CASE REPORT

Cardiac tamponade as a manifestation of severe dermatomyositis

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Accepted 5 September 2016

SUMMARY

In connective tissue disorders, the incidence of pericardial disease and pericardial effusion can be up to 58%, and if untreated, it can lead to cardiac tamponade which can be fatal. Physicians must have a high index of suspicion for this disease as diagnosis can be delayed while evaluating more common causes of tachycardia and hypotension in the immunosuppressed (ie, sepsis). We present a 55-year-old woman with a severe case of dermatomyositis, marked by significant weight loss, a bedridden state and hallmark cutaneous findings. On evaluation of interstitial lung disease, a pericardial effusion was incidentally noted. Serial examination revealed increasing fluid accumulation and progressive tachycardia, and diagnosis of cardiac tamponade was made on echocardiography. After initial pericardiocentesis, the effusion rapidly re-accumulated requiring a pericardial window. In severe presentations of rheumatic disease, cardiac tamponade should be considered as a cause of tachycardia, with or without associated hypotension.

BACKGROUND

Dermatomyositis (DM) is an idiopathic autoimmune disease classically involving the skin and the muscle. The myopathy is characterised by a perivascular and interfascicular infiltrate composed primarily of CD4+ T cells, B cells and macrophages causing perifascicular atrophy and muscle breakdown. The cutaneous involvement is characterised by a CD4+ T-cell infiltrate within the dermis along with perivascular inflammation leading to varying stages of microvascular inflammation.¹ Although the diagnosis of DM suggests skin and muscle involvement alone, it is actually a systemic disorder and is associated with DM-associated malignancy, interstitial lung disease (ILD), functional disability, vascular disease and as in this case, cardiac complications. To better understand all of the clinically relevant complications of this disease, we present a case of DM-associated cardiac tamponade.

CASE PRESENTATION

A 55-year-old Filipino woman presented with concerns of weight loss, weakness and a rash. Her medical history was significant for a ST-elevation myocardial infarction with placement of two drug-eluting stents, hypertension, hyperlipidaemia and recent diagnosis of amyotrophic DM by her dermatologist with skin biopsy showing interface dermatitis; patient was lost to follow-up until current presentation (3 months later). Her

medications included clopidogrel, aspirin, atorvastatin, metoprolol and hydroxychloroquine. She reported a 6-month history of progressive weakness, most pronounced in the proximal muscle groups, evidenced by difficulty in rising from a seated position. Her weakness was severe enough to cause a bedridden state in the weeks preceding presentation. She also noted an unintentional 11 kg weight loss.

Physical examination was significant for multiple distinct dermatologic findings, cachexia, muscle atrophy, fine inspiratory bibasilar crackles and profound weakness in proximal muscle groups including shoulders, hips, arms and legs. Head and neck examination showed a violaceous periorbital rash with oedema consistent with a heliotrope rash and an erythematous rash in a distinct V shape along her neck and upper chest (see [figure 1](#)). Examination of her hands showed beefy violaceous plaques with ulceration along the extensor surfaces of her metacarpophalangeal joints characteristic of Gottron's papules (see [figure 2](#)) and multiple fissures of her erythematous and hyperkeratotic fingertips, typical of mechanic's hands. Given the pathognomonic skin findings and clinical evidence of muscle involvement, a diagnosis of DM was made.²

INVESTIGATIONS

Laboratory studies were significant for thrombocytopenia (73 000/ μ L), an elevated aspartate aminotransferase (61 U/L, normal 0–32), elevated erythrocyte sedimentation rate (57 mm/hour, normal 0–25) and C reactive protein (2.56 mg/dL,

Figure 1 Gottron's papules.

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To cite: Kurth BJ, Wagler V, Keith M. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-216860

Figure 2 'V-neck' sign.

normal 0–0.5) and normal levels of creatine kinase (CK) and aldolase (129 U/L and 4.1 U/L, respectively). Antinuclear antibodies and antibodies against HIV 1/2 were negative. Imaging obtained on admission included a chest x-ray which showed increased interstitial markings.

In order to improve the yield of muscle biopsy, a lower-extremity MRI was obtained with short tau inversion recovery (STIR). This modality was chosen to improve visualisation of muscle inflammation by suppressing fat signals. This study revealed diffuse, bilateral muscle and fascial oedema consistent with DM (see [figure 3](#)). However, muscle biopsy was deferred given patient's unstable clinical status and pretest diagnostic certainty.

In an attempt to further delineate her clinical phenotype,^{3 4} myositis-specific and myositis-associated autoantibodies were

obtained. Serology can guide therapeutic strategies, help determine prognosis and help target surveillance of certain diseases (eg, cancer and ILD). The patient was negative for antibodies against Jo-1, PL-7, PL-12, EJ, OJ, Mi-2, SRP, p155/140/TIF1- γ , CADM-140/MDA-5, NXP-2, U2 sn RNP, PM/Scl complex, SS-A, Ku and U1-RNP.

Since the association between cancer and DM is well documented, and twice as likely to occur in hospital-based presentations,⁵ a thorough evaluation for malignancy was performed. Serologic testing found that carcinoembryonic antigen (CEA) was elevated at 6.6 ng/mL (normal 0.2–4.7). Cancer antigens 125 and 19-9 were found to be normal. Given the elevated CEA, upper and lower GI endoscopies were performed. Colonoscopy only revealed a benign sigmoid polyp confirmed by biopsy. Cervical cancer screening was not performed as patient had had a hysterectomy for uterine fibroids. Mammography was deferred until hospital discharge.

Further evaluation for possible ILD was obtained with high-resolution chest CT of the chest which revealed fine intralobular, septal thickening along subpleural regions, consistent with non-specific interstitial pneumonitis. Additionally, there was a moderate-sized pericardial effusion. Initial transthoracic echocardiography (TTE) revealed a large effusion without signs of cardiac tamponade. Over the next 2 days, the patient's heart rate steadily climbed from the 90s to the low 100s, and repeat TTE showed interval enlargement of the effusion with signs of cardiac tamponade (see [figure 4](#)). Pericardiocentesis retrieved 340 mL of straw-coloured fluid. Gram's, fungal and acid-fast stains were negative, cultures remained negative, adenosine deaminase was normal, and no cellular atypia was found on cytology. The patient's haemodynamics improved immediately.

Over the next 36 hours, the patient grew increasingly tachycardic and suddenly became hypotensive. Manual sphygmomanometry revealed an inspiratory drop of 14 mm Hg systolic, indicating pulsus paradoxus and cardiac tamponade. Bedside echocardiography revealed a large re-accumulated pericardial effusion with advanced tamponade. Emergent pericardiocentesis with pericardial window retrieved sanguineous fluid with a red

Figure 3 MRI STIR of lower extremities showing diffuse muscular and fascial oedema. Black arrow points to uninflamed vastus lateralis. White arrow points to inflamed adductor magnus. STIR, short tau inversion recovery.

Figure 4 Apical four-chamber view demonstrating right atrial collapse during atrial diastole. White arrow points to the right atrial wall. Black arrow points to the large pericardial effusion.

blood cell (RBC) count of 973 000/ μ L. RBC count $>100\,000/\mu$ L is diagnostic of haemorrhagic pericardial effusion.⁶ The patient's haemodynamics immediately improved. Repeat analysis of the pericardial fluid was again negative for infection and malignancy. During the thoracoscopic surgery, fluid samples from the pericardial and a pleural effusion were taken, along with tissue samples from her pleura, pericardium and epicardium. Both effusions had acute and chronic inflammatory cells and occasional reactive mesothelial cells. Fungal and mycobacterial stains and cultures were negative. A pericardial biopsy showed fibrosis and mild chronic inflammation, epicardial biopsy revealed mild chronic inflammation and pleural biopsy showed active inflammation. These findings are consistent with DM with pleural and myocardial involvement.

TREATMENT

The patient was started on daily infusions of 1 mg/kg intravenous methylprednisolone. After 5 days, the patient's response was minimal, and the addition of a course of 2 g/kg intravenous immunoglobulin over 5 days was initiated. Therapeutic pericardiocentesis was chosen for initial treatment of cardiac tamponade. Subsequent pericardial window was performed given the rapidity of recurrence of pericardial effusion and with the initially poor control of the patient's DM with the weight-based glucocorticosteroids and is concordant with the 2015 European Society of Cardiology's Guidelines for the Diagnosis and Management of Pericardial Diseases.⁷

OUTCOME AND FOLLOW-UP

The patient tolerated the pericardial window without complication. The patient was eventually discharged to a skilled-nursing facility on azathioprine and a slow prednisone taper; later, she was transitioned to mycophenolate mofetil due to intolerance of azathioprine.

Within 6 months of hospital discharge, the patient had regained all of her lost weight, was fully ambulatory and able to do all of her activities of daily living, and has not had a re-accumulation of her pericardial or pleural effusions.

DISCUSSION

Pericardial disease is common among certain connective tissue diseases including systemic sclerosis, systemic lupus and mixed

connective tissue disease. In contrast, it is relatively uncommon among DM and polymyositis, thought to involve only 11.4% of cases. The vast majority of these cases represented only small and haemodynamically insignificant pericardial effusions.⁸ To the best of our knowledge, cardiac tamponade secondary to DM in adults has only been reported in three other cases.^{9–11}

The likelihood of a pericardial effusion progressing to cardiac tamponade is dependent on the effusion size and rate of accumulation. The pathophysiology of tamponade is determined by the minimally compliant, fibrous nature of the pericardial sac. When accumulation of pericardial fluid exerts pressure on the four chambers of the heart, cardiac filling and output are diminished, resulting in cardiogenic shock and death.¹² As such, the American Society of Echocardiography and the European Society of Cardiology recommend TTE for suspected pericardial effusion/pericarditis, and emergent echocardiography if cardiac tamponade is suspected.^{7 12}

Haemorrhagic pericardial effusions are typically related to trauma/iatrogenic and malignancy. The diagnosis is determined by the somewhat arbitrary RBC count of $>100\,000/\mu$ L.⁶ Our patient recently underwent a pericardiocentesis while on dual antiplatelet therapy, thereby increasing the likelihood of this haemorrhagic effusion being a postprocedural complication rather than a manifestation of DM.

Of note, our patient had normal CK and aldolase levels despite evidence of muscle atrophy and weakness on examination. In retrospective cohort studies, 19.6–26% of presentations of DM had normal CK values.^{13 14} This underlies the importance of patients with clinically amyopathic DM to be followed closely by a dermatologist and a rheumatologist, both of which are necessary for the management of a disease with multiple clinically important manifestations.

Our case illustrates the importance of keeping a broad differential for tachycardia when evaluating severe presentations of rheumatic disease. Cardiac tamponade is straightforward to treat, but it can be rapidly fatal if unrecognised.

Learning points

- Pericardial disease is common in connective tissue disease and can range from incidental findings to life-threatening conditions.
- Cardiac tamponade should be considered in evaluating tachycardia and/or hypotension in acute presentations of connective tissue disease.
- A normal creatine kinase does not exclude dermatomyositis (DM) from the differential for myopathy with associated rash.
- Muscle biopsy is not necessary for diagnosis when pathognomonic skin findings are seen in cases of DM.
- Pericardial window should be considered for recurrent pericardial effusions.

Contributors All of the above authors contributed to the patient's care and helped in the preparation of this manuscript. BJK drafted the manuscript; performed the literature search; obtained figures 1, 2 and 4; and was responsible for carrying out revisions of the draft. VW is the editor of the manuscript prior to submission, helped with format and case structure, obtained figure 3 and helped with other logistics. MK is co-editor of the manuscript and recommended using the MR images obtained.

Competing interests None declared.

Patient consent Patient consent was obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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