Cardiac Tamponade: Initial Clinical Presentation of Fibrosing Mediastinitis

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Introduction

Fibrosing mediastinitis (FM) is a rare condition characterized by the proliferation of fibrotic tissue in the mediastinum, presenting as a concentrated mass or diffuse infiltration. Clinical conditions can range from benignity to obstruction of critical mediastinal structures, such as vessels, airways, and the esophagus. Common complaints are cough, dyspnea, recurrent pulmonary infection, hemoptysis, and pleuritic pain. Pericardial involvement is not frequently reported in the literature. In this article, we describe a rare case of major pericardial effusion with cardiac tamponade as an initial presentation of FM in a patient with cancer.

Description

A 64-year-old female patient with hormone receptor-positive and human epidermal growth factor receptor 2-positive breast cancer of the invasive ductal carcinoma subtype underwent a mastectomy in 2018 and received adjuvant treatment with paclitaxel and trastuzumab, followed by anastrozole. During routine oncology follow-up, a chest radiograph revealed enlargement of the cardiac area. Additional investigation with echocardiography showed significant pericardial effusion with hemodynamic repercussions (Figure 1, Video 1) requiring urgent pericardial drainage. Pericardial fluid analysis and histology did not indicate neoplastic involvement but showed inflammatory findings. Pericardial fluid cultures were performed and were negative. Following the surgical procedure, the echocardiogram revealed a significant increase in thickness in the right atrium and atrioventricular groove. Computed tomography (CT) of the chest revealed significant evidence of FM, with extensive involvement of the middle and posterior mediastinum, including the arch and descending aorta, and extension into both pleural spaces (Figure 2). One year later, the patient continues to receive treatment with anastrozole and colchicine and remains asymptomatic.

Discussion

FM is a rare disorder resulting from excessive fibrotic proliferation in the mediastinum. Although the pathogenesis of this condition is not fully understood, the most probable cause is an immunological trigger for the proliferation of fibroblast cells. FM can be idiopathic or secondary to other conditions such as infections and malignancies. Infectious causes include tuberculosis, aspergillosis, cryptococcosis, and histoplasmosis, the latter being the most common one, especially in endemic regions. Other conditions associated with FM are radiation therapy, malignancies (especially lymphoma and mesothelioma), sarcoidosis, and medications. In addition to these, some chemotherapy drugs such as paclitaxel, cyclophosphamide, bleomycin, busulfan, Carmustine, and gemcitabine are associated with pulmonary fibrosis and may be related to FM.

After left mastectomy, the patient received adjuvant chemotherapy treatment with paclitaxel and trastuzumab, followed by anastrozole, without exposure to other known risk factors for FM. Due to the paucity of publications about FM, epidemiological data are still insufficient.

Cardiac tamponade, on the other hand, is caused by the accumulation of fluid in the pericardial sac, hindering the physiological functioning of the heart. The most common symptoms are tachypnea, tachycardia, and hemodynamic instability, in addition to an increase in systemic venous pressure. The occurrence of tamponade due to compression of the cardiac chambers by a mass, such as the FM described, is uncommon. Considering the aforementioned, to the best of our knowledge, this is the first report in the literature of FM initially presenting as cardiac tamponade.

Most patients with FM present with signs or symptoms related to obstruction or compression of critical mediastinal structures, such as the central airways, superior vena cava, pulmonary veins, pulmonary arteries, and the esophagus. The heart, pericardium, coronary arteries, aorta, and aortic branch vessels are much less frequently involved. Interestingly, our patient only presented mild complaints related to large pericardial effusion and the extensive involvement of the mediastinum and retroperitoneum.

Clinical and laboratory findings in FM are nonspecific. Imaging tests such as chest radiography, contrast-enhanced chest CT, positron emission tomography (PET), and chest magnetic resonance imaging (MRI) are important to

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Video 1 – Two-dimensional echocardiogram showing significant anechoic pericardial effusion with signs of hemodynamic repercussion. Parasternal long-axis view (A); Parasternal short-axis view (B); Apical four-chamber view (C); Subcostal view (D).
Link: http://abcimaging.org/supplementary-material/2023/3603/2023-0038_RC_video_1.mp4

Figure 1 – Transthoracic echocardiogram showing a large pericardial effusion (A), respiratory variation in tricuspid valve higher than 40% (B), and subxiphoid view of the dilated inferior vena cava greater than 2.1 cm (C).

assess the extent of fibrosis and to guide and monitor therapeutic interventions. However, they do not confirm the diagnosis—they only suggest it. Confirmation is only possible through histopathological evaluation performed after thoracotomy, as ultrasound- or CT-guided biopsy does not obtain enough fragments for the differential diagnosis.

Chest radiography has low sensitivity and specificity for FM. Its findings include mediastinal enlargement, presence of pulmonary infiltrates, and a mediastinal or hilar mass that may or may not contain calcifications, airway narrowing, or septum enlargement. In the present case, the only change in the chest radiograph was an increase in the cardiac area due to pericardial effusion. Contrast-enhanced chest CT is the imaging method of choice for diagnosis, although it does not differentiate between malignant and benign conditions in the absence of clear signs of metastasis. CT may have a focal or diffuse FM pattern. The focal pattern, marked by the presence of a localized, often calcified mediastinal mass of soft-tissue attenuation in the right paratracheal or subcarinal region, is associated with histoplasmosis. On the other hand, the diffuse pattern is marked by a diffusely infiltrating, often noncalcified mass affecting multiple mediastinal compartments and that may extend into the retroperitoneum. PET-CT also does not allow the differentiation between malignant and benign causes, but may assist in the diagnosis and determination of target sites for biopsy, in addition to providing information on response to therapeutic interventions. MRI is similar to CT in determining the extension and involvement of adjacent structures in FM, but it does not consistently demonstrate calcifications. Unfortunately,
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Echocardiographic evaluation of the mediastinum is challenging. However, some findings, such as pericardial effusion, constrictive pericarditis, and thickening of cardiac structures surrounding the heart, may be present in FM.

FM usually has an unpredictable course, with spontaneous remission or symptom exacerbation. The most common causes of death are recurrent infection, hemoptyis, or cor pulmonale. The mortality rate of patients with bilateral subcarinal or mediastinal involvement is higher than that of patients with more localized mediastinal or hilar fibrosis. Awareness of this condition is low, and it is often suspected only in the presence of clear signs. Although the prognosis tends to be severe, as the diagnosis is often delayed and most patients die due to compression of critical structures within the mediastinum, our patient no longer presents pericardial effusion, and all symptoms have resolved.

Conclusion

FM is a rare and severe disease whose diagnosis is challenging and often delayed. The importance of multimodal imaging, particularly in the oncologic setting, cannot be understated, as it plays a crucial role in the early detection and management of this condition. In terms of prognosis, FM is often associated with a poor outcome, as patients frequently experience compression of critical structures within the mediastinum, resulting in life-threatening complications. Further research and awareness are needed to achieve timely diagnosis and optimize treatment strategies for FM.

Figure 2 – CECT (mediastinal window). A-C: axial plane, shows a soft-tissue attenuation mass diffusely infiltrating the middle and posterior mediastinum, involving the arch (A) and descending (B) aorta and extending into both pleural spaces (arrows in B). Coronal plane highlighting enlargement and signs of fibrosis in the mediastinal region (C). Maximum intensity projection (MIP) (D), the periaortic mass continued into the upper abdomen encasing and narrowing the origin of celiac trunk (white arrow) and superior mesenteric artery (black arrow).

Author Contributions

Conception and design of the research, writing of the manuscript, critical revision of the manuscript for intellectual content: Monteiro Neto AJO, Delgado VM, Leite FMA, Dutra AB, Melo MDT; acquisition of data, analysis and interpretation of the data: Melo MDT.

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Study Association

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References


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