J HK Coll Radiol. 2010;13:29-31

CASE REPORT

Segmental Pulmonary Artery Dissection in Systemic Lupus Erythematosus

DD Rasalkar, BK Paunipagar, WCW Chu

Department of Diagnostic Radiology and Organ Imaging, The Chinese University of Hong Kong, Prince of Wales Hospital, Shatin, Hong Kong.

ABSTRACT

We report a unique case of segmental pulmonary artery dissection in a 48-year-old female, with confirmed systemic lupus erythematosus and pulmonary hypertension. Several cardiopulmonary manifestations of systemic lupus erythematosus have been described in literature and the association with pulmonary hypertension is well known. Among the 7 radiologically reported cases of pulmonary artery dissection, only one had segmental pulmonary artery dissection, and to our knowledge no case of pulmonary artery dissection has been reported in systemic lupus erythematosus.

Key Words: *Aneurysm, dissecting; Hypertension, pulmonary; Lupus erythematosus, systemic; Pulmonary heart disease; Tomography, X-ray computed*

中文摘要

系統性紅斑狼瘡症患者的肺動脈段支夾層剝離

DD Rasalkar、K Paunipagar、朱昭穎

本文報道一例罕見的肺動脈段支夾層剝離。患者為系統性紅斑狼瘡症確診伴肺動脈高血壓的48歲女性。文獻報道過幾類由系統性紅斑狼瘡症誘發的心肺病變，而此症跟肺動脈高血壓有關亦是已知的事實。七例有關肺動脈夾層剝離的影像學報告中，只有一例是段支夾層剝離。據我們所知，並未有系統性紅斑狼瘡伴發肺動脈夾層剝離的病例報道。

CASE REPORT

A 48-year-old woman, known to have systemic lupus erythematosus (SLE) with secondary pulmonary hypertension, presented to the emergency department with shortness of breath, palpitations, and associated constricting central chest pain. Physical examination revealed an increased but regular pulse rate and blood pressure of 140/70 mm Hg. An electrocardiogram showed sinus rhythm and biventricular hypertrophy. On auscultation, the chest was clear and there was a tricuspid regurgitation heart murmur. D-dimer levels were elevated about 1156 ng/ml (reference level, <500 ng/ml). Chest X-ray revealed mild cardiomegaly with an enlarged pulmonary trunk and a small right pleural effusion (Figure 1). Using the smart-prep technique, a contrast-enhanced pulmonary angiogram was performed to exclude pulmonary thromboembolism. The contrast-enhanced computed tomographic (CT) images revealed an intimal dissection originating in the lobar pulmonary artery of the right lower lobe, and extending to its

Correspondence: *Dr Darshana D Rasalkar, Department of Diagnostic Radiology and Organ Imaging, The Chinese University of Hong Kong, Prince of Wales Hospital, Shatin, Hong Kong.*
Tel: (852) 2632 2290; Fax: (852) 2636 0012; Email: darshana@cuhk.edu.hk

Submitted: 30 Oct 2009; Accepted: 1 Feb 2010.

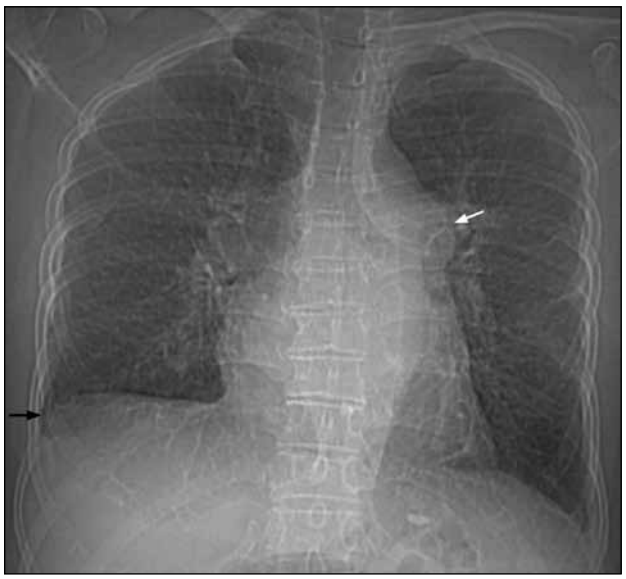

Figure 1. Frontal chest radiograph reveals mild cardiac enlargement and prominent left pulmonary trunk (white arrow). Note the small right pleural effusion (black arrow).

distal segmental branches (Figure 2). No pulmonary thromboembolism was identified. The patient received vasodilators and warfarin and is clinically stable at present.

DISCUSSION

SLE is an autoimmune disorder with multisystem involvement. Among the cardiac manifestations, the most frequently observed complications are pericarditis and or pericardial effusion (39%), arterial hypertension (22%), ischaemic heart disease (16%), myocarditis (14%), congestive heart failure (10%), pulmonary hypertension (9%), valvular heart disease (9%), pleural effusion (7%), and cerebrovascular accident (3%).[1]

Pulmonary arterial dissection is an extremely rare but usually lethal complication of chronic pulmonary hypertension. It often manifests as cardiogenic shock or sudden death and is therefore typically diagnosed at necropsy rather than during life. Sudden death occurs most commonly due to cardiac tamponade as the vessel dissects into the pericardium. Recent isolated reports, however, have described pulmonary artery dissection in surviving patients.[2] In that series, chest pain occurred in 67%, dyspnoea in 82%, and central cyanosis in 52%. Of 7 cases previously described, one had segmental pulmonary artery dissection[3] and the remainder showed involvement of the main pulmonary trunk. Chronic pulmonary arterial hypertension is the likely major cause of pulmonary artery dissection; other rare causes include chronic inflammation of the pulmonary arteries, right heart endocarditis, amyloidosis, trauma, and severe atherosclerosis.[4]

Imaging methods used to detect pulmonary artery dissection include transthoracic echocardiogram,[5,6] CT,[5] magnetic resonance imaging,[7] and pulmonary angiography.[8] Multidetector CT is the first-line investigation if pulmonary embolism is suspected[9] as in our case. Moreover, CT provides accurate information of an

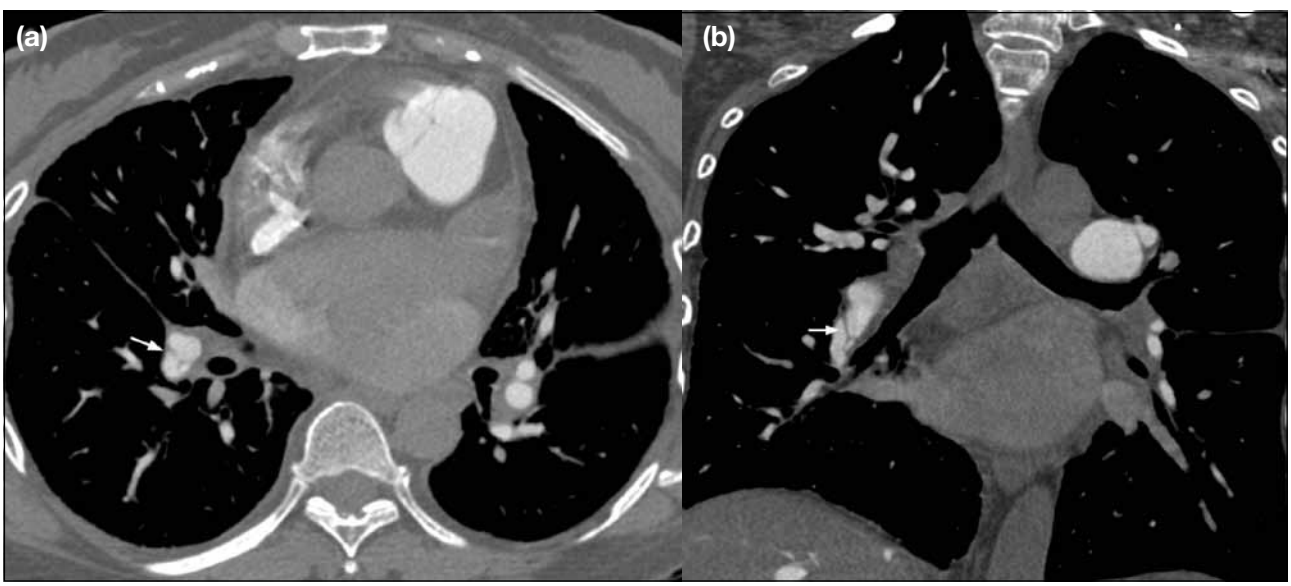

Figure 2. Computed tomographic thorax, pulmonary angiogram, (a) axial and (b) coronal. Note the thin intimal flap in the right lower segmental branch (white arrows) consistent with segmental pulmonary artery dissection.

intimal flap, the extent of dissection, and the presence of intraluminal thrombi. It can also demonstrate related complications such as haemopericardium secondary to pulmonary artery dissection into the pericardium as well as cardiac tamponade.[10] None of these complications nor pulmonary embolism were identified in our patient, who therefore probably survived with conservative medical treatment alone.

Although it is well known that SLE can have almost any potential effect on vessels anywhere in the body, segmental arterial dissection is extremely rare. To the best of our knowledge, no case of segmental pulmonary artery dissection has been reported in association with SLE in the literature.

REFERENCES

1. Badui E, Garcia-Rubi D, Robles E, et al. Cardiovascular manifestations in systemic lupus erythematosus. Prospective study of 100 patients. Angiology. 1985;36:431-41.
2. Khattar RS, Fox DJ, Alty JE, Arora A. Pulmonary artery dissection: an emerging cardiovascular complication in surviving patients with chronic pulmonary hypertension. Heart. 2005;91:142-5.
3. Hoye SL, Gale CP, Tolan DJ, Pepper CB. An unusual presentation of pulmonary artery dissection. Thorax. 2009;64:368.
4. Wunderbaldinger P, Bernhard C, Uffmann M, Kürkciyan I, Senbaklavaci O, Herold CJ. Acute pulmonary trunk dissection in a patient with primary pulmonary hypertension. J Comput Assist Tomogr. 2000;24:92-5.
5. Tønder N, Køber L, Hassager C. Pulmonary artery dissection in a patient with Eisenmenger syndrome treated with heart and lung transplantation. Eur J Echocardiogr. 2004;5:228-30.
6. Areco D, Pizzano N. Pulmonary artery dissection: echocardiographic findings and diagnosis. Echocardiography. 2003;20:375-7.
7. Stern EJ, Graham C, Gamsu G, Golden JA, Higgins CB. Pulmonary artery dissection: MR findings. J Comput Assist Tomogr. 1992;16:481-3.
8. Steurer J, Jenni R, Medici TC, Vollrath T, Hess OM, Siegenthaler W. Dissecting aneurysm of the pulmonary artery with pulmonary hypertension. Am Rev Respir Dis. 1990;142:1219-21.
9. Neimatallah MA, Hassan W, Moursi M, Al Kadhi Y. CT findings of pulmonary artery dissection. Br J Radiol. 2007;80:e61-3.
10. Song EK, Kolecki P. A case of pulmonary artery dissection diagnosed in the Emergency Department. J Emerg Med. 2002;23:155-9.