

In our patient, bronchoscopy showed oedematous supraglottic and glottic regions. The tracheobronchial mucosa showed high vascularity, which is logical but not well described in the medical literature. Doppler imaging while performing EBUS TBNA, showed a positive color sign around the paratracheal lymph nodes, suggestive of multiple collaterals in the mediastinum.

The conventional TBNA performed without EBUS guidance caused 7.7–13.3% of procedures to be terminated early due to hemorrhage [7]. The use of integrated color Doppler during EBUS enables real-time detection of collaterals and helps reduce bleeding. We performed EBUS-guided forceps biopsy in addition to FNAC because more tissue will be more useful for a confident diagnosis of lymphoma. Intranodal forceps biopsy is a well-described technique. In a meta-analysis, bleeding due to intranodal forceps biopsy occurred in 0.8% of cases. It is important that real-time Doppler is used to reduce the complications [8].

The mainstay of treatment for SVCS is the treatment of the underlying cause. Steroid therapy prior to mediastinal lymphoma biopsy may negatively impair pathological accuracy or cause delays in definitive diagnosis [9]. However, in high-risk patients, prebiopsy steroids seem to reduce the likelihood of cardiorespiratory morbidity [9]. Fortunately, in our patient, we obtained an accurate diagnosis, although the patient was on long-term steroids.

Conclusion

Non-Hodgkin's lymphoma is a rare cause of superior vena cava syndrome. When considering an unconfirmed diagnosis of lymphoma, it is best to avoid using glucocorticoids as they can be lympholytic and may impede the diagnostic process. In chronic SVC syndrome, due to the formation of extensive mediastinal collateral vessels, biopsies may be difficult. Doppler imaging while performing EBUS TBNA can reveal multiple mediastinal collaterals, which is useful in reducing the risk of bleeding. Bronchoscopy in SVC syndrome includes edematous supraglottic and glottic regions mimicking angioedema and multiple mucosal collaterals in the tracheobronchial tree.

Abbreviations

SVC	Superior vena cava
SVCS	Superior vena cava syndrome
EBUS	Endobronchial ultrasound
TBNA	Transbronchial needle aspiration
CT	Computed tomography
ROSE	Rapid on-site examination

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Authors' contributions

All authors were actively involved in the case report's drafting and modification. AE and BT did the literature search and manuscript writing. BT, BM, and TA were involved in the design, conceptualization, manuscript review, and editing. The authors reviewed and approved the final paper.

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Availability of data and materials

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Declarations

Ethics approval and consent to participate

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Consent for publication

Signed consent was taken from the patient.

Competing interests

The authors declare that they have no competing interests.

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