Case Report

Detecting jellyfish sign in a patient with systemic lupus erythematosus: An added value of the echocardiography

Marha M. Menaisy

From Menaisy, MBBch, Department of Cardiology, Alexandria Main University Hospital, Alexandria, Egypt

ABSTRACT

Systemic lupus erythematosus (SLE) is an autoimmune disease that affects nearly every organ of the body. Cardiopulmonary manifestations are common in SLE, almost in the form of serositis, and include pleural and pericardial effusions. In these patients, echocardiography is an excellent non-invasive tool for detecting pericardial effusion and may provide a hint about pleural effusion. We report the case of a 20-year-old woman with SLE who had been experiencing dyspnea for 1 month and had no prior history of cardiovascular or pulmonary disease. A rheumatologist referred her to our echocardiography lab for a cardiac evaluation. In the echocardiographic examination, pleural mass depended from the left visceral pleura was discovered, surrounded by massive pleural effusion that induced left lung atelectasis (Jellyfish sign, and complete atelectasis of a lung lobe floats above a massive pleural effusion).

Key words: Echocardiography, Jellyfish sign, Pleural effusion, Systemic lupus erythematosus

Systemic lupus erythematosus (SLE) is an autoimmune disease affecting almost every organ of the body. Its prevalence is estimated to be around 15–50 per 100,000 individuals with a female predilection [1]. Cardiopulmonary manifestations are common in SLE, often in the form of serositis, and include pleural and pericardial effusion [2]. Pleuropulmonary manifestations occur in nearly half of SLE patients throughout the disease and may present in 4–5% of SLE patients. Pleuritis with or without pleural effusion is the most prevalent sign of acute lung involvement in SLE and these are the most prevalent complications directly related to the condition [3]. Cardiac abnormalities are common in lupus patients even if they are clinically asymptomatic. Pericarditis is the most common clinical presentation of heart involvement, with a 20–30% frequency, while examination rubs are less prevalent [4]. It is usually associated with small effusions but it can lead to more significant effusions when uremia is present.

Echocardiography is an excellent non-invasive tool for detecting pericardial effusion and may provide a hint about the presence of pleural effusion, which can be confirmed by chest imaging studies such as radiography and computed tomography (CT) scanning [5]. As a result, it is used to evaluate all patients with cardiopulmonary symptoms such as dyspnea, tachypnea, and chest pain.

Access this article online	
Received - 24 June 2021 Initial Review - 09 July 2021 Accepted - 16 July 2021	Quick Response code
DOI: 10.32677/IJCR.2021.v07.i08.2968	

In this case, we would like to highlight the importance of familiarizing ourselves with the basics of pulmonary echocardiography, as these findings are prevalent during the performance of echocardiography in our daily practice.

CASE REPORT

A 20-year-old female with SLE was suffering from dyspnea for 1 month. The patient had no history of cardiovascular or pulmonary disease. On general examination, the patient was fully conscious with a distended abdomen and bilateral lower limb edema. Auscultation of the chest revealed decreased air entry on the left side and heart auscultation revealed well heard S1 and S2 with no adventitious sounds. The patient was vitally stable with a blood pressure of 120/70 mmHg, a heart rate of 70 beats/min, and oxygen saturation of 95%.

Her rheumatologist referred her to our Echo lab for cardiac evaluation. Echocardiographic examination shows good left ventricular systolic function, no valvular abnormalities, and no pericardial effusion. There was no evidence of pulmonary hypertension but there was an extracardiac (pleural mass) derived from the left visceral pleura which was surrounded by a massive pleural effusion causing left lung atelectasis (Fig. 1). Following the echocardiographic findings, a computed tomography (CT) scan was performed revealing a significant left-sided pleural effusion with near-total lung collapse and a mediastinal shift to the right side (Fig. 2).

Correspondence to: Marha M. Menaisy, Department of Cardiology, Alexandria Main University Hospital, Alexandria, Egypt. E-mail: menecymareha@yahoo.com

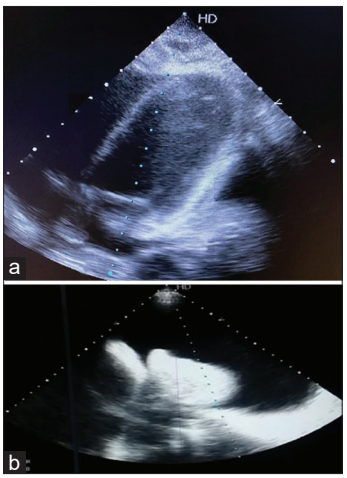

Figure 1: Trans-thoracic echocardiogram (TTE) (a) apical four chamber view showing an extra cardiac mass depended from the left visceral pleura surrounded by massive pleural effusion. (b) Modified apical four chamber view with lateral and pendular modulation for better focus on the mass

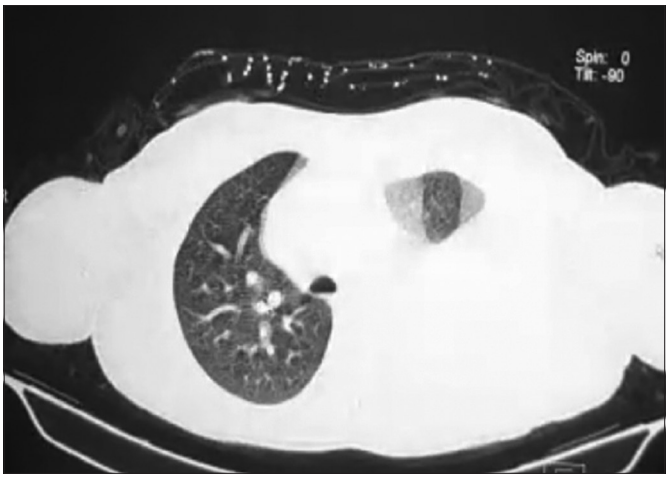

Figure 2: CT chest showing marked left-sided pleural effusion with near total lung collapse and mediastinal shift to the right side

Pathological examination of the pleural fluid obtained through thoracocentesis revealed reactive mesothelial cells and few lymphocytes and neutrophils in an eosinophilic proteinaceous background. The laboratory examination of the pleural fluid was also performed, which includes microscopic, physical, and biochemical analyses. The microscopic analysis revealed neutrophils 80 cell/mm^3, lymphocytes 105 cell/mm^3, RBCs 1,750 cell/mm^3, the physical analysis revealed clear yellow fluid free of deposits, and the chemical analysis revealed proteins 1.7g/dL, glucose 114mg/dL, and lactate dehydrogenase (LDH) 44U/L. The use of multi-imaging modalities followed by a pathological examination confirmed that an extracardiac mass depended from the left visceral pleura was an atelectatic lung floating above a massive pleural effusion which is known as the Jellyfish sign.

After the diagnosis is confirmed, the patient was referred to an interventional radiologist who could insert an ultrasound-guided percutaneous pigtail catheter to drain pleural fluid. It is easy, safe, and successful, and it is less traumatic than traditional chest tube installation. After 2 weeks of recurrence, fluid accumulations were successfully drained as confirmed by a follow-up chest X-ray. Following that, pleurodesis was performed to provide the best palliation.

DISCUSSION

Serositis can be a significant cause of morbidity. Lupus serositis symptoms range from pleuritic pain and pericardial friction murmur to the potentially fatal consequences of massive pleural effusion and congestive heart failure.

The diagnosis of lupus-associated serositis is made by excluding malignant, thromboembolic, and transudation causes such as hypoalbuminemia and heart failure. Identifying a subgroup of patients with SLE who are at higher risk of serositis provides the opportunity for early intervention. The risk factors for serositis are the risk factors of serositis are active SLE (serositis was found to be significantly more common in active SLE than in inactive SLE), a high D-dimer level (this finding indicates the role of hyper-coagulability as a mechanism in the pathogenesis of serositis in SLE), and fever (the precise mechanism is unknown) [6].

Echocardiography can help to distinguish between pericardial and pleural effusions. Because the descending thoracic aorta is interposed between the pericardium anteriorly and pleura posteriorly. Echocardiography may be a helpful landmark in the (a) in the parasternal long axis and apical 4-chamber views, the pericardial effusion tracks anteriorly to the descending aorta; (b) pericardial effusion is always present in the dependent segments (posterior wall, lateral wall, and inferior wall); and (c) in the parasternal long-axis and apical 4-chamber views, a left-sided pleural effusion appears posterior and lateral to the descending aorta [7]. However, due to the abnormal position of the effusion, it may be difficult to distinguish between pericardial and pleural effusion. Another drawback of echocardiography is that it cannot determine the nature of the mass, necessitating the use of multi-imaging modalities followed by a pathological examination to confirm the diagnosis and guide treatment, as in our case.

Similar to our case, Yéboles *et al*. reported a case of a 21-month-old child who underwent arthrotomy due to infectious arthritis. During the procedure, the child displayed remarkable desaturation with high oxygen input and difficulty breathing with obvious hypercapnia in the operating room. Because of these factors, the postoperative follow-up was performed in a pediatric intensive care unit. Following a lung examination that revealed hypoventilation, they decided to perform a lung ultrasound scan, which revealed left pleural effusion with the jellyfish sign [7]. Izaguirre *et al*. also reported a similar case of a 71-year-old man with lymphoma and a complaint of dyspnea for 6 months. The Jellyfish sign was discovered during the echocardiographic examination [8].

CONCLUSION

Not all echocardiographic findings are related to heart disease. We would like to emphasize the importance of becoming acquainted

with the fundamentals of pulmonary echocardiography, as these findings are common during echocardiography performance in our daily practice. The use of lung echocardiography (with cardiac probes) has recently been developed as a new approach. Extravascular lung fluid assessment is a challenging task for clinical cardiologists and an elusive goal for echocardiographers. With lung ultrasonography, echocardiography can also determine the most common problematic differential diagnosis of pleural effusion (pericardial effusion). Transthoracic echocardiography is usually used to rule out the diagnosis.

REFERENCES

1. Cheo SW, Low QJ. Massive pericardial effusion-an uncommon initial presentation of systemic lupus erythematosus (SLE). Med J Malaysia 2019;74:439-40.
2. Tselios K, Urowitz MB. Systemic lupus erythematosus. In: The Heart in Rheumatic, Autoimmune and Inflammatory Diseases. United States: Academic Press; 2017. p. 235-67.
3. Torre O, Harari S. Pleural and pulmonary involvement in systemic lupus erythematosus. La Presse Méd 2011;40:e41-51.
4. Dein E, Douglas H, Petri M, Law G, Timlin H. Pericarditis in lupus. Cureus 2019;11:4166.
5. Shazzad MN, Islam MN, Ara R, Ahmed CM, Fatema N, Azad AK, *et al*. Echocardiographic assessment of cardiac involvement in systemic lupus erythematosus patients. Mymensingh Med J 2013;22:736-41.
6. Liang Y, Leng RX, Pan HF, Ye DQ. The prevalence and risk factors for serositis in patients with systemic lupus erythematosus: A cross-sectional study. Rheumatol Int 2017;37:305-11.
7. Yéboles R, Arroyo-Marin MJ, Jaraba-Caballero S, Gómez-Guzman E, Frías-Pérez M, Ruiz-Sáez B, *et al*. A pleural ultrasound image of a collapsed lung surrounded by pleural fluid ("jellyfish sign") may correspond to an intrapericardial mass. J Ultrasound 2021;1-4.
8. Izaguirre L, Prat C, Alzaga U, Urizar C, Alcalde C, Bajeneta A, *et al*. P1735 Jellyfish sign, not all echocardiographic findings are related to heart disease. Eur Heart J Cardiovasc Imaging 2020;21:jez319-1096.

Funding: None; Conflicts of Interest: None Stated.

How to cite this article: Menaisy MM. Detecting Jellyfish Sign in a Patient with Systemic Lupus Erythematosus: An Added Value of the Echocardiography. Indian J Case Reports. 2021;7(8):336-338.