Case report

Sarcoidosis as unusual cause of massive pleural effusion

Sharad Joshi*, Pallavi Periwal, Vikas Dogra, Deepak Talwar

Department of Pulmonary & Critical Care Medicine, Metro Centre for Respiratory Diseases, Metro Multispeciality Hospital, L-94, Sector-XI, Noida, U.P., 201301, India

A R T I C L E  I N F O

Article history:
Received 12 September 2015
Accepted 24 September 2015

Keywords:
Sarcoidosis
Effusion
Thoracoscopy

A B S T R A C T

Sarcoidosis is a multisystem granulomatous disease of unknown etiology. Pleural involvement is relatively rare. Development of pleural effusion in sarcoidosis needs to be evaluated for other causes, especially tuberculosis in endemic countries. Sarcoid pleural effusion responds to systemic corticosteroids. We are presenting case of 42 year old male patient of sarcoidosis who developed massive pleural effusion while on treatment with steroids, which was attributed to disease per se. Sarcoidosis as a cause of massive pleural effusion has not been mentioned before in published literature.

© 2015 The Authors. Published by Elsevier Ltd. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 42 year old male, non-smoker, with no history of diabetes or hypertension, presented with 10 days history of shortness of breath which increased since 1 day. He was diagnosed as a case of sarcoidosis 2 months back based on clinical and radiologic findings when he had presented with history of intermittent fever, shortness of breath, loss of appetite and weight-loss for 2 months. Chest X Ray (CXR) done at that time showed diffuse nodular shadows (Fig. 1A). CT chest demonstrated diffuse micronodules with predilection for fissures and bronchovascular bundles and associated mediastinal lymphadenopathy (Fig. 1B). MRI abdomen revealed multiple subcentimetric focal lesions diffusely scattered in liver, spleen and renal parenchyma. Serum angiotensin converting enzyme (ACE) was raised (126 U/L). Quantiferon Gold for tuberculosis was negative. Rheumatoid factor and anti nuclear anti-

In view of acute increase in breathlessness for last one day, pulmonary embolism was suspected. CT pulmonary angiography was done which showed no evidence of embolus. Pleuroscopy revealed multiple large nodules over parietal and visceral surfaces (Fig. 3A and B). Their biopsy demonstrated extensive inflammation with well-formed epithelioid granulomas without necrosis. Tuberculosis cultures from biopsy, pleural fluid and bronchoalveolar fluid were negative. 3.5 L of fluid was drained over a period of 24 h. To confirm the diagnosis of sarcoidosis, patient underwent further investigative work up. On fibreoptic bronchoscopy mucous in left bronchial tree was grossly oedematous and irregular (Fig. 4A). Endobronchial biopsy showed non-caseating epithelioid cell granulomas with langhans cells (Fig. 4B). Transbronchial lung biopsy revealed nonspecific inflammation of interstitium with ill formed epithelioid granulomas (Fig. 4C). A diagnosis of sarcoidosis with massive pleural effusion was made.

For treatment Prednisolone was continued and Hydroxychloroquine was added. 2 months later effusion resolved completely and he remains asymptomatic.

1. Discussion

Pleural involvement is rare in sarcoidosis. In a review of published studies about pleural-sarcoidosis it may manifest as pleural effusion, pneumothorax, pleural thickening, hydro-pneumothorax, trapped lung, hemothorax, or chylothorax[1]. However, effusion of sarcoid pleural effusion, pneumothorax, pleural thickening, hydro-pneumothorax, trapped lung, hemothorax, or chylothorax [1]. However, effusion continues to be a rare manifestation in all published series of sarcoidosis to the extent that it's frequently considered to exclude its diagnosis.

A low prevalence was further reported by Huggins and...
colleagues who described 2.8% (5/181) prevalence of pleural effusions defined by thoracic ultrasonography in 181 consecutive outpatients with sarcoidosis where it was related to sarcoidosis in only 1.1% (2/181) of patients [2]. They concluded that pleural effusion in patients with sarcoidosis should not be assumed to be related to sarcoidosis.

The mechanism of pleural effusion formation in sarcoidosis is presumably,

a) Involvement of the pleura which leads to increased capillary permeability
b) Superior vena cava obstruction [3]

c) Endobronchial sarcoidosis leading to bronchial stenosis and lobar atelecatsis [4]
d) Trapped lung [5]
e) Lymphatic disruption with the development of chylothorax [6].

Szwarcberg and colleagues published their findings in a series of 61 patients with sarcoidosis using thoracic CT scans. The authors noted that 25 (41%) of the 61 patients had “pleural involvement” detected on CT. Of the 25 patients with pleural involvement, 20 (80%) had pleural thickening and five (20%) patients had pleural effusions; [8.2% (5/61)]. But, cause of pleural disease in this cohort of sarcoidosis patients was not determined [7].
In a case report published previously, authors retrospectively analyzed clinical data obtained from 624 sarcoid patients, the only patient who showed pleural effusion and histologically proven sarcoidosis of the pleura was the case they described, making frequency of sarcoid pleural effusion to be 0.16% [8].

In an isolated description of a case of bilateral massive pleural effusion secondary to sarcoid authors mentioned growth of one colony of *Mycobacterium tuberculosis* on culture from the lesion in their patient [9].

Pleural fluid characteristics are similar in sarcoidosis and tuberculosis, both being exudative, lymphocytic and is usually considered tuberculous in endemic areas for tuberculosis [10]. A definitive diagnosis of sarcoid pleural effusion relies on a pleural biopsy demonstrating non-necrotizing granulomas, with the exclusion of granulomatous diseases of known etiology by microbiological evaluation. In the majority biopsy is necessary to rule out malignancy or other infections that can mimic sarcoid-pleural effusion.

Oral steroids are the primary treatment while in some cases spontaneous resolution has also been reported for sarcoid pleural effusions. Development of pleural effusion during steroid therapy points more towards infections secondary to usual or opportunistic organisms.

In a patient with multisystem sarcoidosis, responding well on steroid treatment; development of massive effusion secondary to sarcoidosis per se has not been reported in medical literature so far, to the best of our knowledge.

2. Conclusion

In an endemic country, tuberculosis is a frequent cause for effusion in patients on steroid treatment for sarcoidosis. Hence, pleural effusion in all sarcoidosis patients must be appropriately confirmed with pleural biopsies and culture studies to distinguish between pleural involvement due to sarcoid or tuberculosis, and medical thoracoscopic pleural biopsy is an important and safe intervention to ascertain diagnosis. Also, emphasized in this case is development of massive pleural effusion secondary to sarcoidosis while on oral steroid therapy.

Supplementary video related to this article can be found online at http://dx.doi.org/10.1016/j.rmcr.2015.09.011.

References

[1] N.T. Soskel, O.P. Sharma, Pleural involvement in sarcoidosis: case presentation and detailed review of the literature, Curr. Opin. Pulm. Med. 6 (2000) 455–468.
[2] J.T. Huggins, P. Doelken, S.A. Sahn, et al., Pleural effusions in a series of 181 outpatients with sarcoidosis, Chest 129 (2006) 1599–1604.
[3] J. Gordonson, S. Trachtenberg, E.N. Sargent, Superior vena cava obstruction due to sarcoidosis, Chest 63 (1973) 292–293.
[4] R.H. Poe, Middle-lobe atelectasis, N. Y. State J. Med. 78 (1978) 2895–2897.
[5] J.T. Heidecker, M.A. Judson, Pleural effusion caused by a trapped lung, South Med. J. 96 (2003) 510–511.
[6] H. Aberg, M. Bah, A.W. Waters, Sarcoidosis complicated by chylothorax, Minn. Med. 57 (1966) 1065–1070.
[7] J.B. Szwarzberg, N. Glajchen, A.S. Teirstein, Pleural involvement in chronic sarcoidosis detected by thoracic CT scanning, Sarcoidosis Vasc. Diffuse Lung Dis. 22 (2005) 58–62.
[8] A. Tommasini, G. Di Vittorio, F. Facchinetti, et al., Pleural effusion in sarcoidosis: a case report, Sarcoidosis 11 (2) (1994) 138–140.
[9] P. Balasubramanian, J. Mathew, R. Cherian, O.C. Abraham, Bilateral massive pleural effusion-A rare presentation of sarcoidosis, J. Postgrad. Med. 5 (4) (2005) 335–336.
[10] S. Akcay, V. Pinelli, G.P. Marchetti, G.F. Tassi, The diagnosis of sarcoidosis pleurisy by medical thoracoscopy: report of three cases, Tuberk. Toraks 56 (4) (2008) 429–433.