LETTER TO EDITOR (VIEWERS CHOICE)

SPONTANEOUS PNEUMOTHORAX: A RARE COMPLICATION OF ANA NEGATIVE PEDIATRIC SYSTEMIC LUPUS ERYTHEMATOUS

Sudhir Mehta, Jyoti Sanghvi, Ravindra Kumar

Keywords: ANA negative, Pneumothorax, Systemic Lupus Erythematous

Systemic Lupus Erythematous (SLE) is a chronic multisystemic disease and a number of pulmonary complications including pleuritis, infectious pneumonia, pulmonary hemorrhage, pulmonary hypertension and pneumothorax are known to occur in these patients. (1). Of these well known complications, spontaneous pneumothorax is rare to occur in pediatric patients. (2) An 8 years old girl, suffering from SLE presented with cough for 2 days, sudden onset right sided chest pain and breathlessness. Her chest X ray before onset of breathlessness was normal and she was on oral antibiotics for past 2 days. She was diagnosed to have SLE one year ago, when she presented with symptoms of low grade fever, joint pains, pallor, oral ulceration, photosensitivity without involvement of lung. ANA was negative but anti dsDNA done by immunofluorescence was positive. She met 6 out of 11, ACR (American college of Rheumatology) revised classification criteria for SLE. (2) In view of diffuse proliferative glomerulonephritis on renal biopsy, she was started on NIH (National institute of health) protocol of monthly methyl prednisolone and cyclophosphamide pulse therapy for management of SLE nephritis (3) and was on regular follow up. On current admission, she was afebrile, but had respiratory distress (respiratory rate – 48/min). Examination of chest revealed decreased air entry on right side. Chest X-ray showed right sided pneumothorax with collapse of underlying lung. Immediate chest tube insertion and pneumothorax evacuation was carried out. There was no evidence of any underlying chest infection and underlying lung functions were also normal. Gradually her lungs expanded and chest tube was also removed. She was kept on regular follow up and she never had recurrence of pneumothorax.

Although pulmonary involvement in SLE has rarely been systematically investigated in children, pulmonary involvement is reported to be 5 to 67% in few studies. (4) All parts of pulmonary system can be involved, but out of all these complications, spontaneous pneumothorax has not been reported in pediatric SLE in literature. A number of putative mechanisms have been presented. It has been proposed that underlying pneumonitis predisposes to pneumothorax either by formation of macroscopic blebs or by causing microscopic involvement of pulmonary parenchyma in form of infarction. (5) In general glucocorticoids have antagonistic effect on collagen deposition during wound healing, resulting in tissue fragility. (6) On the other hand, it has been reported that few patients with various systemic disorders, who are treated with cyclophosphamide develop late onset lung disease complicated by pneumothorax. But this was not the case with our patient, who had received only two cycles of cyclophosphamide therapy prior to pneumothorax. Review of literature identified total 11 cases of SLE complicated by pneumothorax in adults of which 10 had evidence of underlying parenchymal lesions including 9 who had evidence of pleurisy. Ten out of 11 patients had received glucocorticoid therapy for at least 3 weeks before the occurrence of pneumothorax. (7) Literature review revealed primarily four factors contributing to pneumothorax that include underlying immune process of disease, lung infections, role of glucocorticoids and cyclophosphamide therapy. Tissue fragility caused by these might contribute to pneumothorax in these patients. Therefore in this case, disease process itself contributed to the formation of pneumothorax probably potentiated by the glucocorticoid and cyclophosphamide therapy.

Funding: None

Conflict of Interest: None

References :

1. Haupt HM, Moore GW, Hutchins GM. The lung in systemic lupus erythematosus. Analysis of the pathologic changes in 120 patients. Am J Med 1981;71:791–98.
2. Delgado EA, Malleson PN, Pirie GE, Petty RE. The pulmonary manifestations of childhood onset systemic lupus erythematosus. Semin Arthritis Rheum 1990;19:285–93
3. Gourley MF, Austin HA 3rd, Scott D, Yarboro CH, Vaughan EM, Muir J, et al. Methyl prednisolone alone or in combination in Lupus Nephritis. A randomized controlled trial . Ann Intern Med 1996;125:549-57.
4. Caeiro FM, Michielson FMC, Bernstein R, Hughes GR, Ansell BM. Systemic lupus erythematosus in childhood. Ann Rheum Dis 1981;40:325–31.
5. Wilhelm M, Van Why SK. Pneumothoraces complicating systemic lupus erythematous with nephritis. Pediatr Nephrol 2002;17:261-63
6. Wicke C, Halliday B, Allen D, Roche NS, Scheuenstuhl H, Spencer MM, et al. Effects of steroids and retenoids on wound healing. Arch Surg 2000;135:1265-70.
7. Tanaka N, Kusunoki Y, Kaneko K, Yamamoto T, Kaburaki M, Muraoka S, et al. Systemic lupus erythematous complicated by recurrent pneumothorax: case report and literature review. J Clin immunol 2010;33: 162-68.

From: Department of Pediatrics, Sri Aurobindo medical college and PG Institute, Indore, Madhya Pradesh, India.

Address for Correspondence: Dr. Sudhir Mehta, Senior Resident, DCH, DNB Pediatrics, Department of Pediatrics, Sri Aurobindo medical college and PG Institute, Indore, Madhya Pradesh, India.

Email: samsudhir123@yahoo.co.in

DOI: 10.7199/ped.oncall.2015.21