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Case Report

Acute Fibrinous and Organizing Pneumonia with Excellent Response to Steroid Therapy

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Abstract: Acute Fibrinous and Organizing Pneumonia (AFOP) is a diffuse infiltrative pulmonary disease characterized by the presence of intra alveolar fibrin and organized pneumonia. It's a rare and relatively unknown "entity" with only a few cases described. Herein, we describe the association of acute fibrinous and organizing pneumonia with excellent response to steroid therapy. We suggest that, such an association has been very rarely reported. A32 year's old Sudanese female was presented to us in ELmek Nimer university hospital complaining of fever, cough and shortness of breath for 10 days and received a full course of antibiotics without any improvement, her condition deteriorated. The initial chest radiography showed well defined inhomogeneous pacification in the upper lobe of the left lung with no air bronchogram, Computed Tomography of the chest showed a rather well defined Lt Upper lobe mass lesion closely related to the arch of the aorta. Ultrasound guided biopsy of the lung showed a histological diagnosis of acute fibrinous and organizing pneumonia. After establishing the diagnosis of fibrinous pneumonia the patient received oral steroid (prednisone 1mg/kg) for 10 days then tapering. The patient showed rapid improvement and excellent response and discharged in good condition. Chest radiography after one month showed complete resolution of the radiological finding in her previous imaging. We recommend that AFOP should be considered in the differentials of a suspected pulmonary infection unresponsive to optimal antibiotic "therapy". AFOP may present as pulmonary mass on chest X-ray and diagnosed made with biopsy and histological examination. We conclude that AFOP may be treated with "steroids" therapy.

Keywords: Acute Fibrinous and Organizing Pneumonia, Chest X-Ray, And Steroid

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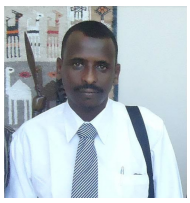

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1. INTRODUCTION

Acute fibrinous and organizing pneumonia is a rare diffuse pulmonary disease which recognized as pattern of acute lung injury, but it is not yet known whether it is a distinct form of interstitial pneumonia or simply a reflection of a tissue sampling issue. Histological patterns are characterized predominantly by presence of intra alveolar "fibrin" in the form of "fibrin" balls within patchy distribution and organizing "pneumonia". Acute Fibrinous and Organizing Pneumonia (AFOP) was first described by Beasely et al. in 2002 as a distinct pattern of lung "injury" with the histological analogy to diffuse alveolar damage, organizing pneumonia and eosinophilic pneumonia[1,2]. The features that distinguish it histologically include intra alveolar "fibrin" balls, the absence of typical hyaline membranes and eosinophils, and numerous foci of fibroblastic activity [1, 2] . Since then, there have been essentially isolated reports describing diverse causes and clinical courses [3-6]. Consequently, whether AFOP is a distinct pattern of interstitial pneumonia or whether it simply reflects a tissue sampling issue remains to be elucidated [2].It has been described in all age groups with numerous associations including connective tissue and autoimmune diseases, drugs, occupational and environmental exposures, and less commonly infectious agents. Occasionally, no cause has been found [2]. The signs and symptoms are variable; two forms of the illness have been described; a severe form which leads to rapid respiratory failure and a sub-acute form, which has a good prognosis with treatment [1.] Diagnosis, is by histological analysis of a biopsy obtained from the affected pulmonary tissue [1,2,7-9] . There are no standard guidelines on treatment; steroids and immunosuppressant have been used with varying success rates, with best results in the sub-acute form of the disease. Mechanical ventilation is often necessary for severe disease and carries poor prognosis. [1, 3, 5] We present the case of a young Sudanese female patient who presented to us in ELmek Nimer university hospital complaining of cough, and her signs, radiological features suggested community acquired pneumonia on underlying tumor not responding adequately to full antibiotic course. A lung biopsy was thus performed which revealed AFOP. The disease was of the acute variety and responded well to steroids therapy. To the best of our knowledge this is the twenty fifth cases report on AFOP and the first to be reported from Sudan and Africa.

2. CASE REPORT

A 32 years old Sudanese female referred to us in ALmek Nimir university hospital with fever and cough. She had a similar condition before 10 days received two courses of antibiotic from a general practitioner without any response.

2.1 Physical examination

The patient was unwell not pale or jaundiced, BP 110/70 heart rate 96 b/min, respiratory rate 22/min, Oxygen saturation in room 98% and Temperature was 37^0 C. Lung auscultation revealed a fine inspiratory crackle in the left upper lung field. Other systems were normal.

2.2 Initial laboratory finding: Complete blood cell count showed

TWBCs 13*10 3 u/l, Hemoglobin concentration 12 g/dl, Platelet count 282 *10 3u\l the differential count showed neutrophil 67% lymphocyte 21% and monocyte 8%. Serum sodium, potassium, creatinine, calcium was all within normal limits.

2.3 Radiological finding

The initial chest radiography showed well defined inhomogeneous pacification in the upper lobe of the left lung with no air bronchogram. Figure No (1)

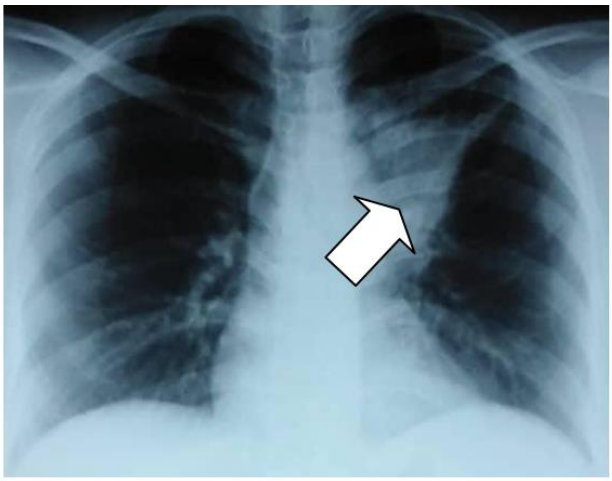

Fig 1: Chest Radiographs during the first admission

2.4 CT chest without contrast

Showed rather well defined Lt Upper lobe mass lesion closely related to the arch of the aorta. No air bronchogram seen within it. Figure No (2&3)

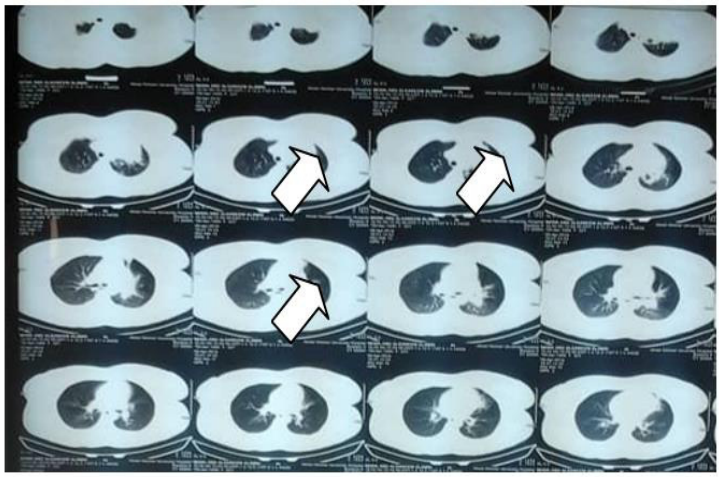

Fig 2: CT chest without contrast lung window

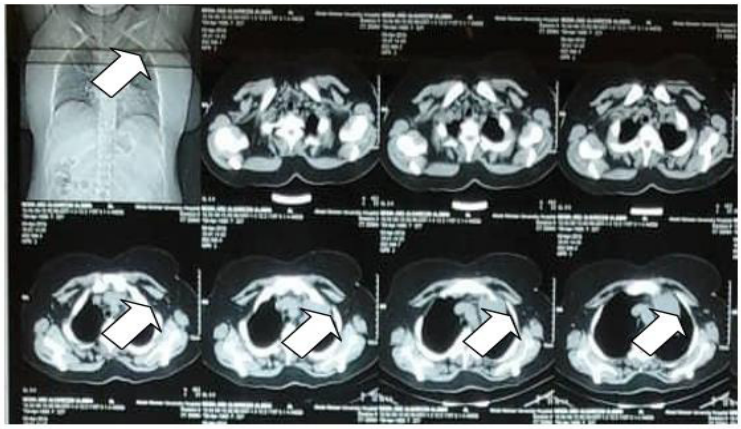

Fig 3: CT chest without contrast mediastinal window

2.5 CT chest with contrast

No enhancement on post contrast study. Figure No (4)

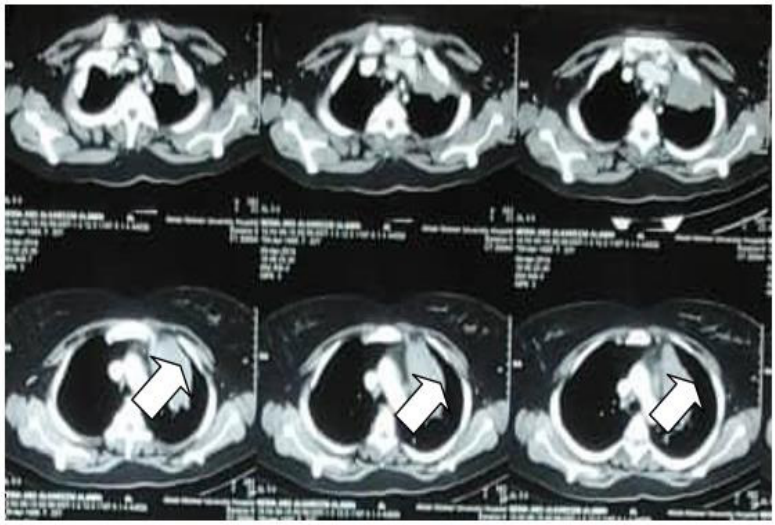

Fig 4: CT chest with contrast

2.6 Ultrasound guided biopsy

U\S guided biopsy to area of consultation in the left lung localized and biopsy was taken using size 16G true cut biopsy needle specimen sent for histopathology

2.7 Pathological findings

2.8 Macroscopic appearance

Multiple pieces of needle lung biopsy longest 1.5 cm

2.9 Microscopic appearance

Microscopy of needle biopsy of left lung shows widening of alveolar septa by mixed inflammatory infiltrate with massive fibrinous exudation with organization within alveolar spaces, and scattered neutrophilic infiltrates. Silver stains for pneumocystis carinii are negative. No evidence of malignancy in the section of adequate biopsy submitted. The features are those of fibrinous organization consolidation. Figure No (5)

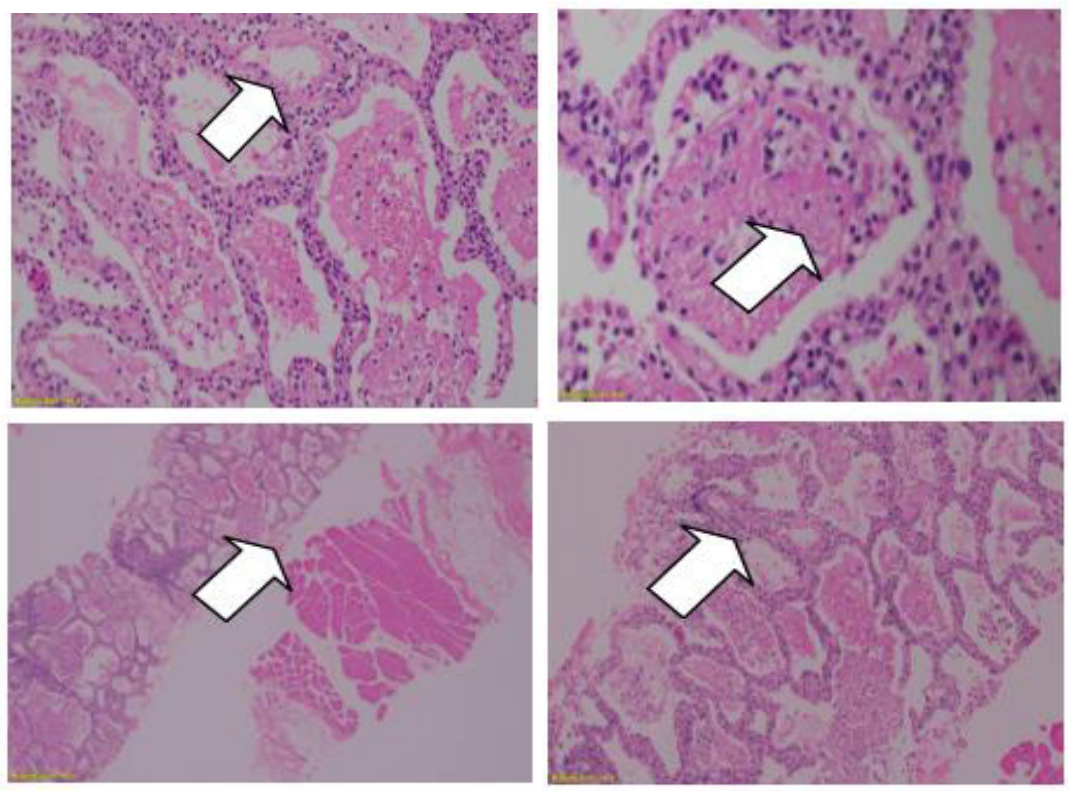

Fig 5: Histopathology of the lung biopsy shows widening of alveolar septa by a mixed inflammatory infiltrate with massive fibrinous exudation with organization within alveolar spaces, and scattered neutrophilic infiltrates

2.10 Hospital course

On the first hospital day, the patient received I.V third generation cephalosporin for 5 days; Paracetamol infusion per need without any improvement in her condition. Her condition was worse and became more tychaepnic and distressing. Then "we" put other diagnosis and sent for further investigations for lung biopsy to exclude lung tumors. Fortunately her histopathology was reported as fibrinous organized pneumonia. After establishing the diagnosis of fibrinous pneumonia the patient received oral steroid (prednisone 1mg/kg) and Proton pump inhibitor for 10 days then tapering. The patient showed rapid and excellent improvement and discharge in good condition. Chest radiography after one month showed complete resolution of the radiological finding with compared to previous studies done on the first day of admission Figure No (6).

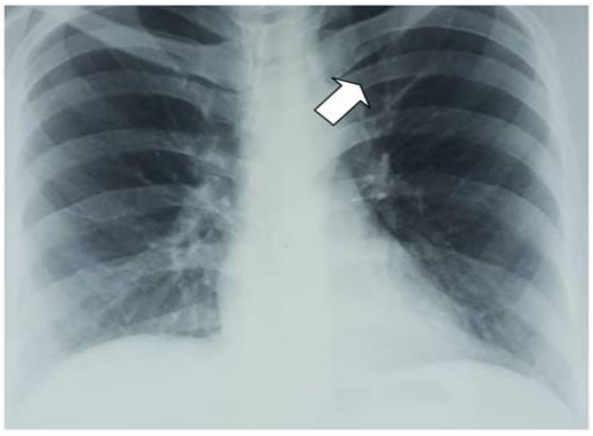

Fig 6: Chest Radiography after treatment with steroid

3. DISCUSSION

The AFOP is associated with several conditions such as rheumatologic disease. Infections, drug reactions, environmental exposure, connective tissue disease and lung transplantation [1,4,7-13] . This makes it difficult to identify clinical characters that are specific to AFOP. The histological pattern of AFOP is similar to that of diffuse alveolar damage, COP cryptogenic organizing pneumonia and eosinophilic pneumonia. However, it has its own distinct histopathology. The course of AFOP can be "acute" or subacute and clinical outcomes are directly related to the mode of onset. Acute and severe ill patients typically have poor prognosis [1, 6]. The main clinical manifestation is coughing respiratory distress and asthma like symptoms and chest tightness [1, 6] as our patient presented with symptoms and signs suggesting community acquired pneumonia. The most common radiological findings are diffuse patchy "opacities" with both peripheral and bilateral distribution and the lesion may be limited to the lung "base" the image may appear similar to those in other lung disease like interstitial pneumonia ,pulmonary edema and infectious pneumonia[1,14] .The initial chest radiography of our patient showed well defined homogeneous pacification in the upper lobe of the left lung with no air bronchogram. Definitive diagnoses of AFOP require histopathological evaluation. Tissue histopathology is characterized by intra alveolar fibrin in balls and organizing pneumonia with patch distribution. The optimal treatment of AFOP is remained controversial. Kuzet summarized 111 cases reported in the "literature" they found that glucocorticoid were the most common and successful treatment modality the therapeutic effect of drugs may be associated with onset of disease. Most patients with acute onset "died" however the patients with subacute response well to glucocorticoids and had a good prognosis[15, 16]. Our patient exhibits an excellent response to glucocorticoids and does not need a mechanical ventilator.

4. CONCLUSION

From this case , we recommend that AFOP is a rare clinical pattern of lung injury and should be considered as a differential diagnosis in patients with a suspected pulmonary infection unresponsive to standard antibiotic therapy. AFOP may present as pulmonary mass on chest X-ray and diagnosed made with biopsy and histological examination of the obtained tissue. Treatment is with corticosteroid and the response is dramatic with good compliance.

5. AUTHORS CONTRIBUTION STATEMENT

MIA consultant physician was involved in making the decisions regarding the patient's treatment while in the intensive care unit. He was the primary consultant physician of the patient. He reviewed the manuscript critically and approved it for final submission. MHE was the resident registrar of medicine who took care of the patient. She was involved in planning of various investigations for the patient and performed the literature review, drafted the manuscript, reviewed it critically and approved it the end for submission. She was also involved in retrieving high quality images for publication from the pathology and radiology archives. Both authors read and approved the final manuscript.

6. CONFLICT OF INTEREST

Conflict of interest declared none.

7. REFERENCES

1. 1Beasley 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. Doi: 10.5858/2002-126-1064-AFAOP, PMID 12204055.
2. 2Tzouvelekis A, Koutsopoulos A, Oikonomou A, Froudarakis M, Zarogoulidis P, Steiropoulos P, Et Al. Acute Fibrinous And Organising Pneumonia: A Case Report And Review Of The Literature. J Med Case Rep. 2009;3(1):74. Doi: 10.1186/1752-1947-3-74.
3. 3Damas C, Morais A, Moura CS, Marques A. Acute Fibrinous And Organizing Pneumonia. Rev Port Pneumol. 2006;12(5):615-20. Doi: 10.1016/S0873-2159(15)30453-0, PMID 17117329.
4. 4Balduin 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. PMID 18069424.
5. 5Prahalad 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. Doi: 10.1016/J.Jpeds.2004.09.023, PMID 15689928.
6. 6López-Cuenca S, Morales-García S, Martín-Hita A, Frutos-Vivar F, Fernández-Segoviano P, Esteban A. Severe Acute Respiratory Failure Secondary To Acute Fibrinous And Organizing Pneumonia Requiring Mechanical Ventilation: A Case Report And Literature Review. Respir Care. 2012;57(8):1337-41. Doi: 10.4187/Respcare.01452, PMID 22348347.
7. 7Yokogawa N, Alcid DV. Acute Fibrinous And Organizing Pneumonia As A Rare Presentation Of Abacavir Hypersensitivity Reaction. AIDS. 2007;21(15):2116-7. Doi: 10.1097/QAD.0b013e3282f08c5a, PMID 17885309.
8. 8Müller-Wening D, Koschel D, Stark W, Sennekamp HJ. Humidifier-Associated Disease In The General Population. Dtsch Med Wochenschr. 2006;131(10):491-6. Doi: 10.1055/S-2006-932548, PMID 16511738.
9. 9Bhatti S, Hakeem A, Torrealba J, Mcmahon JP, Meyer KC. Severe Acute Fibrinous And Organizing Pneumonia (AFOP) Causing Ventilatory Failure: Successful Treatment With Mycophenolate Mofetil And Corticosteroids. Respir Med. 2009;103(11):1764-7. Doi: 10.1016/J.Rmed.2009.07.009, PMID 19666216.
10. 10Cho JY, Lee HK, Lee SS, Lee HK, Lee YM, Lee HP, Et Al. A Case Of Acute Fibrinous And Organizing Pneumonia. Tuberc Respir Dis. 2006;61(5):479-83.7. Doi: 10.4046/Trd.2006.61.5.479 Rapaka V. Rapaka V, Hussain MA, Niazi M, Diaz-Fuentes G. Severe Acute Fibrinous And Organizing Pneumonia Causing Acute Respiratory Distress Syndrome And Shock. J Bronchology Interv Pulmonol. 2011;18(3):269-73. Doi: 10.1097/LBR.0b013e318222a4f2, PMID 23208573.
11. 11Otto C, Huzly D, Kemna L, Hüttel 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. Doi: 10.1186/1471-2466-13-30. PMID 23683442.
12. 12Valim V, Rocha RH, Couto RB, Paixão TS, Serrano EV. Acute Fibrinous And Organizing Pneumonia And Undifferentiated Connective Tissue Disease: A Case Report. Case Rep Rheumatol. 2012;2012:549298. Doi: 10.1155/2012/549298, PMID 22957292.
13. 13Hariri LP, Unizony S, Stone J, Mino-Kenudson M, Sharma A, Matsubara O, Et Al. Acute Fibrinous And Organizing Pneumonia In Systemic Lupus Erythematosus: A Case Report And Review Of The Literature. Pathol Int. 2010;60(11):755-9. Doi: 10.1111/J.1440-1827.2010.02586.X, PMID 20946526.
14. 14Sauter JL, Butnor KJ. Expanding The Spectrum Of Pulmonary Histopathological Manifestations Of Anti-Synthetase Syndrome: Anti-EJ-Associated Acute Fibrinous And Organizing Pneumonia. Histopathology. 2014 [Epub Ahead Of Print];65(4):581-2. Doi: 10.1111/His.12420. 12. Guimarães C, Sanches I, Ferreira C. Acute Fibrinous And Organising Pneumonia. BMJ Case Rep. 2012;2012. Doi: 10.1136/Bcr.01.2011.3689, PMID 22605688.
15. 15Bhatti S, Hakeem A, Torrealba J, Et Al. Severe Acute Fibrinous And Organizing Pneumonia (AFOP) Causing Ventilator Failure: Successful Treatment With Mycophenolate Mofetil And Corticosteroids. Respir Med. 2009;103(1764)_7.
16. López-Cuenca S, Morales-García S, Martín-Hita A, Frutos-Vivar F, Fernández-Segoviano P, Esteban A. Severe Acute Respiratory Failure Secondary To Acute Fibrinous And Organizing Pneumonia Requiring Mechanical Ventilation: A Case Report And Literature Review. Respir Care. 2012;57(8):1337-41. Doi: 10.4187/Respcare.01452, PMID 22348347.