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Letters to the Editor

Diagnosing pulmonary alveolar proteinosis: Is there any role of radiology?

Dear Editor,

We read with interest the article titled “Primary alveolar proteinosis – A report of two cases” written by Bhattacharyya et al¹ and published in MJAFI 2013;69:90-93. Having gone through the article, we would like to emphasize certain facts which might be relevant to all of us. The authors have presented four axial images of CT scan of chest, one photomicrograph of histopathology of open lung biopsy, one photograph depicting lung lavage fluid samples in serial order from case 1 and one axial image of CT scan of chest from case 2 in support of the case reports.

The key words mentioned in the article are 1) Pulmonary alveolar proteinosis, 2) Whole lung lavage and 3) Granulocyte–macrophage colony stimulating factor. It is surprising that HRCT scan chest is not listed in the key words. As mentioned in the case history, both the cases were treated empirically with ATT despite sputum being negative for AFB and chest radiograph depicting bilateral extensive alveolar opacities. It would not be incorrect to assume at this point that the working diagnosis before the CT scan studies was pulmonary tuberculosis. Having gone through the cases mentioned in the article, it is quite evident that CT scan chest had

played a decisive role in directing further investigations for confirming the diagnosis.

The authors portray the HRCT findings as ‘crazy paving pattern’ in both the cases without any mention about the radiological diagnosis. It is on record that pulmonary alveolar proteinosis (PAP) was given as the first radiological diagnosis based on CT scan findings in both the cases despite the clinical diagnosis being pulmonary tuberculosis. It would be pertinent to mention here that radiological diagnosis of PAP was not made on the basis of ‘crazy paving pattern’ alone. Due consideration was given to the clinical setting in each case before suggesting the possibility of PAP. In fact, in case 2, bronchoalveolar lavage (BAL) was suggested after the CT scan study. It may also be relevant here to mention that, although ‘crazy paving pattern’ on HRCT chest in an appropriate setting is highly characteristic of PAP, there are a host of other conditions e.g. infectious (pneumocystis carinii pneumonia), neoplastic (mucinous bronchoalveolar carcinoma), idiopathic (sarcoidosis, non-specific interstitial pneumonia, organizing pneumonia), inhalational (lipoid pneumonia), misc (adult respiratory distress syndrome, pulmonary hemorrhage) and many more which can cause similar appearance on HRCT chest.

Fig. 1 – Serial axial CT scan sections (lung window) at immediate infra-carinal (a–c) and mid cardiac level (d–f) depicting partial clearance of lesions after nearly six months (b, e) and two years (c, f).

The authors mention that both the cases were successfully managed with whole lung lavage. It is also mentioned that there was marked improvement in the condition of both the patients after one week of BAL. There is however, no follow up available for either of the patients. It is well known that patients of PAP may require repeat BAL after a certain interval of time which may vary from patient to patient. Some of the patients do not show any remission and rather worsen in severity. Hence, is it appropriate to comment on the success of BAL just on the basis of one week of observation? For the benefit of readers of the MJAFI, we illustrate serial CT scan findings of case 2 who was followed up with CT scan once after nearly six months and then exactly after 2 years (Fig. 1). The follow up images available with us, depict only partial resolution of lesions even after two years.

Overall, we find that the role of radiology has been understated by the authors in the diagnosis and management of both these cases. This is likely to give an erroneous message to the readers of the esteemed journal of the AFMS.

Reply

Dear Editor,

On behalf of all the authors, I thank the writers for their deep interest, thorough study and in depth imaging analysis and follow up of the case report titled "Primary alveolar proteinosis – A report of two cases".¹ I agree with the writers that HRCT scan chest is an important test in the diagnosis and follow up of a case of pulmonary alveolar proteinosis (PAP).

However, we disagree with the writers about listing HRCT scan chest as one of the key words in the present case report. Key words are listed by authors of scientific articles to help readers to search for the articles they are interested in reading or doing research. HRCT scan chest gives a clue to the diagnosis of PAP in an appropriate clinical set up. However, PAP is a rare disease. Hence, out of more than a few hundreds of HRCT scan chest done in a centre, only a few cases will be finally diagnosed as PAP. The readers have also given at least eight causes of HRCT scan chest findings similar to PAP. Hence, listing HRCT scan chest as one of the key words in the present case report is not justified as it is unlikely to help many readers searching for articles on PAP or whole lung lavage (WLL).

Both the cases were treated at peripheral hospitals as pulmonary tuberculosis. This has been mentioned in the case report. The patients were evaluated subsequently at the tertiary care centre with HRCT scan chest, bronchoscopy, bronchoalveolar lavage (BAL) and lung biopsy, which confirmed the diagnosis of PAP. Readers will agree that HRCT scan chest alone cannot confirm a diagnosis of PAP without BAL and/or histopathological findings.

Response to WLL is evaluated clinically (both symptoms and signs), and by objective physiological parameters (which

REFERENCE

1. Bhattacharyya D, Barthwal MS, Katoch CDS, et al. Primary alveolar proteinosis – a report of two cases. *MJAFI*. 2013;69:90–93.

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include forced vital capacity (FVC), DLCO, PO₂ at rest and with exercise), along with X-ray and HRCT scan chest.² In the present case report, both patients showed improvement in clinical and physiological parameters following WLL. Similar responses were maintained when they came for follow up subsequently. But case 2 showed partial resolution in HRCT scan chest as mentioned by the readers. Similar observation has also been recorded in literature: "Some patients are asymptomatic despite significant radiographic abnormalities: others undergo spontaneous remission and do not require treatment".²

We agree with the writers that response of patients of PAP to WLL is variable; and the same has been mentioned in the case report in discussion.^{1,3}

There was no intention of the authors to understate the role of radiology in diagnosis and management of both these cases. Rather we very much appreciate the role of radiology in both diagnosis and management of PAP. Actually management of PAP is a multispecialty team approach with pulmonologists, radiologists, pathologists, and anaesthesiologists (for single lung ventilation during WLL).

On behalf of all the authors, I again thank the writers for their critical evaluation and discussion of both the cases and for publishing the serial HRCT scan chest findings of case 2.

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1. Bhattacharyya D, Barthwal MS, Katoch CDS, et al. Primary alveolar proteinosis – a report of two cases. *MJAFI*. 2013;69: 90–93.