Radiology pearl

Progressive massive fibrosis

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Clinical Presentation

A 40 year old male presented with cough, expectoration and dyspnea (grade 2 MMRC) over last several years. There was no history of fever or loss of appetite .General examination was unremarkable. Respiratory examination revealed bilateral diffuse rhonchi on auscultation. Chest X ray showed rounded mass opacities in bilateral upper zone and sequential x rays showed a static course. He was on antitubercular treatment for more than one year without clinicoradiological improvement. His sputum was repeatedly negative for AFB and was referred as a case of suspected drug resistant tuberculosis.



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What is the diagnosis?

Add to this his occupational exposure history - he had worked in a stone crushing unit for almost 10 years and had stopped working a few years back when his complaints started. In view of definite occupational exposure and suggestive radiological picture he was diagnosed as a case of silicosis(complicated)

Discussion

Failure to respond to antitubercular treatment both clinically and radiologically should not only prompt consideration of drug resistance or noncompliance to treatment but also reconsideration of diagnosis especially if microbiological evidence is lacking. Inadequate response to treatment on the background of co-morbidities like diabetes mellitus and human immunodeficiency syndrome must also be kept a possibility.

Progressive massive fibrosis (PMF) refers to the formation of large conglomerate mass like lesions with irregular margins predominantly in the upper lobes. Though classically described in the context of pneumoconiosis like Coal worker's pneumoconiosis and silicosis, it has occasionally been described with talcosis¹, berylliosis¹ kaolin pneumoconiosis² and pneumoconiosis from carbon compounds,² such as carbon black, graphite, and oil shale. Conglomerate masses can also develop in Sarcoidosis³.The diagnosis of PMF requires the presence of a large opacity exceeding 1 cm (on Chest x-ray). PMF can be mistaken for bronchogenic carcinoma and vice versa. PMF lesions tend to grow very slowly, so any rapid change in size or development of cavitations, should prompt a search for either alternative cause or secondary disease. Carcinoma and tuberculosis are two potential complications. The large opacities gradually migrate toward the hilum, leaving emphysematous lung tissue between the fibrotic tissue and the pleural surface. Treatment is conservative with avoidance of further exposure but it is usually progressive even after cessation of exposure. The diagnosis of pneumoconiosis relies heavily on a proper meticulous history and is one of the DPLDs where only history is rewarding in making a proper diagnosis.

Reference

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