Idiopathic fibrosing mediastinitis

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Fibrosing mediastinitis is rare in settings where histoplasmosis is not endemic. An idiopathic form of the disease may present with indistinguishable features and requires methodical exclusion of competing differential diagnoses. We report the case of a 30-year old female patient who presented with intermittent haemoptysis for the past 2 years with no constitutional symptoms. Computed tomography of the chest revealed a prominent right bronchial arterial circulation with a mass-like lesion, which encased and attenuated the right pulmonary trunk and adjacent structures. Endobronchial ultrasonography with transbronchial fine-needle aspiration showed a paucicellular aspirate with no evidence of malignancy or granulomas. Fungal infection, tuberculosis, sarcoidosis, IgG4-disease, and connective tissue disease were ruled out by appropriate serological, molecular, and microbiological tests. A diagnosis of idiopathic fibrosing mediastinitis was therefore made by exclusion and the patient was successfully treated with oral corticosteroids.

Keywords. idiopathic fibrosing mediastinitis; fibrosing mediastinitis; haemoptysis.

Afr J Thoracic Crit Care Med 2021;27(2):60-62. https://doi.org/10.7196/AJTCCM.2021.v27i2.064
was initiated and the patient remains clinically stable without evidence of disease progression during follow-up care. She has had no further episodes of haemoptysis.

Discussion

Idiopathic fibrosing mediastinitis is a metabolically active space-occupying fibro-inflammatory disease in the mediastinum. Many hypotheses have been advanced for its aetiology; however, the exact cause remains unknown. IgG4-related disease has been recognised as having significant overlap with idiopathic fibrosing mediastinitis and must be actively excluded with use of serum levels of IgG4 and histological markers. Idiopathic fibrosing mediastinitis has also been associated with other idiopathic fibro-inflammatory disorders and autoimmune disease (Fig. 2).

The microscopic characteristics of fibrosing mediastinitis reveal abundant, paucicellular, fibrous tissue infiltrating and obstructing adipose tissue. Granulomas are usually absent in patients with idiopathic fibrosing mediastinitis. Idiopathic fibrosing mediastinitis is progressive when left untreated and can be staged as follows:

- stage 1 lesions are characterised by oedematous fibromyxoid tissue associated with an inflammatory reaction, thin-walled vessels, and lacking cellular atypia and necrosis.
- stage 2 lesions are poorly demarcated lesions consisting of haphazardly arranged hyaline material encircling and infiltrating mediastinal structures with minimal inflammatory reaction.
- stage 3 lesions are obliterative, which involve mediastinal structures, and are characterised by a cellular dense collagen and occasional lymphoid aggregates. Spindle cells and inflammatory cells are absent. Dystrophic calcification is commonly seen.

Idiopathic fibrosing mediastinitis may present with one of two main radiological patterns – focal or diffuse disease. Focal fibrosing mediastinitis usually involves the right mediastinum, hilar and subcarinal region. The diffuse type can extend to the soft-tissue structures of the neck, posterior mediastinum and the lung. The majority of patients will experience symptoms and exhibit signs of compression of mediastinal structures, which include pulmonary arterial or venous narrowing, superior vena cava obstruction, and airway narrowing.

Affected patients are usually young and present with symptoms related to obstruction of vital mediastinal structures, such as the oesophagus, airways, pulmonary arteries or veins and central systemic veins. The most common presenting complaints include cough, dyspnoea, haemoptysis and pleuritic chest pain. Haemoptysis can affect up to 20% of patients and has several potential causes in patients with fibrosing mediastinitis. Airway obstruction with a post-obstructive necrotising pneumonia, invasion of a bronchus by fibrous tissue, and pulmonary hypertension from pulmonary vascular compression are the most common clinically encountered mechanisms. Obstruction of the pulmonary arteries, in particular, may lead to extensive anastomoses with intercostal or bronchial arteries, which increases the risk for massive haemoptysis.

Idiopathic fibrosing mediastinitis is usually a progressive disease with no evidence-based therapeutic options. Oral corticosteroids are the most commonly used medical treatment and have been associated with variable success. Other potentially efficacious therapies include tamoxifen, methotrexate and mycophenolate mofetil. Rituximab has also been shown to be associated with a favourable therapeutic response in patients with progressive and refractory disease. Surgical biopsy should be performed in patients with poor response to therapy. Surgical resection is curative in localised disease and may ameliorate symptoms. A complete resection may require vascular and airway reconstruction, which is associated with high morbidity and mortality. Patients with bilateral mediastinal involvement, extensive fibrosis, calcifications and collateral vessels are generally not suitable for surgery. Symptomatic patients can also be treated with local therapies directed towards re-establishing patency of occluded airways, pulmonary arteries or vena cava.

Conclusion

This case highlights a common presentation of a rare disease and the challenge of establishing the diagnosis of idiopathic fibrosing mediastinitis. The diagnosis of idiopathic fibrosing mediastinitis requires suggestive radiological and pathological findings, and the methodical exclusion of competing differential diagnoses.
Fig. 2. Classification of mediastinal fibrosis. (RPF = retroperitoneal fibrosis; ANCA = antineutrophil cytoplasmic antibodies.)

Declaration. None.

Acknowledgements. None.

Author contributions. FDM and RP contributed equally to the writing of this case report. GS and LM edited the manuscript. All authors approved the manuscript for submission.

Funding. None.

Conflicts of interest. None.

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Accepted 9 March 2021.