Low Cellular Sclerosis of Anterior Mediastinum CT-guided Biopsy with Hydrodissection of this Rare Entity - Case Report

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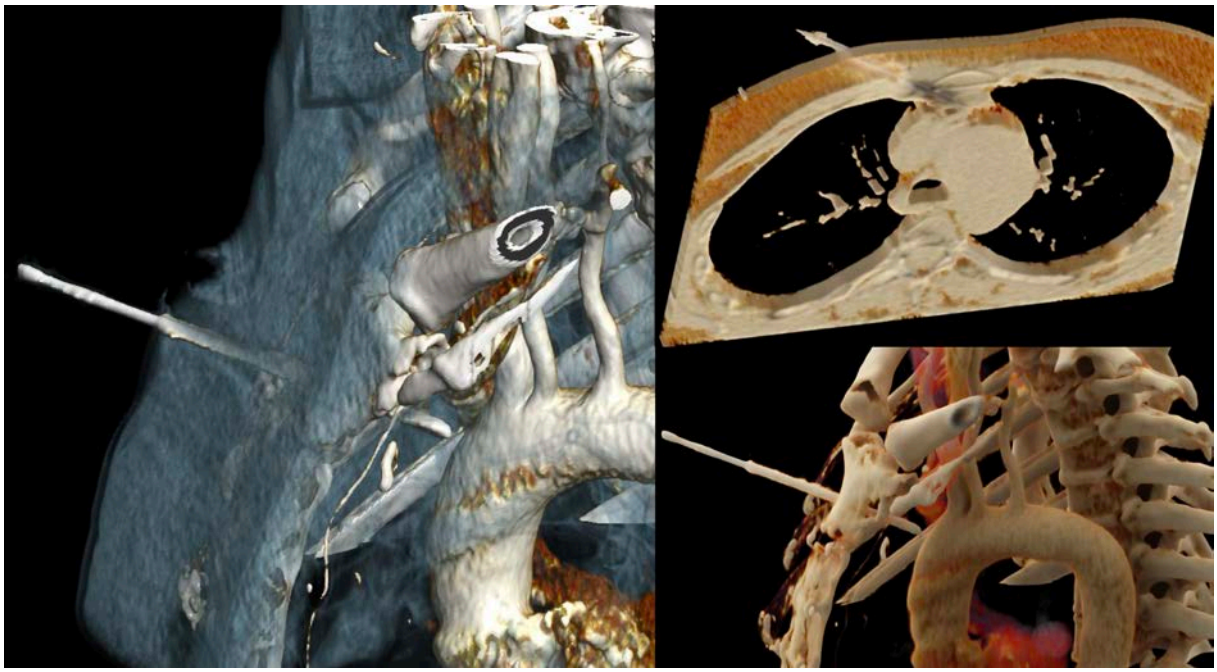
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Abstract

Conducting a CT-guided interventional procedure involving retrosternal pleural hydro-dissection of the juxta-aortic anterior mediastinum under local anesthesia revealed a rare histopathological condition known as benign mediastinal sclerosis. This challenging CT-guided intervention within the anterior mediastinum is scarcely documented in the existing scientific literature. In this presentation, we share our case along with relevant clinical data.

What makes this case distinctive is that the clinicians initially questioned the representativeness of the biopsy sample we obtained. Consequently, an additional sample was acquired through a highly invasive video-assisted mediastinoscopy procedure under intubation anesthesia. Remarkably, this second sample yielded the same histopathological result.

Mediastinal sclerosis is an exceedingly rare condition, often referred to as the "burnt-out" stage of sclerosing mediastinitis. While infectious etiologies are frequently identified, the majority of cases remain idiopathic.

Keywords: CT-guided biopsy – low cellular mediastinal sclerosis – sclerosing mediastinitis – aortic arch mass - retrosternal pleural hydrodissection

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Introduction:

Sclerosing mediastinitis was first described by Oulmont in 1855 and was considered a consequence of syphilis or tuberculosis until 1925 when Knox suggested an association with fungal infections. Sclerosing mediastinitis, also known as fibrosing mediastinitis, is a rare syndrome characterized by an aggressive fibroinflammatory process in the mediastinum. The progressive fibrosis caused by the proliferation of invasive fibrous tissue in the mediastinum [16] often leads to compression and functional impairment of vital mediastinal structures [9].

It has been suggested that mediastinal sclerosis represents the advanced, long-term phase of sclerosing mediastinitis. Given the rarity of sclerosing mediastinitis itself and its association with inflammatory and compressive symptoms in the upper mediastinum, the development of mediastinal sclerosis frequently requires medical intervention. In our specific case, the patient presents with no symptoms and was incidentally diagnosed. Reliable epidemiological statistics are not available due to the low number of cases. As far as is known, it mainly affects young adults and occurs from the first [20] to the seventh decade of life; a sexual, racial, or geographical predilection has not been reliably established so far [13].

Clinical data:

1) History:

A 62-year-old female patient, who is asymptomatic, had an incidental discovery of a paraaortic mass in the upper mediastinum during a chest X-ray examination. The subsequently performed CT examination of the thorax with contrast medium revealed the exact positional relationship of the paraaortic mass in the upper mediastinum (see Fig.1 A +C).

2) Laboratory parameters:

TB Eli-Spot: negative
Sputum, direct microscopy: negative
Antibodies for Toxoplasmosis: negative
Antib. for [22] Histoplasmosis: negative
IgG4-associated [17] laboratory parameters were within normal ranges.
IgG4 immunohistochemistry was not performed.

Radiological findings:

In the chest X-ray overview in the posteroanterior and lateral projection, as well as in the contrast-enhanced computed tomography of the chest, a paraaortic mass is observed in the anterior upper mediastinum without evidence of compression or deviation of vascular structures. There is no evidence of superior vena cava syndrome which is one of the principal complications [19] of this seldom entity.

Procedure (see Fig.1 and 2)

We began by creating a sterile environment at the puncture site and administered local anesthesia. The patient underwent a planning CT scan while lying in supine position. The previously identified paraaortic anomaly was clearly visible. We inserted a needle between the sternum and the internal thoracic artery, followed by a smooth hydrodissection procedure using a 40% glucose solution. While maintaining continuous hydrodissection, we isolated the lung and carefully advanced the needle into the parapleural mediastinum. Subsequent imaging confirmed that the needle was in an optimal position right in front of the target lesion. We then used an 18 G Tru-Cut needle to obtain multiple biopsy samples from the sclerotic mediastinal tissue immediately adjacent to the aortic arch (see Fig. 1G). These biopsy samples were preserved in formalin and sent to the Pathology Institute for further analysis. After removing all foreign materials, we applied a sterile adhesive bandage. During the follow-up examination, there were no signs of any primary complications.

Procedural strategy

We chose an anterolateral approach that would pass through the lung [1] but then used a mixture of contrast medium (CM), glucose, and mepivacaine to perform hydrodissection of the pleura. There are two key points to consider here:

a) In this case, a small dorsal pleural effusion was observed as a result of hydrodissection.

b) Glucose in the tissue can be painful. Therefore, we always mix it with a few milliliters of local anesthetic.

The further course

The intervention was carried out without any complications, and the patient was discharged without experiencing any issues.
However, due to the absence of malignancy in the pathological findings, an additional mediastinoscopy was conducted. As previously mentioned in the patient's medical history, the medical team had reservations about the histologic result obtained from our biopsy and opted to perform a video-assisted thoracoscopy with mediastinoscopy with intubation anesthesia. Once more, no malignancy was detected, and the findings remained consistent. Even a completely resected mediastinal lymph node (region 10L) showed no pathological findings.

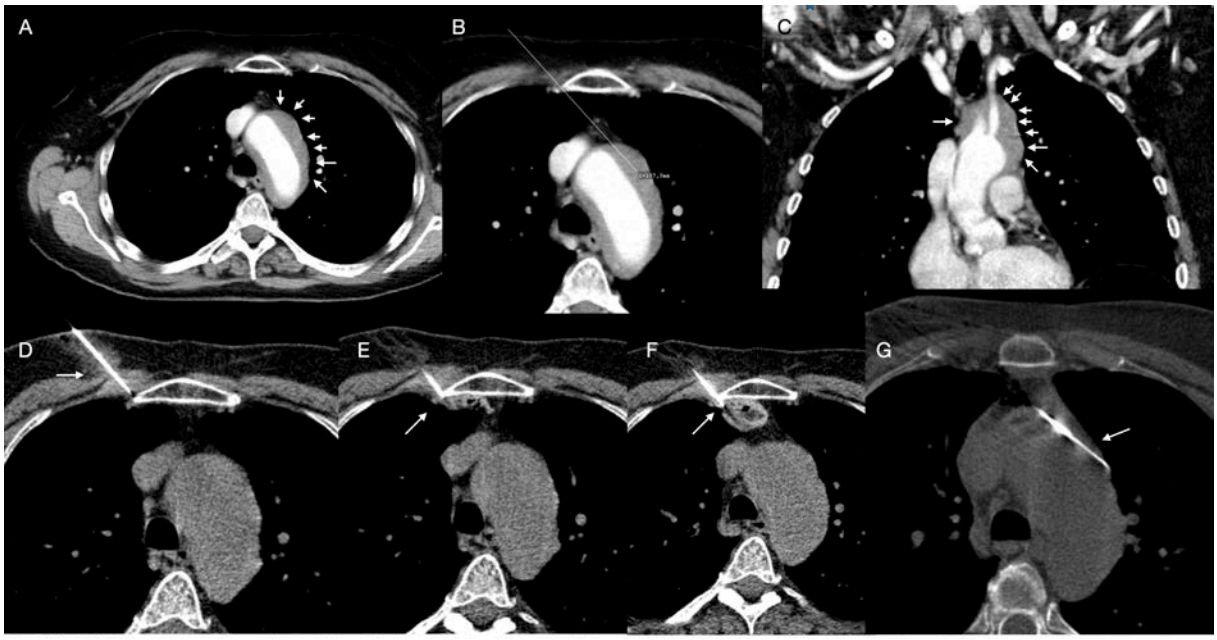


Fig. 1: A) and C) Axial and coronal CT images in arterial phase showing target lesion. Arrowheads. B) Planned access route. White line. D) Puncture and local anaesthesia with needle in position between sternum and internal thoracic artery. Arrow. E) Incipient pleural hydrodissection. Arrow. F) Hydrodissection of the pleura. Arrow. G) Biopsy needle in the sclerotic mediastinal tissue immediately adjacent to the aortic arch. Arrow.

Histo-pathological results

- Material: Mediastinal CT biopsy
Assessment: Low-cellular sclerosis of the mediastinum. No evidence of malignancy, including lymphoma.

- Material: Mediastinoscopic biopsy
Assessment (several samples): inconspicuous mediastinal fat tissue. Sparse cellular sclerosis of the mediastinum. No malignancy, especially no lymphoma.

- Material: Lymph node region 10L
Assessment: One normal lymphnode from region 10L. No malignancy, especially no manifestations of lymphoma.

Outcome

Four weeks after the surgical mediastinoscopic intervention, the patient visited our thoracic surgery outpatient clinic for a postoperative evaluation following the so-called VATS (Video-Assisted Thoracoscopic Surgery). The patient reported feeling generally well and had made satisfactory physical improvement. Both chest X-ray and laboratory results indicated improving postoperative recovery. Oxygen saturation while breathing room air was at 97%, and there was no longer a need for analgesics. As a result, the overall postoperative out-come is favorable. The wound conditions are not causing irritation.

Pathological-Radiological Correlation

The histological report revealed low cellular mediastinal sclerosis. The findings are consistent with the images. There is concordance of findings. Nonetheless, we recommend a follow-up assessment.



Fig. 2: Post-interventional findings after puncture. The juxtaaortic puncture channel is just barely visible in the target lesion. No complications.

Etiology still unknown

More than 150 years after Oulmont's initial description, the causes of mediastinal sclerosis remain largely unclear. While there are several theories and associative approaches, each can only provide a partial explanation (Tab. 1). Infections with Aspergillus flavus and terreus have been associated with sclerosing mediastinitis (SM) [10]. In the active proliferative phase of IgG4-related SM there is a notably FDG-uptake in PET-CT [6] [14] [17]. Pulmonary hypertension is one of the important [4] complications in patients with sclerosing mediastinitis (SM) [2, 3]. From a histological perspective SM is a rare, benign condition which occurs due to extensive proliferation of acellular collagen and fibrous tissue within the mediastinum [5].

Sclerosing Mediastinitis - Etiology unknown
Potentially Etiologic Associations

• Infections	**Infections**
• Autoimmune disease	**Fibroinflammatory disorders**
• Behçet disease	**Autoimmune disorders**
• Rheumatic fever	
• Radiation therapy	
• Hodgkin disease	
• Methysergide drug therapy for migraine headache	
• Retroperitoneal fibrosis	
• Sclerosing cholangitis	
• Riedel thyroiditis	
• Pseudotumor of the orbit	
• Sclerosing cervicitis	
• IgG-4 related syndromes	
• Systemic mastocytosis	
• Mast cell activation syndrome	

Tab. 1: Sclerosing Mediastinitis - Etiology unknown - Potentially Etiologic Associations modified according to [13]

Discussion

A CT-guided dorsal biopsy approach in the prone position is viable when the mediastinal mass extends sufficiently into the dorsal regions of the mediastinum [7]. However, in our case, we had no alternative but to approach from the ventral direction.

Although rare, sclerosing mediastinitis can result in extrinsic compression of both mediastinal and neighboring structures, including the cervical carotid artery. This compression, along with its hemodynamic implications, can be effectively addressed through innovative and durable endovascular therapy in the mid-term, offering a solution for this uncommon clinical problem [11]. In some cases SM was discovered with severe bronchial stenosis one year after external thoracic radio-therapy [24]. In histopathological exams often there are dense bundles and sheets of hyalinized collagen, accompanied by a relatively

sparse inflammatory infiltrate, serving as microscopic indicators of the disease. There are three different stages classified based on the ratio of fibrous tissue to the inflammatory component. The differential diagnosis for sclerosing mediastinitis encompasses intrathoracic desmoid tumors, solitary fibrous tumors, desmoplastic mesothelioma, and inflammatory myofibroblastic tumors (IMT) [15]. Evaluation for IgG4-related disease requires correlation of clinical findings, histopathology and, where appropriate, tissue immunostaining and serum IgG4 measurements. A normal serum IgG4 concentration alone does not exclude IgG4-related disease [28]. In this case, IgG4 immunohistochemistry was not performed. If the underlying cause of sclerosing mediastinitis is indeed IgG4-related, surgical resection may not be necessary for curing the disease [17, 18].

The mechanism by which Tamoxifen® operates in these sclerosing mediastinitis conditions remains uncertain. Tamoxifen® [18] is most widely recognized for its role as a partial agonist at the estrogen receptor. Fibroblasts express these receptors, and in approximately one-third of cases, they can be observed in desmoid tumors and desmoid like lesions, where they manifest as proliferations of fibroblasts [25].

A case of intrathoracic sclerosing mediastinitis, which resulted in a mass that resembled lymphoma [12], is reported in connection with long-term Propranolol® therapy [26].

Antifungal and antituberculous agents have been recommended for cases showing evidence of active granulomas. However, these, along with steroids, are typically ineffective in established cases with extensive fibrosis. Most reports highlight the benign nature of the condition and recommend minimal intervention unless symptoms become lifethreatening [21] [27].

Conclusions

SM can manifest as a mass lesion in the anterior upper mediastinum, requiring differentiation from other mass-forming sclerosing conditions like Nodular Sclerosis Hodgkin's lymphoma, sclerotic mesothelioma, oligo-metastatic phenomena and desmoplastic carcinomas. Therefore, the significance of an early micro-invasive biopsy cannot be overstated, as it is crucial for reaching a diagnosis and determining the histologic subtype of sclerosis, enabling the initiation of appropriate treatment.

This case highlights possibility of rare presentations of rare diseases in rare sites which need to be approached with high index of suspicion in an endeavor to achieve an early tissue diagnosis [8] for effective management and favourable outcome.

Conflict of interest:

The authors declare that there were no conflicts of interest within the meaning of the recommendations of the International Committee of Medical Journal Editors when the article was written.

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