Radiology Case Reports 13 (2018) 867–870

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Acute fibrinous and organizing pneumonia: Imaging features, pathologic correlation, and brief literature review☆☆

Ji Yeon Kim[a], *Kyung Won Doo*[b,*], *Hang-Jea Jang*[c]

[a] *Department of Pathology, Inje University College of Medicine, Haeundae Paik Hospital, Haeundae-ro 875, Haeundae-gu, Busan 48108, Republic of Korea*

[b] *Department of Radiology, Inje University College of Medicine, Haeundae Paik Hospital, Haeundae-ro 875, Haeundae-gu, Busan 48108, Republic of Korea*

[c] *Department of Internal Medicine, Division of Pulmonology and Critical Care Medicine, Inje University College of Medicine, Haeundae Paik Hospital, Haeundae-ro 875, Haeundae-gu, Busan 48108, Republic of Korea*

ARTICLE INFO

Article history:
Received 31 August 2017
Revised 20 April 2018
Accepted 21 April 2018

Keywords:
Computed tomography
Acute fibrinous and organizing pneumonia
Idiopathic interstitial pneumonias

ABSTRACT

Acute fibrinous and organizing pneumonia is a histopathologic entity of lung injury with the hallmark feature of intra-alveolar fibrin "balls." We report 2 cases of acute fibrinous and organizing pneumonia in patients without a significant medical history, who presented with cough and worsening dyspnea and experienced a fulminant course of disease progression with diffuse lung parenchymal abnormalities on chest computed tomography. These cases suggest that this rare histologic pattern of idiopathic interstitial pneumonias can be included in the differential diagnosis with other conditions leading to acute respiratory failure.

1. Introduction

Acute fibrinous and organizing pneumonia (AFOP) has been described as a histopathologic entity of lung injury with the hallmark features of intra-alveolar fibrin deposits within the alveolar spaces. It may represent a part of a spectrum of diffuse alveolar damage (DAD) and organizing pneumonia (OP) associated with a similar acute or subacute clinical presentation, but histologically does not meet the criteria for either DAD or OP [1].

There have been few reports primarily addressing imaging of in AFOP [2]. We describe radiological and pathologic findings in 2 cases of AFOP, which are included among the rare histologic patterns of interstitial lung disease (ILD).

☆ Competing Interests: The authors have declared that no competing interests exist.

* Corresponding author.
E-mail address: kwdoo@inje.ac.kr (K.W. Doo).

https://doi.org/10.1016/j.radcr.2018.04.028

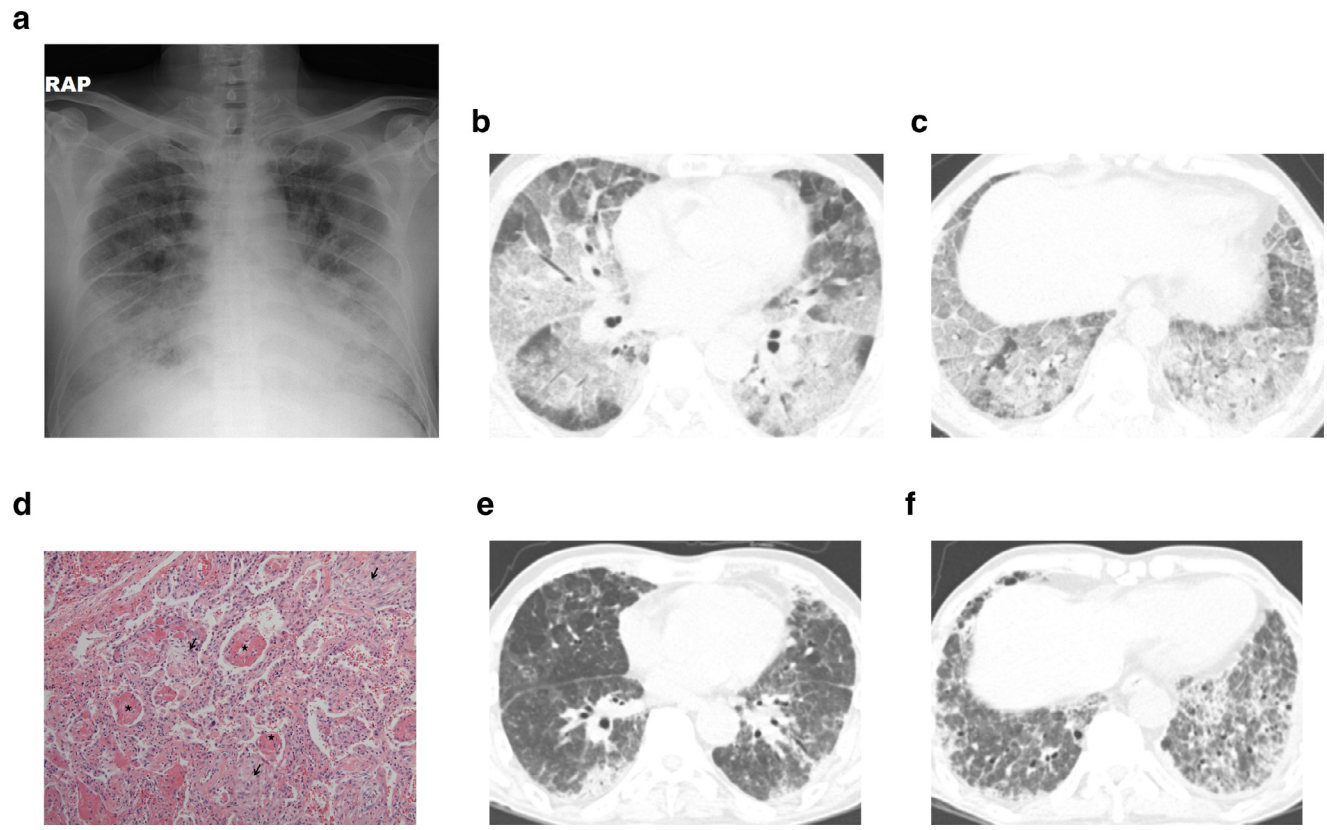

Fig. 1 – Acute fibrinous and organizing pneumonia in a 63-year-old man with dyspnea and cough. (a) Chest radiograph shows bilateral diffuse basal opacities. (b, c) Computed tomography scan shows bilateral geographic and patchy areas of ground glass opacities with interlobular septal thickening predominant in basal portions in the lungs. (d) Photomicrograph reveals fibrin balls (asterisks) in the airspaces alternating with organizing fibroblastic tissue (arrows; hematoxylin and eosin stain, ×200). (e, f) Axial computed tomography images obtained 10 weeks after the initial study demonstrate clearing of diffuse opacities, with residual peribronchial fibrosis and distortion.

2. Case report

2.1. *Case 1*

A 63-year-old man presented to the emergency room due to worsening of dyspnea, with a 5-day history of cough and myalgia. He had no significant medical history. Initial laboratory investigations revealed elevated white blood cell count $17.14 \times 10^3/\mu L$ with neutrophil predominance (82.9%).

Initial chest radiography revealed bilateral diffuse basal opacities (Fig. 1a). Chest computed tomography (CT) was performed and demonstrated bilateral geographic and patchy areas of ground glass opacities (GGO) with interlobular septal thickening predominant in the basal portions (Fig. 1b and c). Findings of "crazy paving appearance" are observed in various diseases, and reflect a combination of airspace exudates, interstitial edema, inflammation, and alveolar collapse that usually exhibit a pattern of DAD. Therefore, primary radiological differential diagnoses include pulmonary edema, acute respiratory distress syndrome, or *Pneumocystis jiroveci* pneumonia. The patient was administered empirical therapy for community-acquired pneumonia. Despite initial treatment, his respiratory status continued to decline and required intubation. On admission day 7, he underwent wedge biopsies of the left lower lobe through video-assisted thoracoscopic surgery (VATS).

On histologic examination, the alveolar spaces were filled with fibrin balls and organizing fibroblastic tissue (Fig. 1d). The alveolar septa were thickened by moderate chronic inflammatory cell infiltration and organized loose connective tissue. Hyaline membranes were not detected and significant eosinophil presence was not observed, thereby excluding the diagnoses of DAD and eosinophilic pneumonia. Special stain (Grocott methenamine silver) for fungal infection was negative, essentially excluding the possibility of *P jiroveci* pneumonia. Based on the characteristic intra-alveolar fibrin balls and associated OP, a diagnosis of AFOP was made.

Intravenous corticosteroid (methylprednisolone, 60 mg/day) was administered, and his clinical symptoms and chest radiography findings improved. At day 7 of the steroid treatment, opacity in both lungs decreased, with concomitant improvement in his respiratory symptoms and oxygen saturation. After 10 weeks, follow-up CT revealed clearing of diffuse opacities, with residual peribronchial fibrosis and distortion (Fig. 1e and f). After 1 year with steroid

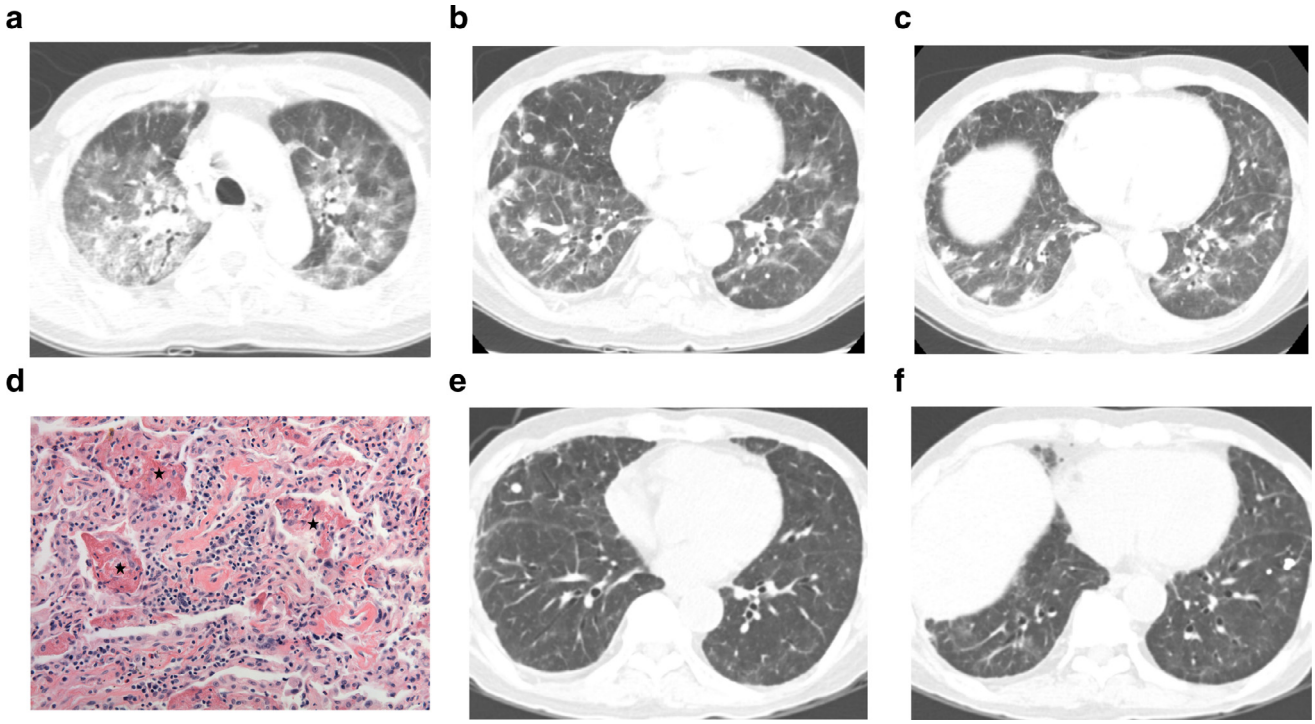

Fig. 2 – Acute fibrinous and organizing pneumonia in a 75-year-old man with dyspnea, cough, and fever. (a-c) Computed tomography scan reveals ground glass opacities, consolidation with patchy and peribronchial distribution, and multifocal ill-defined nodular opacities in both the lungs. (d) On high-power microscopic view, intra-alveolar fibrin balls (asterisks) are noted (hematoxylin and eosin stain, ×400). (e, f) Follow-up computed tomography after 3 weeks shows markedly decreased extent of bilateral ground glass opacities and nodular infiltrations.

tapering, respiratory symptoms relapsed; consequently he was followed-up as an outpatient with administration of oral steroid. He died of pneumonia 31 months after the diagnosis of AFOP.

2.2. *Case 2*

A 75-year-old man was admitted with a 5-day history of worsening dyspnea. He complained of fever, cough, sputum, headache, and dizziness. He had history of variant angina, pulmonary tuberculosis, and had been treated for diabetes mellitus. Initial laboratory investigations revealed an elevated white blood cell count ($15.96 \times 10^3/\mu L$) with neutrophil predominance (90.1%).

Chest radiography revealed bilateral diffuse and patchy opacities. CT revealed bilateral GGO and consolidation predominantly in the upper lungs, with patchy and peribronchial distribution, and multifocal, ill-defined small nodules (Fig. 2a–c). On admission day 4, his respiratory status and blood pressure rapidly declined and required intubation. He underwent video-assisted thoracoscopic surgery-wedge biopsies of the right middle lobe and right lower lobe.

Histologically, the dominant finding was intra-alveolar fibrin deposition in the form of fibrin balls (Fig. 2d). The remaining lung parenchyma exhibited organized loose connective tissue within the alveolar spaces. Sparse eosinophils were noted, but an eosinophilic pneumonia pattern was not evident. These histologic findings were most consistent with AFOP.

He underwent intravenous corticosteroid therapy (methylprednisolone, 60 mg /day). At day 8 of steroid treatment, parenchymal opacities on chest radiography decreased with concomitant improvement in his respiratory symptoms. Follow-up CT after 3 weeks revealed markedly decreased extent of bilateral GGO and nodular infiltrations (Fig. 2e and f). During follow-up as an outpatient with administration of oral steroid, his respiratory symptoms relapsed and improved several times. He died of pneumonia 33 months after the diagnosis of AFOP.

3. Discussion

The American Thoracic Society/European Respiratory Society (ATS/ERS) classification of idiopathic interstitial pneumonias (IIPs), revised in 2013, described a category of rare IIPs and rare histologic patterns of interstitial lung disease, including AFOP and a group of bronchiolocentric patterns [3,4].

AFOP is a histopathologic diagnosis that was initially described in 2002 by Beasley et al. in a case series involving 17 patients [1]. The dominant histopathologic features are intra-alveolar fibrin deposition in the form of fibrin “balls” within the alveolar spaces, with type II pneumocyte hyperplasia and OP, typically in a patchy distribution. The AFOP pattern differs

histologically from acute lung injury patterns of DAD or OP, and also differs from eosinophilic pneumonia. It differs from DAD in that the fibrin, which is organized into balls within the alveolar spaces, is typically patchy, and classic hyaline membranes are not evident. Therefore, AFOP may represent a histologic pattern that can occur in the clinical spectrum of DAD and OP, or may reflect a tissue sampling issue [1].

AFOP can either be idiopathic or associated with a variety of disease states including infections, collagen vascular diseases, adverse drug or chemicals reactions, hematological malignancy, altered immune status, and inhalation diseases [5–11]. Our patients did not appear to be associated with any known significant exposures or previous medical diseases.

The prognosis of AFOP appears to have 2 distinct patterns of disease progression: one is an acute and fulminant course leading to respiratory failure with rapid progression to death; and the other is a subacute, less fulminating course with recovery. Overall, AFOP exhibits poor prognosis, with up to 50% mortality [1]; the clinical course and radiological findings mirror one another. Although variable radiographic findings have been described in several cases [11], there have been few reports primarily addressing imaging in AFOP. Patients who experience a rapidly progressive course exhibit imaging findings similar to DAD, with diffuse but basilar-predominant consolidation and GGO. Those with a more subacute course may have similar radiological findings of cryptogenic OP, and can exhibit both focal and diffuse parenchymal abnormalities. Both cases of AFOP in our institution presented with a rapidly progressive clinical course leading to respiratory failure. One exhibited bilateral geographic and patchy areas of GGO, with interlobular septal thickening predominant in basal portions, and findings of "crazy paving appearance." These findings give rise to a differential diagnosis of radiological DAD pattern. The other patient exhibited bilateral GGO, predominantly in the upper lungs, with patchy and peribronchial distribution and multifocal ill-defined small nodules identified at presentation, which represented a combined manifestation of OP and DAD.

We encountered 2 cases of AFOP that presented with rapid progression of dyspnea and radiological findings. Recognition of this pattern is important to avert potentially adverse clinical outcome(s). These 2 cases emphasize the importance of being aware of this uncommon pattern of lung injury and to consider AFOP in the differential when encountering diffuse, basilar-predominant consolidation and GGO suggesting DAD or multifocal consolidation along bronchovascular bundles, and ill-defined nodular infiltration similar to OP on chest imaging. Although the imaging findings are nonspecific, our cases suggest that AFOP can be included in the differential diagnosis with other conditions leading to respiratory failure in patients with a fulminant clinical course. This information is valuable to clinicians and radiologists who are unfamiliar with these rare histologic patterns of IIP.

REFERENCES

[1] Beasley MB, Franks TJ, Galvin JR, Gochuico B, Travis WD. Acute fibrinous and organizing pneumonia: a histological pattern of lung injury and possible variant of diffuse alveolar damage. Arch Pathol Lab Med 2002;126(9):1064–70.
[2] Garcia BA, Goede T, Mohammed TL. Acute fibrinous organizing pneumonia: a case report and literature review. Curr Prob Diagn Radiol 2015;44(5):469–71.
[3] Travis WD, Costabel U, Hansell DM, King TE, Lynch DA Jr, Nicholson AG, et al. An official American Thoracic Society/European Respiratory Society statement: update of the international multidisciplinary classification of the idiopathic interstitial pneumonias. Am J Respir Crit Care Med 2013;188(6):733–48.
[4] Johkoh T, Fukuoka J, Tanaka T. Rare idiopathic intestinal pneumonias (IIPs) and histologic patterns in new ATS/ERS multidisciplinary classification of the IIPs. Eur J Radiol 2015;84(3):542–6.
[5] Prahalad S, Bohnsack JF, Maloney CG, Leslie KO. Fatal acute fibrinous and organizing pneumonia in a child with juvenile dermatomyositis. J Pediatr 2005;146(2):289–92.
[6] Balduin R, Giacometti C, Saccarola L, Marulli G, Rea F, Bartoli M, et al. Acute fibrinous and organizing pneumonia in a patient with collagen vascular disease "stigma". Sarcoidosis Vasc Diffuse Lung Dis 2007;24(1):78–80.
[7] Heo JY, Song JY, Noh JY, Yong HS, Cheong HJ, Kim WJ. Acute fibrinous and organizing pneumonia in a patient with HIV infection and Pneumocystis jiroveci pneumonia. Respirology 2010;15(8):1259–61.
[8] Yokogawa N, Alcid DV. Acute fibrinous and organizing pneumonia as a rare presentation of abacavir hypersensitivity reaction. AIDS 2007;21(15):2116–17.
[9] Lee SM, Park JJ, Sung SH, Kim Y, Lee KE, Mun YC, et al. Acute fibrinous and organizing pneumonia following hematopoietic stem cell transplantation. Korean J Internal Med 2009;24(2):156–9.
[10] Otto C, Huzly D, Kemna L, Huttel A, Benk C, Rieg S, et al. Acute fibrinous and organizing pneumonia associated with influenza A/H1N1 pneumonia after lung transplantation. BMC Pulm Med 2013;13:30.
[11] Kligerman SJ, Franks TJ, Galvin JR. From the radiologic pathology archives: organization and fibrosis as a response to lung injury in diffuse alveolar damage, organizing pneumonia, and acute fibrinous and organizing pneumonia. Radiographics 2013;33(7):1951–75.