

Cardiac Tamponade as a Rare Manifestation of Systemic Lupus Erythematosus: A Report on Four Cases in the Philippine General Hospital

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Abstract

Synopsis: Cardiac tamponade among systemic lupus erythematosus (SLE) patients is an unusual event. The pericardial effusion may be a consequence of uremia, infections in the pericardium, or the lupus pericarditis itself. We present four atypical cases of cardiac tamponade from pericarditis of connective tissue disease (CTD), all of which were treated with drainage and immunosuppressants. Due to the rarity of this combination, management was a challenge.

Clinical Presentation: Four females each sought consult for dyspnea associated with typical manifestations of connective tissue disease such as arthritis, characteristic rashes, serositis, typical laboratory features, and a positive ANA and/or anti-dsDNA. The first three cases fulfilled the criteria for SLE, while the fourth fulfilled the criteria for SLE-dermatomyositis overlap syndrome. Echocardiography was done due to suspicion of pericardial involvement and revealed massive pericardial effusion in tamponade physiology in all cases.

Diagnosis: Cardiac tamponade from serositis due to connective tissue disease (SLE (case 1 to 3) or SLE-dermatomyositis overlap (case 4). Other common etiologies of tamponade such as bacterial, tuberculous, malignant, and uremic pericardial effusion were ruled out by clinical and laboratory tools, including Gram stain and culture, cytology, PCR, and biochemical testing. The pericardial fluid of the first case tested positive for lupus erythematosus (LE) cells, indicative of lupus serositis.

Treatment and Outcome: All patients underwent pericardial drainage via tube pericardiostomy. They received high dose glucocorticoids after infectious etiologies for the pericardial effusion were ruled out. The fourth case with the overlap syndrome, however, required more immunosuppressants using azathioprine and methotrexate. Resolution of pericardial effusion was noted with this approach. Three of four were discharged improved, however, the third case suffered from worsening nephritis and pulmonary hemorrhage leading to her demise.

Significance and Recommendations: Four cases of cardiac tamponade as a manifestation of connective tissue disease were presented. Literature underlines the rarity of this condition anytime during the course of SLE. Despite this, SLE should be considered as one of the differential diagnosis of cardiac tamponade, especially in patients who manifest with multi-systemic findings. Likewise, massive pericardial effusion should be considered in patients with a connective tissue disease presenting with subtle evidence of pericardial involvement. It requires timely identification and treatment with high dose steroids, after other causes such as infections have been excluded. Immediate drainage through pericardiocentesis or pericardiostomy in combination with immunosuppressants may be life-saving.

Keywords: systemic lupus erythematosus, dermatomyositis, cardiac tamponade

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Introduction

The prevalence of systemic lupus erythematosus (SLE) is between 15 and 50 per 100,000 individuals, with a female preponderance of six to 10:1 in the age group 15 to 40 years. On the other hand, polymyositis and dermatomyositis are regarded as rare rheumatic diseases and the prevalence of overlap syndromes, especially mixed connective tissue disease, is unknown.¹ Pericardial involvement is more common in SLE, as compared to dermatomyositis/polymyositis; and cardiac tamponade is rare in all these diseases.²

We present four cases of SLE presenting as cardiac tamponade. In all cases, diagnosis of SLE was made

based on the American Rheumatism Association (ARA) criteria. Our fourth case also fulfilled a diagnosis of dermatomyositis (as an overlap syndrome) based on the Bohan and Peter criteria, fulfilling four of five (see appendix). Early recognition of this serious condition among lupus patients may be life-saving. Table I summarizes the highlights of the four cases.

4. Hypernasality - excessively nasal speech resonance, probably due to muscle weakness
5. Beck's Triad - jugular venous distention, hypotension, muffled heart sounds

Table I: Pertinent points in the four cases described

	Case 1	Case 2	Case 3	Case 4
General Data	32 year old, Filipina, Nulligravid	35 year old Filipina, G3P3	19 year old Filipina, Nulligravid	38 year old, Filipina, Nulligravid
Comorbidities	None	Post-partum	None	None
Diagnosis	Pericardial Effusion from SLE-Serositis Cardiac Tamponade Systemic Lupus Erythematosus			Pericardial Effusion from Serositis Cardiac Tamponade SLE-Dermatomyositis Overlap Autoimmune Thyroiditis
Criteria fulfilled for CTD	SLE: Serositis, anemia, LE cells, proteinuria, polyarthritis, ANA	SLE: Serositis, anemia, proteinuria, polyarthritis, malar rash, photosensitivity, ANA	SLE: Serositis, pancytopenia, polyarthritis, nephritis, ANA	SLE: Serositis, anemia, proteinuria, polyarthritis, ANA, anti-dsDNA, discoid Dermatomyositis: muscle weakness, muscle biopsy, EMG findings, skin findings (gottron's sign, heliotrope rash, mechanic's hand)
Chief Complaint	Dyspnea and chest heaviness	Dyspnea and edema	Dyspnea and fever	Proximal muscle weakness
Physical Examination	Neck vein engorgement Muffled heart sounds Normotensive, tachycardic Edema No pulsus paradoxus	Neck vein engorgement Distinct heart sounds Normotensive, tachycardic Edema With pulsus paradoxus	No neck vein engorgement Distinct heart sounds Normotensive, tachycardic Edema No pulsus paradoxus	No neck vein engorgement Distinct heart sounds Normotensive, tachycardic Edema No pulsus paradoxus
ECG	Sinus tachycardia, low voltage complex			
CXR	Cardiomegaly with Signs of Pericardial Effusion			
2D Echo	Massive pericardial effusion RV diastolic collapse	Massive pericardial effusion RA and RV diastolic collapse	Moderate pericardial effusion RV diastolic collapse	Massive pericardial effusion RA and RV diastolic collapse
Pericardial Fluid	(+) LE cells in pericardial fluid No organisms/ malignant cells AFB negative	No organisms / malignant cells AFB negative TB PCR negative	No organisms / malignant cells AFB negative	No organisms / malignant cells AFB negative TB PCR negative
Laboratory	(+) ANA Anemia, proteinuria	(+) ANA, Anti-dsDNA Anemia, proteinuria	(+) ANA Pancytopenia, proteinuria	(+) ANA, Anti-dsDNA Anemia, proteinuria Elevated TSH, thyroglobulin, TPO antibodies
Management	MPPT Prednisone Tube Pericardiostomy	High dose hydrocortisone Prednisone Tube Pericardiostomy	High dose hydrocortisone Tube Pericardiostomy	MPPT Prednisone Azathioprine and Methotrexate Tube Pericardiostomy
Outcome	Discharged	Discharged	Mortality	Discharged

MPPT: Methylprednisolone Pulse Therapy

ANA: Anti-nuclear Antibodies

Anti-dsDNA: Anti-double stranded DNA

Definition of Terms:

1. Gottron's Sign - erythema of knuckles with a raised violaceous scaly eruption
2. Mechanic's Hand - lateral and palmar areas of fingers may become rough and cracked, with irregular, dirty horizontal lines
3. Raynaud's Phenomenon - episodic digital ischemia, manifested by digital blanching, cyanosis and erythema of fingers & toes following cold exposure & subsequent

CASE 1: Systemic Lupus Erythematosus Presenting as Cardiac Tamponade

A 32-year-old nulligravid Filipina with no prior medical illness was admitted for a three month history of symmetric polyarthritis involving the metacarpals, proximal interphalangeal, distal interphalangeal, and ankle joints lasting from two to three days which would be relieved spontaneously. Over the next two months, she developed intermittent low-grade fever, malaise, myalgias, and anorexia. One month prior

to consult, she had tea-colored urine, facial edema, ascites, and oliguria. Two days prior to admission, she manifested with high-grade fever, cough, shortness of breath, easy fatigability, and chest pain. She denied photosensitivity, malar rash, oral ulcers, headache, or Raynaud's phenomenon although she had livedo reticularis.

Admission examination showed blood pressure of 130/90 mmHg, heart rate of 104 per minute and respiratory rate of 28 per minute. She was pale with neck vein engorgement slightly displaced apex

beat, muffled S1 and presence of an S3. There was ascites and bipedal edema. Pertinent laboratory work up revealed anemia, hypoalbuminemia, and a serum creatinine of 120 $\mu\text{mol/L}$. Chest radiograph showed cardiomegaly. Electrocardiogram showed sinus tachycardia and low voltage QRS complexes in all leads. Urinalysis showed +4 albuminuria and hematuria

Transthoracic echocardiography showed moderate global hypokinesia and moderate pericardial effusion with early diastolic right ventricular collapse, suggesting cardiac tamponade. She immediately underwent tube

Table II: Pertinent laboratory results

Variable	Reference Value	Case 1	Case 2	Case 3	Case 4
Complete Blood Count					
Hemoglobin	120-180g/L	100	93	70	118
WBC	4-11x10 ⁹ /L	7.00	3.98	1.57	7.44
Platelet	150-450x10 ⁹		197	41	70
Blood Chemistry					
Fasting Blood Sugar	< 125mg/dL	85	81	N/A	82
Creatinine	53-133 $\mu\text{mol/L}$	120	154	122	58
BUN	3.2-8.0mmol/L	7.91			4.80
Sodium	135-145mmol/L		134	140	136
Potassium	4-4.5mmol/L	4.0	4.1	3.3	3.9
Calcium	2.12-2.75mmol/L				1.92
Magnesium	0.70-1.05mmol/L				0.62
Albumin	38-51g/L	34		18	10
AST	0-34U/L			86	107
ALT	0-30U/L			29	49
Alkaline Phosphatase	36-92 $\mu\text{mol/L}$				259
CKMM	8-97U/L				56
CK Total	21-232U/L				123
LDH	100-190U/L				399
Thyroid Function Tests					
TSH	0.35-4.94 mIU/L	N/A	N/A	N/A	6.7
FT4	11-24 pmol/L				12.7
FT3	2.2-6.8 pg/L				3.5
Thyroid Antibodies					
Thyroglobulin Ab	< 4.1U/mL	N/A	N/A	N/A	3,500
TPO-Antibody	<5.6U/mL				1,900
Urine Studies					
Proteinuria (Dipstick)	None		+3	+3	+1
Hematuria (Dipstick)	None		+3	Innumerable	0
24 hour Total Protein	0-0.1 g/240	1.7	0.096/240		0.391
Pericardial Fluid Studies					
AFB and TB-PCR		Negative	Negative	Negative	Negative
GS/CS		No growth	No growth	No growth	No growth
RBC		3090	3650		21,200
WBC		1	12		470
Glucose		1.16 (↓)	6.78 (↑)		6.0 (↑)
Protein		26 (↓)	44 (↑)		43 (↑)
Cytology		No atypical cells	No atypical cells	No atypical cells	No atypical cells
Lupus Erythematosus Cells		Positive	N/A	N/A	N/A
Immunology					
ANA		+2 homogenous	+4 Speckled	+4 Homogenous	+4 Speckled
Anti-dsDNA		Negative	N/A	N/A	Positive
Arterial Blood Gases					
pH	7.35-7.45	N/A	7.431	7.49	7.441
PCO2	35-45mmHg		20	22	31
PO2	90-100mmHg		82	251	74
HCO3	22-28mHg		13	17	21

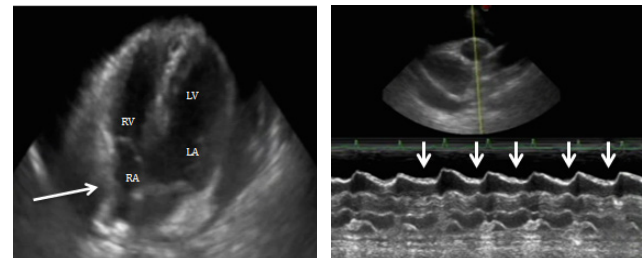
pericardiostomy, draining 150mL of straw-colored fluid, which on analysis showed 3,090 RBC/mm³ and 1.0 WBC/mm³. Gram stain and cultures of the pericardial fluid did not reveal any organisms. Pericardial fluid lupus erythematosus (LE) cell determination was positive. Laboratory results are summarized in Table II.

SLE was suspected on the basis of the serositis (pericardial effusion), history of arthritis, anemia, albuminuria, granular casts in the urine, and the presence of LE cell in the pericardial fluid. Antinuclear antibody (ANA) determination presented a +2 homogenous pattern by immunofluorescence. Detection of antibody to dsDNA using Crithidia lucillae immunofluorescence was negative. She was initially treated with prednisone at 1.0mg/kg/day for seven days. However, due to active nephritis (albuminuria of 1.7 g/24 hours and active urinary sediments,) and a repeat echocardiogram showing persistent depression of left ventricular systolic function interpreted to be myocarditis, methylprednisolone 500mg intravenously over four hours daily was given for three consecutive days. Subsequently, prednisone was resumed at a dose of 0.8mg/kg/day. Her pericardiostomy tube was removed after four days. Repeat echocardiogram showed improvement in the myocardial contractility and decrease in the amount of pericardial effusion with resolution of the right ventricular diastolic collapse. She was discharged improved after another week and prednisone was tapered off on follow up.

CASE 2: SLE Presenting as Myopericarditis, Complicated by a Cardiac Tamponade

A 35-year-old Filipina sought consult for dyspnea. Three years prior to consult, she was diagnosed with SLE when she presented with malar rash, polyarthritis, and pallor, supported by a positive ANA determination. She was maintained on prednisone, but was non-compliant. Two months prior to admission after giving birth to her third child, she noted worsening dyspnea on exertion and progressing bipedal edema. This prompted consult at the Philippine General Hospital (PGH). She was received with a blood pressure of 120/80 mmHg, tachycardic at 106 beats per minute, and tachypneic. She was afebrile. She had bibasal rales, distended neck veins, distinct heart sounds, and a regular rhythm. She had grade 2 bipedal edema, hepatomegaly, and minimal ascites. An electrocardiogram showed low voltage complexes. Echocardiogram at the emergency room showed massive pericardial effusion with a tamponade physiology (Figure 1). There were multisegmental wall motion abnormalities and a depressed systolic function with an ejection fraction of 36%, indicative of myocardial involvement (myocarditis). Other labs are in Table III. Immediate tube pericardiostomy was done yielding serosanguinous fluid. She was started

on high dose steroids after bacterial and tuberculous effusion were ruled out. On the seventh hospital day, a repeat 2D echo showed no recurrence of pericardial effusion. She was discharged on prednisone and heart failure medications.



A. Echocardiogram (four-chamber view) demonstrating right atrial collapse during diastole (arrow), indicative of tamponade physiology

B. Echocardiography (M-mode) demonstrating right ventricular free wall collapse (arrows) during diastole, also indicative of tamponade physiology

Figure 1: Transthoracic Echocardiogram of Case #2

CASE 3: Newly Diagnosed SLE Presenting as Myopericarditis, Complicated by a Cardiac Tamponade

A 19-year old nulligravid Filipina presented with dyspnea and undocumented febrile episodes. Three weeks prior to admission, she experienced undocumented febrile episodes, joint pains, and multiple vasculitic rashes on her lower extremities. Two weeks prior to admission, she complained of dyspnea on exertion, orthopnea, paroxysmal nocturnal dyspnea, and edema. She was admitted twice in a span of two weeks where multiple blood transfusions were made, allegedly due to low hemoglobin. Initial assessment was an acute leukemia, hence referral to PGH. On examination, she had a blood pressure of 100/60 mmHg, heart rate of 180 beats per minute, no neck vein engorgement, and bibasal rales. She had distinct heart sounds, an S3 gallop, and a slightly displaced apex beat laterally. She had alopecia, Raynaud's phenomenon (Figure 2), vasculitic lesions on her lower extremities, and bipedal edema. An electrocardiogram revealed sinus tachycardia, low voltage QRS complexes, and poor R-wave progression. Chest radiograph showed cardiomegaly, suggestive of pericardial effusion. A 2D echocardiogram was done revealing multisegmental wall motion abnormalities with a depressed systolic function (ejection fraction of 50%). She had moderate pericardial effusion and with right ventricular diastolic collapse, indicative of tamponade physiology. She had pancytopenia, +4 proteinuria on urinalysis, and a positive ANA (+4 homogenous). The assessment was a newly diagnosed SLE with myopericarditis in cardiac tamponade. She was in nephritic and hematologic flare. An immediate tube pericardiostomy was inserted, draining 500cc of serosanguinous fluid. Fluid did not grow any

bacterial organisms and was negative for Acid Fast Bacilli (AFB) stain. Other laboratory work up is in Table II. She was started on high dose hydrocortisone. Clinical improvement was noted on the fourth hospital day when she suddenly experienced dyspnea and refractory hypoxemia. Bloody secretions were noted upon intubation. She then went into refractory shock and renal failure. She arrested after a few hours. Cause of death was probably acute respiratory failure secondary to adult respiratory distress syndrome with a consideration of pulmonary hemorrhage from vasculitis.

seek a second opinion. About one month prior to consult, progressive dyspnea and muscle weakness made her bedridden. It was at a local hospital, where a repeat chest radiograph was normal, and the impression of systemic lupus erythematosus or kidney disease was considered. She was eventually referred to our institution and admitted.

On admission, the patient was conscious and oriented to person, place, and time. She was normotensive, tachycardic, tachypneic, with low



A. Cyanotic phase



B. Ischemic (Pallor) Phase



C. Hyperemic Phase

Figure 2: Raynaud's Phenomenon demonstrated in Case #3

CASE 4: SLE-Dermatomyositis Overlap Presenting with Cardiac Tamponade

A 38-year-old female, married, Filipina, was admitted for extreme muscle weakness. She had no other known prior comorbidities. She enjoyed good health as a nanny in Sweden, until four months prior to admission, when evanescent erythematous petechial lesions appeared on the extensor surfaces of both legs. Subsequently, she experienced generalized muscle pain, pains affecting the knees, elbows, and ankles, and proximal muscle weakness (described as difficulty in climbing stairs and combing her hair). She also experienced easy fatigability and grade 2 bipedal edema. A physician in Sweden assessed her to have autoimmune hypothyroidism. Other laboratory work up also revealed anemia and an elevated SGPT. She took prescribed but unrecalled medications with no relief of her weakness.

Three months prior to consult, her proximal muscle weakness worsened, now described as difficulty standing from a sitting position. However, she was still able to dress herself and to dorsiflex and plantarflex her ankles. At this time, she had difficulty swallowing liquids and solid food, dyspnea at rest, hypernasality, change in voice and weight loss from approximately 120 to 90 pounds within three months. Upon her return to the Philippines, she was admitted at a local hospital in Laguna. She developed erythematous plaques on the glabella, circumscribed lesions on her cheeks and forehead, and persistent hyperpigmentation over her finger joints. Chest x-ray was done allegedly showing pulmonary tuberculosis. She did not comply with anti-tuberculosis medications and decided to

grade fever. Facial lesions were a heliotrope rash, nonblanching erythematous patch over bridge of the nose, erythematous papules on cheeks and malar area, and a few discoid rashes on both lower cheeks (Figure 3). Other significant findings were masseter muscle atrophy, left cervical lymphadenopathy, erythema over the chest, and proximal muscle atrophy making her unable to move her extremities against gravity. She had decreased breath sounds on both lower lung fields with fine crackles. Heart sounds were distinct, tachycardic with regular rhythm, and no murmurs appreciated. Kussmauls sign and pulsus paradoxus were not appreciated. Moreover, there was note of Gottron's sign, mechanic's hand and Raynaud's phenomenon (Figure 4). She was unable to lift her arms against gravity and unable to abduct, adduct, internally and externally rotate her shoulders (muscle strength 2/5). She was also unable to stand up and lift her thighs against gravity (muscle strength 2/5).



A. Bluish Discoloration on the upper lids with note of edema (Heliotrope Rash). There is also note of papular erythematous rashes on glabella, cheeks, and malar area.



B. Discoid Rash near hairline

Figure 3: Characteristic Skin Lesions of Case #4

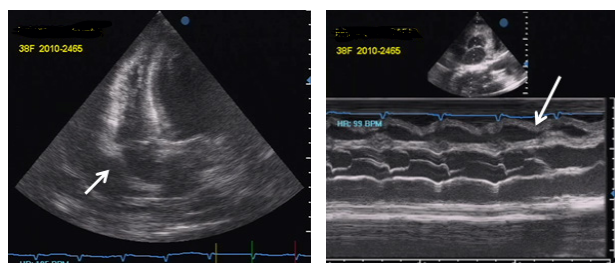


A. Erythematous to slightly hyperpigmented lesions on the knuckles (Gottron's Sign)

B. Rough and cracked palm with irregular, dirty horizontal lines (Mechanic's Hand)

Figure 4: Physical Examination Findings on Hands of Case #4

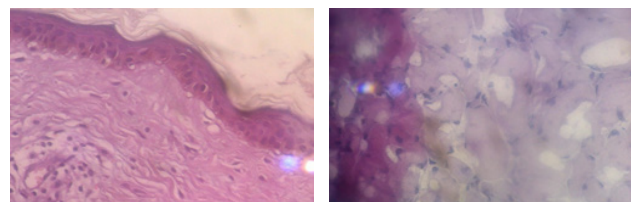
Laboratory test results are in Table II. Pertinent laboratory findings include anemia; positive ANA and anti-dsDNA; high AST and LDH; normal creatine kinase; elevated TSH, thyroglobulin and TPO antibodies; and normal FT4 and FT3. Chest radiograph showed cardiomegaly and bilateral pleural effusion. Sinus tachycardia and low voltage complexes were noted on 12-L ECG. Two dimensional echocardiogram on admission demonstrated a normal sized left ventricle with good wall motion and contractility and preserved overall systolic function (Figure 5). There was moderate pericardial effusion with right atrial and right ventricular diastolic collapse, consistent with cardiac tamponade. A holoabdominal ultrasound revealed minimal ascites. EMG-NCV revealed acute and chronic denervation changes with no volitional activity signifying diffuse muscle membrane irritability and axonal degeneration, consistent with inflammatory changes. Skin biopsy of an erythematous patch on her face showed findings suggestive of dermatomyositis, as seen in Figure 6. Muscle biopsy revealed a chronic myopathic process, as seen in Figure 7. The clinical impression was SLE-dermatomyositis overlap; pericardial effusion secondary to serositis, in cardiac tamponade, and autoimmune thyroiditis.



A. Echocardiogram (four-chamber view) demonstrating right atrial and ventricular collapse during diastole, indicative of tamponade physiology

B. Echocardiography (M-mode) demonstrating right ventricular free wall collapse during diastole, also indicative of tamponade physiology

Figure 5: Transthoracic Echocardiogram of Case #4



A. Dermal Biopsy. Biopsy of maculopapular lesions on the right side of her face revealed changes suggestive of dermatomyositis. Cut sections showed atrophic epidermis, vacuolar interface change, superficial and deep perivascular lymphocytic infiltrates, and neutrophilic dusts around vessels.

B. Muscle Biopsy. Biopsy of atrophied right deltoid muscle showed chronic myopathic process, likely inflammatory myositis

Figure 6: Histopathology done for Case #4

Therapy was initiated with high dose steroids (prednisone 1mg/kg/day). This was followed by azathioprine on the second week. There was gradual improvement of muscle strength. Immediate pericardiocentesis yielded serosanguinous to bloody fluid which was negative for TB PCR and for malignant cells (Table II). A second course of steroid therapy consisted of methylprednisolone pulse which further improved her motor strength and speech. Because of persistent cardiac tamponade on repeat echocardiogram, tube pericardiostomy was inserted and kept for two weeks. Methotrexate was subsequently added on the third week. After five weeks on glucocorticosteroids, four weeks on azathioprine, and two weeks on methotrexate, the pericardial fluid output decreased to less than 20 cc per day, and the tube was subsequently removed. Proximal muscle strength improved, and she could lift her arms against minimal resistance (muscle strength 4/5) and do self care, such as combing her hair. She was sent home on prednisone 1mg/kg/day and methotrexate 12.5 mg/week.

She was seen at the outpatient department after one week, wheel-chair bound but with improved upper extremity strength. Although she still could not stand from a sitting position, she could move her thighs from side-to-side and lift against gravity (muscle strength 3/5), but not yet against resistance. Repeat echocardiography showed improved cardiac function.

Discussion

Cardiac Tamponade in the Philippines

Cardiac tamponade is defined echocardiographically as the presence of pericardial effusion with any one of the following: 1) right atrial late diastolic collapse; 2) right ventricular early diastolic collapse; or 3) both. Echocardiographic tamponade occurs before the onset of overt hemodynamic effects,

and is recognized by cardiac chamber collapse described above. Clinical tamponade represents the most extreme manifestation, causing severe hemodynamic effects (ie. the Beck's triad).^{3,4} A local study in our institution⁵ reviewed 58 patients with cardiac tamponade. The two most common etiologies for cardiac tamponade were malignancies (specifically lung cancer) and tuberculosis. Of note, hemodynamic compromise was only seen in eight (14%) and the Beck's triad was only present in five cases (9.0%). Our present case series documents four Filipinas with SLE and cardiac tamponade. In contrast to foreign literature, tuberculosis may be a more important etiology to be considered in our setting.

Prevalence of Tamponade in SLE

Pericarditis has been the most frequent cardiac finding in connective tissue disease (including overlap syndromes).⁶ Cardiac tamponade among SLE patients is an uncommon event. Pericardial effusion in tamponade was discovered "incidentally" in our patients by 2D-echocardiography and electrocardiogram. Some did not have the typical combination of physical examination findings for tamponade (pulsus paradoxus, hypotension, muffled heart sounds, distended neck veins). Kahl,⁷ in his retrospective review of a database of 395 patients, reported 11 episodes of cardiac tamponade among 75 patients with lupus pericarditis. Three of the 11 patients were uremic. A recent review by Rosenbaum et al.⁸ of 71 patients with SLE documented nine patients with cardiac tamponade. They have concluded that a low C level at presentation was predictive of the development of cardiac tamponade physiology. Almost all other data on cardiac tamponade in SLE is culled from case reports,⁹⁻¹³ wherein most cases of tamponade usually occur during the initial manifestation of SLE. In these reports, SLE was diagnosed by ARA criteria and tamponade was diagnosed by echocardiography. All were given high dose steroids and subsequent drainage was performed.

At present, there are fewer case reports of cardiac tamponade in dermatomyositis. Other similar cases of dermatomyositis were those cited by Chraïbi et al.,¹⁴ Yale et al.,¹⁵ and Martinez et al.¹⁶ It was Chraïbi et al. who recommended that two dimensional echocardiogram be done in all these patients. In our review of literature, cardiac tamponade in SLE-dermatomyositis overlap has not yet been reported to date.

Other Etiologies of Pericardial Effusion

Aside from lupus pericarditis, the pericardial effusion may be a consequence of uremia, infections in the pericardium, or malignancies involving the

pericardium. Our first three cases had proteinuria on admission, hence, this could have contributed to the effusion. The presence of LE cells in the pericardial fluid in our first case favors that the etiology of the tamponade was lupus. In all cases, however, improvement was noted after institution of immunosuppressants, favoring lupus pericarditis. It is mandatory to exclude an infectious cause of pericarditis. In our setting, bacteria and tuberculosis are most common, which were ruled out in all cases by negative pericardial fluid cultures and negative TB-PCR / AFB stains (Table II). Pericardial fluid cytology did not yield atypical cells, making malignant effusion unlikely for all cases.

Autoimmune Thyroiditis and CTD

Autoimmune thyroiditis, as in our fourth case, initially presents with signs and symptoms of hyperthyroidism and serological evidence of thyroid autoimmunity, but without evidence of the immune-mediated thyroid stimulation characteristic of Grave's disease. As autoimmune thyroiditis often irreversibly damages the thyroid gland, this condition frequently progresses to hypothyroidism.¹⁷ In our patient, the diagnosis of autoimmune thyroiditis was made on the basis of the presence of anti-thyroid peroxidase antibodies. Autoimmune thyroiditis may appear in the course of other nonorgan-specific autoimmune diseases, such as SLE.¹⁸ In a study by Mihailova et al.,¹⁹ thyroglobulin antibodies were positive in seven out of 12 cases while antinuclear antibodies were detected in eight out of 12 cases. The high steroid dose and immunosuppressant have added benefits for the autoimmune thyroid disease.

Management

Pericardial tamponade in connective tissue disease is initially managed with pericardiocentesis and steroids. These interventions are instituted immediately. All patients required steroids, but the fourth case needed adjunctive therapy with azathioprine and methotrexate due to recurrence of the effusion.

Conclusion and Recommendation

We have presented four cases of cardiac tamponade as a manifestation of SLE. Literature underlines the rarity of this condition. Despite its rarity, SLE should be considered as one of the differentials of cardiac tamponade, especially in patients who manifest with multi-systemic findings. It requires timely identification and treatment with high dose steroids, after other causes such as infections have been excluded. Immediate drainage through pericardiocentesis or pericardiostomy may be life-saving.

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Appendix: Bohan and Peter Criteria for Dermatomyositis

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| <ol style="list-style-type: none"> 1. Symmetric weakness of proximal muscles with or without dysphagia or respiratory muscle involvement * 2. Positive muscle biopsy * 3. Elevation of muscle enzymes (CK, LDH, AST) 4. EMG findings * 5. Dermatologic features (Gottron's sign, Heliotrope rash) * |
| * : Criteria present in our fourth patient |