Spectrum of radiological appearance of Progressive Massive Fibrosis

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Learning objectives

The purpose of this educational exhibit is:

1. To illustrate and describe the key features of PMF on radiograph and high resolution computed tomography.
2. To correlate imaging findings with pathological meaning, as well as clinical history and investigations.
3. To highlight occupations that can be associated with PMF and explore imaging differentials.
Background

Progressive Massive Fibrosis (PMF) is a radiological term for the accumulation of fibrotic nodules greater than 10 mm diameter in lung tissue.[1] It is also known as a complicated form of the pneumoconioses, such as coal workers’ pneumoconiosis or silicosis. Traditionally, PMF is thought to be associated with extended occupational tenures of ten or more years. However, in the setting of very high dust exposures, PMF has been described as developing within few years. As suggested in its name, the disease is progressive with loss of lung tissue and function continuing beyond the period of dust exposure. The development of PMF is well established in dust-exposed workers such as coal mine workers, stonemasons and hard rock miners.[2,3]
Imaging findings OR Procedure details

This poster highlights the spectrum of imaging appearances of PMF using five cases from experience at the Wesley Dust Disease Research Centre. Clinical history is discussed, as well as medical imaging using the internationally recognized pneumoconiosis grading systems for chest radiograph and high resolution computed tomography (HRCT). On both chest radiographs and HRCT, PMF is described based on size as either A (10-50 mm), B (>50 mm but not exceeding equivalent area of the right upper lung zone), or C (the longest diameter exceeds the equivalent area of the right upper lung zone).[4,5]

Case 1

Textbook descriptions of PMF instruct readers to notice large bilateral opacities with irregular margins, found primarily in posterior upper and middle lung lobes.[6] Compensatory emphysematous change may also occur as well as features of pulmonary hypertension. Case 1 demonstrates an example of the radiographic features of advanced PMF in a 36 year old male with a high dust exposure working with artificial stone benches (Figure 1). The widespread opacifications may be confused with infective processes, with case 1 as an example being originally reported as pneumonia by a general Radiologist.

The equivalent histopathological finding is described as an avascular amorphous fibrotic mass of cross-linked insoluble proteins and peripherally poorly defined coarse hyalinised collagen bundles, with associated pigment found in macrophages and freely within the tissue.[5] Necrosis can be seen with chronic inflammatory cellular infiltrates. Thought to be ischaemic, central necrosis contributes to the differential diagnosis of tuberculosis (TB), further complicated by an association between silicosis and TB.

Case 2

Case 2 shows PMF in a 74 year old male with >20 years history of dust exposure in the coal mining industry, undertaken in both underground and open-cut environments. He was symptomatic with severely obstructed pulmonary function and a significant smoking history (>60 pack-years). His co-morbid diagnosis of emphysema, contributes to his primary symptom of dyspnoea on exertion. PMF is typically a bilateral, symmetric solid spiculated mass but can be unilateral, centrally cavitating or centrally necrotic. PMF can therefore occasionally mimic the appearance of lung cancer, made more difficult by the fact that PMF is also avid on Positron Emission Tomography (PET) (Figure 2). This case might be considered the stereotypical clinical scenario for PMF, however particularly in view of the patient's age and significant smoking history, it raises the important imaging differential of lung malignancy.
Case 3

In contrast, Case 3 is an asymptomatic 37 year old male smoker (10 pack-years) with <10 year history of exposure in underground coal mining in the setting of a trade role in mine development. While pulmonary function tests were normal, bilateral PMF in the setting of silicosis is demonstrated in both radiograph and HRCT (Figure 3). This case particularly highlights the importance of considering PMF in patients with much less exposure history and various occupational roles as auxiliary roles to the mining process can have clinically relevant exposure.

Case 4

Similar to its radiological appearance, PMF exists on a spectrum in terms of severity of disease and its progression in each individual. While prevention is key and there is no cure, earlier identification could inform lifestyle and occupational choices to manage the disease more effectively. Case 4 is a symptomatic 45 year old male ex-smoker (2 pack years) with 27 year history in stonemasonry. No features of PMF were identified on CT performed at baseline (images not shown). However, the progression of large bilateral lesions is evident on HRCT over a 9 month period 7 years later (Figure 4). It demonstrates considerable progression of the disease over several years, but notably of the left sided lesion in particular in less than 12 months.

Case 5

Case 5 demonstrates PMF in a 33 year old stonemason, asymptomatic with unknown smoking history, working in dry cutting with 2 years of extremely high dust exposure and ~75% PPE compliance rate. Importantly, the chest radiograph appears largely normal and was reported as ILO negative (profusion grade 0/0) by a B-reader (Figure 5). The PMF lesion is revealed in HRCT, with associated satellite nodules and subtle ground glass changes. This case also highlights the recommendation to use CT as a follow-up modality.
Fig. 1: Chest radiograph of advanced PMF in a 36 yr old stonemason.

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Fig. 2: HRCT showing mixed dust pneumoconiosis with COPD and PMF in 74 yr old male: type B opacity 70mm right upper zone (a,b) with a type A opacity 40mm left upper zone (not shown). PET-CT imaging demonstrates avid nature of a PMF lesion (c).


Fig. 3: Bilateral PMF in a 37 yr old male with coal mining exposure. (a) Chest radiograph (ILO 2/1) and (b) HRCT showing large bilateral apical A type opacities: 14mm right upper zone, 23mm left upper zone.

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**Fig. 4:** HRCT (a) with ground glass centrilobular lesions in bilateral upper and mid-zone regions, the largest a right upper zone 70mm lesion. Marked mediastinal and hilar lymphadenopathy with "egg shell" calcification was noted, characteristic of silicosis. A subsequent HRCT 9 months later (b) shows progression of nodules to 73mm right upper zone and 53 mm left upper zone.

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**Fig. 5:** Chest radiograph (a) shows no clear features of the PMF nodules revealed in HRCT (b) in a 33 yr old asymptomatic male stonemason.

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Conclusion

This educational exhibit demonstrates the variation of appearance of PMF, as well as highlighting the importance of recognizing the features of PMF in occupations other than coal mining. This is a particularly relevant topic to Australia with its strong resource sector and history of mining and occupations associated with dust exposure. The limitation of chest radiography in assessing for PMF is highlighted, noting that a normal radiograph may not reveal significant disease as seen on HRCT. Imaging differentials can include lung malignancy, granulomatous disease such as sarcoidosis, infections such as tuberculosis and even pneumonia on radiograph. It is essential for the clinician and radiologist to maintain a high level of suspicion and consider carefully the occupational history in these cases.
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