

Post-tuberculous sclerosing mediastinitis: A case report

L. Romane *, M.HINDI, M.Ijim, O.Fikri and L.Amro

Department of Pulmonology, Hospital ARRAZI, CHU Mohammed VI, Laboratories LRMS, FMPM, UCA.

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Abstract

Sclerosing mediastinitis is a rare condition characterized by excessive fibrous tissue formation within the mediastinum, often secondary to chronic infections like tuberculosis. This condition can lead to severe compression of vital mediastinal structures, resulting in significant clinical complications.

We reported the case of a 55-year-old woman with a history of inadequately treated pleuro-pulmonary tuberculosis. She presented with persistent dyspnea and dysphonia, and imaging revealed significant mediastinal fibrosis with compression of the superior vena cava (SVC) and thrombosis of the internal jugular vein. Histopathological analysis confirmed sclerosing mediastinitis with extensive fibrosis encasing the mediastinal structures.

The patient was initiated on anticoagulation therapy with Rivaroxaban to address the thrombotic complications. Due to the severity of SVC compression, she was scheduled for surgical placement of an SVC prosthesis to restore normal blood flow and alleviate symptoms. This comprehensive approach aims to prevent further complications and improve her quality of life.

This case highlights the critical need for early diagnosis and intervention in patients with post-tuberculous sclerosing mediastinitis. Comparisons with similar cases in the literature underscore the importance of a multidisciplinary approach to manage the complex clinical challenges associated with this condition. Early recognition and appropriate treatment are essential to prevent severe complications and ensure better patient outcomes.

Sclerosing mediastinitis, particularly following tuberculosis, requires prompt and effective management to mitigate serious health impacts. This case emphasizes the importance of tailored therapeutic strategies, including anticoagulation and surgical intervention, to address the unique needs of affected patients.

Keywords: Sclerosing Mediastinitis; Vascular Compression; Superior Vena Cava Syndrome; Anticoagulation Therapy; Post-Tuberculous; Multidisciplinary Management

1. Introduction

Sclerosing mediastinitis as known as Fibrosing mediastinitis is a rare entity usually caused by an exaggerated immune response to an infection, it is defined as a granulomatous disease. Most cases develop as a late complication of histoplasmosis or TB.

It is a serious condition characterized by the excessive growth of fibrous tissue within the mediastinum, the central compartment of the thoracic cavity.

* Corresponding author: L. Romane

This condition can lead to significant morbidity due to the compression and obstruction of vital structures such as the trachea, esophagus, and major blood vessels. Although various etiologies exist, one notable cause is a prior infection with *Mycobacterium tuberculosis*, which can trigger a profound fibrotic response even after the primary infection has been treated or resolved.

Tuberculosis remains a major public health issue worldwide, with millions of new cases reported annually. Despite advancements in treatment, complications such as fibrosing mediastinitis underscore the long-term impacts of the disease.

The diagnosis of fibrosing mediastinitis usually involves a combination of imaging tests and a biopsy of the affected tissue.

Treatment options for fibrosing mediastinitis are limited, and the goal is to manage symptoms and prevent complications. In severe cases, surgical interventions like bypass procedures or stenting may be considered to relieve compression of vital structures.

This case report aims to shed light on a unique instance of post-tuberculous fibrosing mediastinitis, highlighting the clinical presentation, diagnostic challenges, and therapeutic considerations associated with this rare disease.

2. Case presentation

Patient E.G, 55-year-old female homemaker with a history of primary amenorrhea that has not been investigated, as well as pleuro-pulmonary tuberculosis. She was first diagnosed with tuberculosis in 1997, but it was inadequately treated due to poor compliance. In 2008, she experienced a reactivation of pulmonary tuberculosis, which was treated successfully.

Over the past 10 years, the patient presented symptoms such as shortness of breath: dyspnea at stage IV of Sadoul's classification, chest pain along with dysphonia and stomach aches.

Upon examination, the patient revealed stable neurological and hemodynamic status, with a Glasgow Coma Scale score of 15. Blood pressure readings were 110/60 mmHg in the left arm and 104/59 mmHg in the right arm. She exhibited regular tachycardia with a heart rate of 90 beats per minute, and oxygen saturation was 98% on room air with a respiratory rate at 25cpm. Pulmonary examination was normal.

Clinical signs included noticeable thoraco-abdominal collateral circulation and a more pronounced jugular venous distension on the left side. Notably, the patient had varicose veins in the lower limbs, and the abdomen was soft epigastric tenderness.

Moreover, the patient lacked secondary sexual characteristics, including the absence of axillary and pubic hair and undeveloped breasts, consistent with her history of primary amenorrhea.

A chest X-ray revealed a para-hilar opacity: heterogeneous, poorly defined located near the right hilum. The opacity was not confluent, suggesting a complex or multi-focal process.

Mediastinal Widening which may indicate the presence of masses, lymphadenopathy, or other mediastinal structures and a Drooping Heart Sign.

For the etiological Workup, A bronchoscopic examination during hospitalization showed a diffuse bilateral inflammatory aspect of the second degree with free orifices. Bilateral staged biopsies revealed subacute and chronic inflammatory changes in the bronchial mucosa without signs of specificity or malignancy.

2.1. Tuberculosis (BK) Test: Negative in bronchial aspiration fluid

Imaging studies, including a thoracoabdominal and pelvic CT scan, revealed a mediastino-hilar mass on the right side with significant fibrotic encasement and infiltration of mediastinal elements, leading to structural compression with the invasion of the superior vena cava (SVC) resulting in marked compression of the vessel. This invasion resulted in the dilation of the azygos vein and ectasia of the inferior vena cava, with collateral circulation observed in the thoracic and abdominal walls. Additional findings included thickening of interlobular septa, bronchial distortions, and pseudo-

nodular condensations in areas of the middle lobe and lower right lung. Bone lesions, classified as IC according to the Lodwick classification, were noted in the right trochanteric mass.

A gastroscopy was also performed showed erythematous and atrophic pangastritis at the fundus and suggested a pseudo-compression of the middle third of the esophagus without luminal invasion. Histopathology confirmed an antral-fundic gastritis.

The patient's laboratory findings indicate mild leukopenia, and anemia, which may be consistent with chronic illness or previous tuberculosis. Elevated CRP points to an active inflammatory process. Other parameters, including coagulation profile, liver and renal functions, are within normal limits. Tumor markers were normal, ruling out active malignancy at the time of testing

A mediastinoscopy was performed to obtain tissue samples for histopathological examination.

- The histopathology results revealed an extensive fibrosis in the mediastinal tissue consistent with a diagnosis of sclerosing mediastinitis.
- The patient was diagnosed with sclerosing mediastinitis based on clinical presentation and diagnostic investigations.
- process and compression of surrounding structures.
- Regarding the impact assessment
- Transthoracic Echocardiography: Good systolic function of the left ventricle (LV) with an ejection fraction (EF) of 65%.
- Non-elevated systolic pulmonary arterial pressure (PAP).
- No valvopathies detected.
- Good function of the right ventricle (RV).
- Six-Minute Walk Test:
- The patient covered a distance of 430 meters, which is 68% of the theoretical distance. This suggests reduced exercise capacity.
- Arterial Blood Gas Analysis was normal.
- Pulmonary Function Tests (PFTs): Moderately restrictive ventilatory impairment, indicating reduced lung capacity to inhale and exhale.

Mildly reduced diffusion capacity with a diffusion capacity of the lung for carbon monoxide

These findings confirm the diagnosis of sclerosing mediastinitis, a rare condition characterized by excessive fibrous tissue formation within the mediastinum, leading to compression and obstruction of vital structures. The patient's clinical symptoms, including dyspnea and dysphonia, are consistent with the effects of this extensive fibrosis. The thrombosis of the SVC and the internal jugular vein further complicate the clinical scenario

Given the findings of internal jugular vein thrombosis and the risk of further thromboembolic complications, the patient was initiated on effective anticoagulation therapy with a Direct Oral Anticoagulant (DOAC), specifically Rivaroxaban.

Rivaroxaban was administered at an effective therapeutic dose, started with an initial higher dose (15 mg twice daily for the first 21 days) followed by a maintenance dose (20 mg once daily),

Goals of Anticoagulation, the patient was also initiated on corticosteroids at a dosage of 1 mg/kg per day.

Due to the significant compression of the superior vena cava (SVC) by the extensive fibrotic tissue associated with sclerosing mediastinitis, the patient is scheduled for the placement of an SVC prosthesis. This surgical intervention is necessary to alleviate the severe vascular obstruction and restore adequate blood flow.

3. Discussion

Sclerosing mediastinitis, also known as fibrosing mediastinitis, is a rare disorder characterized by excessive fibrous tissue growth within the mediastinum, often leading to compression and obstruction of vital structures such as the superior vena cava (SVC) and other mediastinal components. The condition can arise from various etiologies, with tuberculosis being a notable cause, particularly in regions where it is endemic.

Our patient, a 55-year-old woman with a history of inadequately treated pleuro-pulmonary tuberculosis, presented with severe symptoms including persistent dyspnea and dysphonia. Imaging and histopathological findings confirmed the presence of extensive mediastinal fibrosis, leading to significant vascular compression and complications such as thrombosis of the internal jugular vein. Similar cases have been reported in the literature, emphasizing the complexity and severity of post-tuberculous sclerosing mediastinitis.

In a review by Peikert and colleagues (1), it was highlighted that sclerosing mediastinitis often results from chronic inflammatory or infectious processes, including tuberculosis. The review discussed cases where extensive fibrosis led to compression of mediastinal structures, similar to our patient's condition. This review underscores the importance of early recognition and appropriate management of such cases to prevent severe complications.

Loyd et al. (2) reported a case where a patient with a history of tuberculosis developed mediastinal fibrosis, leading to compression of the SVC and necessitating surgical intervention. This case parallels our patient's situation, where significant fibrosis led to SVC compression, prompting the decision for a surgical prosthetic replacement to alleviate the obstruction and restore normal blood flow.

Gams et al. (3) described typical imaging findings associated with sclerosing mediastinitis, such as mediastinal widening and complex opacities near the hilum. Our patient's chest X-ray revealed similar findings, including a poorly defined opacity near the right hilum and mediastinal widening, which are critical indicators for diagnosing this rare condition.

Histopathological examination is crucial for confirming sclerosing mediastinitis. As noted by Denning and Deshaw (4), biopsy findings in such cases typically show dense fibrotic tissue infiltrating and compressing mediastinal structures. Our patient's biopsy results, which revealed extensive fibrosis with compression of the IVC and thrombosis of the internal jugular vein, align with these typical histopathological features, confirming the diagnosis and the severity of the disease.

For managing thrombotic complications, our patient was started on Rivaroxaban, a direct oral anticoagulant. The use of Rivaroxaban in similar clinical scenarios has been discussed in the literature for its effectiveness in preventing and treating thrombosis. Weitz and Eikenboom (5) highlighted the advantages of direct oral anticoagulants like Rivaroxaban over traditional anticoagulation therapies, emphasizing their ease of use and favorable safety profile, which is relevant to our patient's ongoing treatment plan.

The planned surgical intervention to place an SVC prosthesis is supported by various studies that have demonstrated the efficacy of such procedures in relieving symptoms and preventing further complications due to vascular compression. Rajesh et al. (5) discussed the successful outcomes of surgical decompression in patients with SVC syndrome, which is directly applicable to our patient, who faces similar vascular challenges due to the fibrotic process.



Figure 1 Scoot view showing mediastinal enlargement with retraction of the mediastinal elements towards the right side with atelectasis of the middle lobe with interstitial involvement

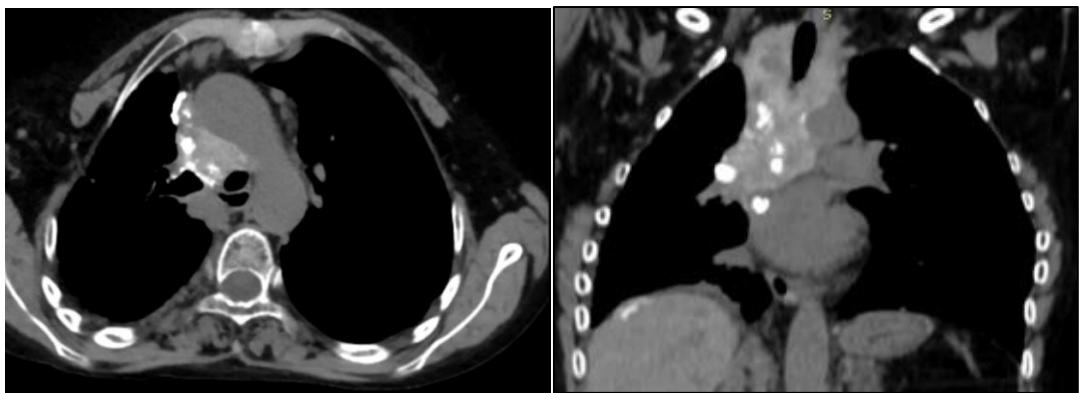


Figure 2 Axial and coronal sections of the SPC mediastinal window showing the spontaneously hyperdense mediastinal mass containing calcifications



Figure 3 CT SCAN: Sagittal Mediastinal window showing the collateral circulation parietal anterior

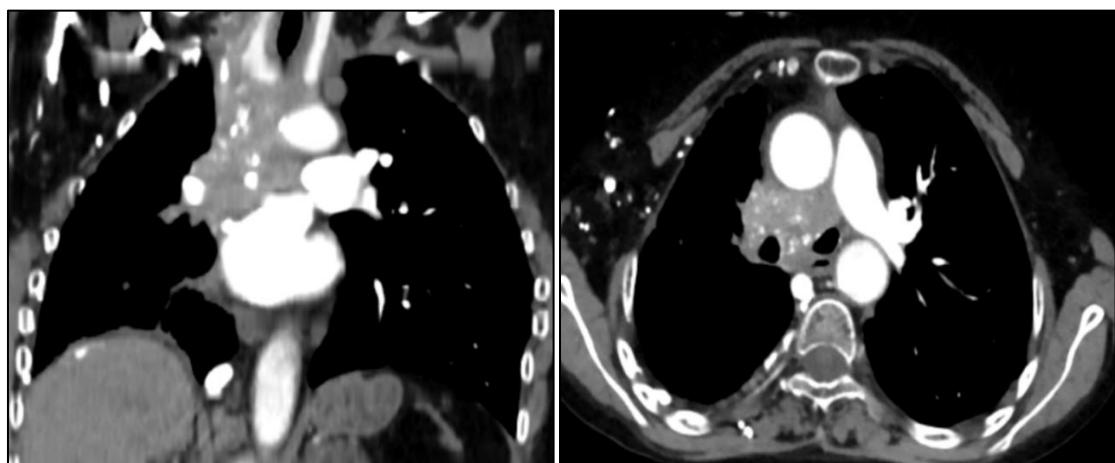


Figure 4 Axial and coronal sections of the mediastinal window injected in the arterial phase showing total inclusion of SVC

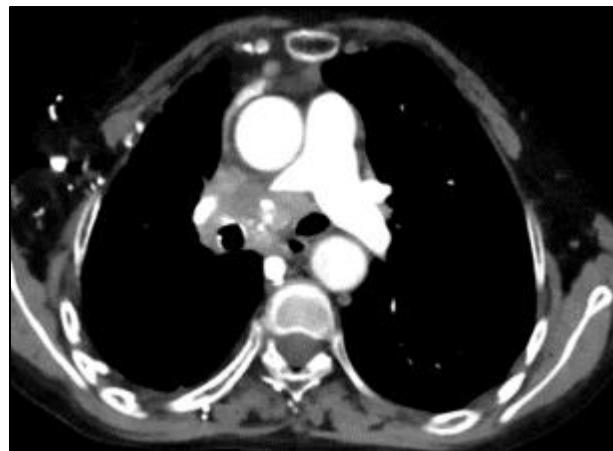


Figure 5 Axial section of the mediastinal window injected in the arterial phase showing total engulfment of the right pulmonary artery

4. Conclusion

This case underscores the critical need for awareness and timely management of sclerosing mediastinitis, particularly in patients with a history of tuberculosis. Early diagnosis, appropriate medical management, and surgical intervention when necessary are essential to improving patient outcomes. The comparison with similar cases in the literature highlights the importance of a comprehensive and multidisciplinary approach to treating this rare but serious condition.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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