ESC European Society of Cardiology

European Heart Journal - Case Reports (2022) **6**, 1–8
https://doi.org/10.1093/ehjcr/ytac145

CASE REPORT
Other

Cardiac tamponade as the initial presentation of autoimmune polyglandular syndrome Type 2: a case report

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Received 9 August 2021; first decision 21 September 2021; accepted 31 March 2022; online publish-ahead-of-print 8 April 2022

Background Cardiac tamponade is a rare but serious manifestation of autoimmune polyglandular syndrome Type 2 (APS 2). Patients often present with symptoms of thyroid dysfunction and adrenal insufficiency, but the insidious onset of the disease may lead to delayed diagnosis, which can progress rapidly to haemodynamic instability requiring urgent intervention.

Case summary A 39-year-old previously healthy male was admitted with cardiac tamponade complicated by cardiac arrest requiring emergent pericardiocentesis. An extensive work up revealed primary adrenal insufficiency and Hashimoto's thyroiditis. His positive autoantibodies to thyroid peroxidase and 21-hydroxylase combined with rapid improvement with initiation of corticosteroids and levothyroxine confirmed a diagnosis of APS 2.

Discussion Although this disease is often difficult to diagnose given its vague symptoms, it should be considered in the differential diagnosis for young patients presenting with pericardial effusion or cardiac tamponade of unknown origin. Early diagnosis and management are critical and often result in rapid improvement after appropriate treatment.

Keywords Cardiac tamponade • Adrenal insufficiency • Autoimmune polyglandular syndrome Type 2 • Case report

ESC Curriculum 2.2 Echocardiography • 7.3 Critically ill cardiac patient

Learning points

- Cardiac tamponade can be a rare but deadly manifestation of primary adrenal insufficiency disease and autoimmune polyglandular syndrome Type 2.
- Even in patients without known risk factors, haemodynamic instability in the setting of an acute pericardial effusion should be considered tamponade until proven otherwise.
- Early, definitive treatment often allows for quick recovery in these patients arrived.
- Patients with rare diseases benefit from a broad differential and a multidisciplinary team approach to diagnosis and management.

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Handling Editor: Davide Stolfo
Peer-reviewers: Maria Mattioli and Giacomo Mugnai
Compliance Editor: Zhiyu Liu
Supplementary Material Editor: Ammenathul Mazaya Fawzy

Introduction

Autoimmune polyglandular syndrome Type 2 (APS 2), also called autoimmune polyendocrine syndrome or polyglandular autoimmune syndrome Type 2, describes the co-occurrence of autoimmune adrenal insufficiency with autoimmune thyroid disease and/or Type 1 diabetes mellitus. It is a rare syndrome, with an incidence estimated as 1.4–4.5 per 100 000. Patients often present with symptoms of thyroid dysfunction and non-specific symptoms of adrenal insufficiency. However, the disease occasionally presents with life-threatening cardiac tamponade. The below case describes a young, otherwise healthy patient who presented with acute onset cardiac tamponade, ultimately leading to a diagnosis of APS 2.

Timeline

Arrival to the ED (Day 0): A 39-year-old male with no past medical history presented with epigastric pain. Vital signs were significant for tachycardia to 108, hypotensive to 80/39, tachypnoeic to 24 with oxygen saturation 100%, prompting aggressive fluid resuscitation

1 h after arrival (Day 0): laboratory findings largely unremarkable but still hypotensive. Echocardiogram with mild pericardial effusion with no evidence of right atrium or ventricle compromise

3 h after arrival (Day 0): Developed progressive hypoxaemia requiring intubation

4 h after arrival (Day 0): Pulseless electrical activity cardiac arrest for 6 min with return of spontaneous circulation (ROSC) following Advanced Cardiovascular Life Support (ACLS)

5 h after arrival (Day 0): Transferred via helicopter to medical intensive care unit at a tertiary care centre on norepinephrine 3 mcg/kg/min, phenylephrine 9 mcg/kg/min, and vasopressin 0.03 mcg/kg/min

7 h after arrival (Day 0): Repeat echocardiogram with large pericardial effusion with reduced ventricular filling. Underwent emergent pericardiocentesis but still required vasopressors

Day 3: Found to have elevated thyroid-stimulating hormone (TSH) and low free thyroxine (T4), random cortisol of <0.05 and his cosyntropin stimulation test revealed an elevated adrenocorticotropic hormone (ACTH) and undetectable cortisol. Started on high-dose corticosteroids and levothyroxine with improvement in haemodynamics

Day 4: Extubated and weaned off of vasopressors

Day 6: Transferred out of the intensive care unit

Day 12: Discharged home

Three months after discharge: Readmitted to the hospital with mild pericarditis (without tamponade), discharged after 2 days on colchicine and ibuprofen.

Case presentation

A 39-year-old male with no significant past medical history presented to the emergency department with 3 months of unintentional weight loss and 2 days of worsening epigastric pain. On initial presentation to the outside hospital, he was afebrile, tachycardic to 108, hypotensive to 80/39, tachypnoeic to 24 with oxygen saturation 100% on room air. His physical examination on arrival was notable only for diffuse hyperpigmentation. His initial work up revealed a slight leucocytosis to 10.3 $\times$ 10^9/L (reference range: 4.0–10.0 $\times$ 10^9/L), sodium of 128 mmol/L (reference range: 136–145 mmol/L), C-reactive protein elevated at 3.1 mg/dL (reference range <0.30 mg/dL), B-type natriuretic peptide pro (proBNP) of 1,428.0 pg/mL (reference range <125 pg/mL) and a negative troponin. He was resuscitated with 4 L of normal saline without improvement in his blood pressure. His initial electrocardiogram (ECG) showed sinus tachycardia and low voltage without electrical alternans (*Figure 1A*). His chest X-ray showed increased bilateral interstitial markings without any focal infiltrates. Computed Tomography Angiography (CTA) was negative for pulmonary embolus but revealed a trace pericardial effusion. An initial transthoracic echocardiogram (TTE) before intubation demonstrated a mild-to-moderate pericardial effusion without evidence of right atrial or ventricular collapse (*Figure 2A*), mildly increased right atrial pressure as evidenced by IVC diameter measuring <2.1 cm and collapsing <50% with a sniff. There was also increased variation of mitral inflow velocities on which the official report did not comment (*Figure 2B*). Left ventricular ejection fraction (LVEF) was estimated by visual assessment to be preserved at 55–60%. The patient became progressively altered requiring intubation for airway protection. Despite these interventions, he was acidotic to an arterial pH of 7.04 and had a pulseless electrical activity arrest for 6 min requiring Advanced Cardiovascular Life Support, after which return of spontaneous circulation was achieved. He subsequently required three vasopressors and was airlifted to the medical intensive care unit at a tertiary care centre for further management.

The patient arrived to our unit on norepinephrine 3 mcg/kg/min, phenylephrine 9 mcg/kg/min, and vasopressin 0.03 mcg/kg/min. Repeat TTE showed evidence of tamponade physiology including a large circumferential pericardial effusion measuring 2.6 cm and diastolic collapse of the right ventricular free wall (*Figure 2C* and *D*). Respiratory variation in inflow velocities is more difficult to interpret in an intubated patient (*Figure 2E* and *F*). A left-sided pleural effusion was also noted on TTE, whereas a right-sided pleural effusion was noted on transesophageal echocardiography (TEE). His TEE also showed mild global hypokinesis with LVEF decreased to 40–45%. The patient underwent emergent pericardiocentesis with the removal of 200 mL of serosanguinous fluid (*Figure 2G* and *H*) leading to improvement in his haemodynamics. The patient arrived at the cath laboratory and pressors were titrated during the procedure to support perfusion. About 1 h after completion of the pericardiocentesis, the patient had been weaned off phenylephrine but remained on norepinephrine and vasopressin. His repeat ECG (*Figure 1B*) continued to show low voltage.

Given the unclear aetiology of his tamponade, a broad infectious and autoimmune work up was sent (*Table 1*). His thyroid-stimulating hormone (TSH) was elevated to 30 μIU/mL (reference range: 0.27–0.42 μIU/mL) with a T3 of 67.3 ng/dL (reference range: 72–153 ng/dL) and a free T4 of 0.34 ng/dL (reference range: 0.8–1.7 ng/dL). His random cortisol was <0.05 μg/dL and his cosyntropin stimulation test revealed an elevated adrenocorticotropic hormone (ACTH) and undetectable cortisol, leading to a diagnosis of primary adrenal insufficiency. Computed tomography of the abdomen

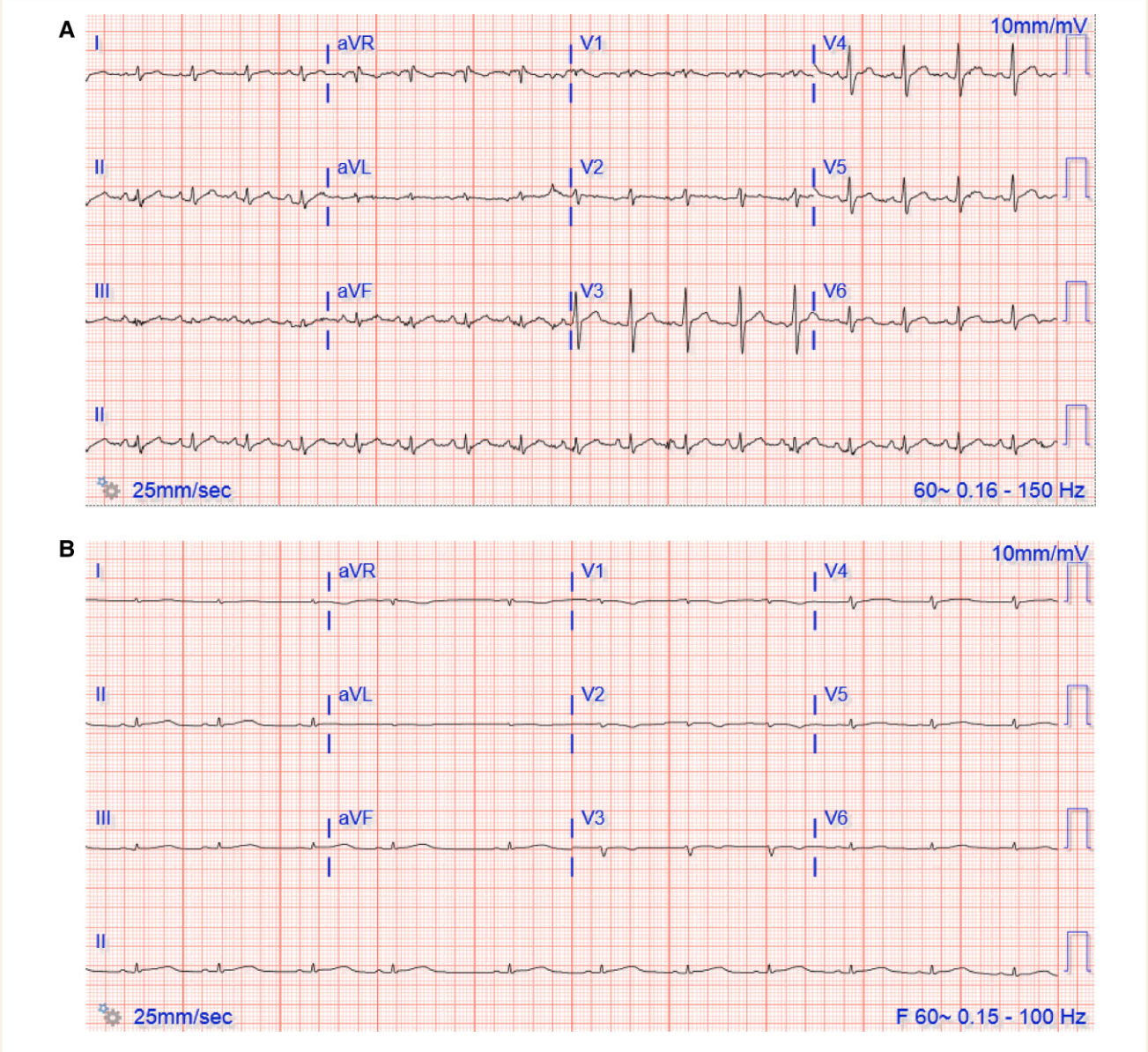

Figure 1 Electrocardiograms. The patient initially presented with tachycardia and low voltage (*A*), the latter of which did not resolve immediately after pericardiocentesis (*B*).

showed slightly atrophic adrenal glands bilaterally. Further autoimmune work up was significant for an elevated antinuclear antibody (1:80, dense fine speckled), as well as an elevated thyroid peroxidase antibody and 21-hydroxylase antibody level. High-dose corticosteroids and levothyroxine were initiated on hospital Day 3, and the patient showed marked improvement; he was extubated and weaned off all vasopressors by hospital Day 4. The combination of autoimmune adrenal insufficiency and autoimmune thyroid disease confirmed a diagnosis of APS 2.

The patient was admitted to the hospital for 12 days (6 of which were in the intensive care unit). Repeat TTE on hospital Day 2 and Day 4 demonstrated bilateral pleural effusions. Following the pericardiocentesis, electrocardiogram continued to show low voltages. The patient's LVEF recovered to 55–60% by hospital Day 7. He was discharged home on hydrocortisone 20 mg in the morning, hydrocortisone 10 mg at night and levothyroxine 125 mcg daily with close follow-up.

Twelve weeks later, he presented to the hospital with pleuritic chest pain, upper abdominal pain, and nausea and was readmitted briefly with pericarditis. CTA showed small pericardial effusion with pericardial thickening and trace bilateral pleural effusions noted, whereas TTE showed trivial pericardial effusion without evidence of haemodynamic compromise. The patient was treated with intravenous fluids, stress dose hydrocortisone, ibuprofen, and colchicine before being discharged home with outpatient follow-up.

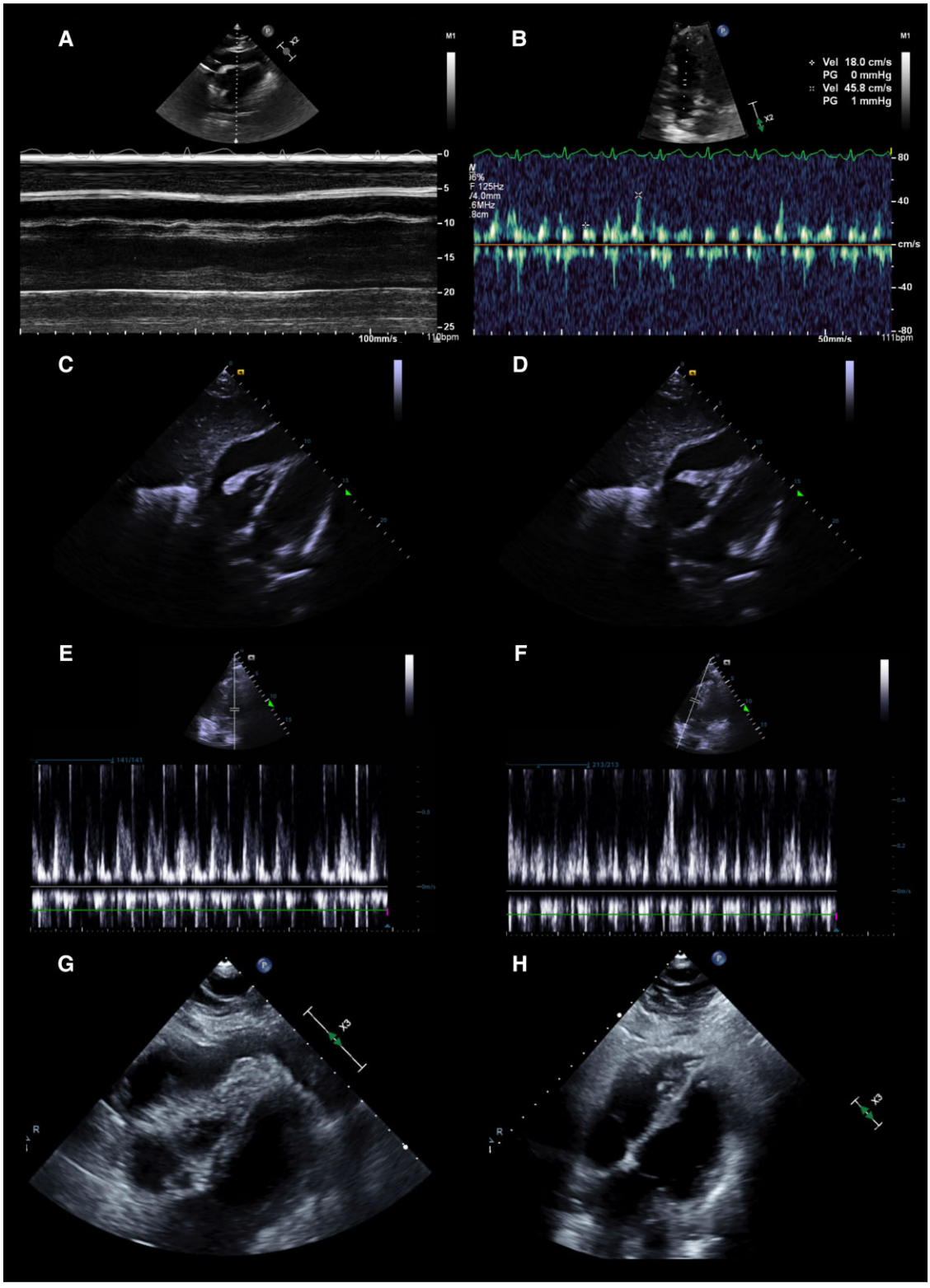

Figure 2 Transthoracic echocardiograms. Initial echo at the outside hospital did not reveal right ventricular diastolic collapse (*A*), but did show increased variability of mitral inflow velocities (*B*). Repeat echo at our tertiary hospital showed right ventricular diastolic collapse (*C* and *D*), while variations in inflow velocities partially reflected the patient's intubated status (*E* and *F*). In the cath laboratory (G), ~200 mL was removed, which essentially resolved the effusion (*H*).

Table 1 Presenting laboratories and work up

Test	Result	Reference range
Chemistry		
TSH	30	0.27–0.42 μIU/mL
T3, total	67.3	72–153 ng/dL
Free T4	0.34	72–153 ng/dL
ACTH	560	7.2–63.3 pg/mL
Cortisol	<0.05	Variable, 0.2–18 μg/dL
Immunology		
Thyroid peroxidase Ab	182	<34 IU/mL
21-hydroxylase Ab	Positive	Negative
Adrenal total auto Ab	Negative	Negative
Glutamic acid decarboxylase	0.0	<0.02 nmol/L
IGF-1	33	53–331 ng/mL
ANA	1:80 (dense fine speckled)	<1:80
ANCA panel (c-ANCA, p-ANCA)	Negative	Negative
Myeloperoxidase Ab	2.8	<3.5 EliA U/mL
Proteinase 3 Ab	<0.7	<2.0 EliA U/mL
RF	<10	<14 IU/mL
dsDNA Ab, IgG	0.8	<0.10 IU/mL
SS-A	0.6	<7 EliA U/mL
SS-B	0.6	<7 EliA U/mL
Smith Ab	2.5	<7 EliA U/mL
Scleroderma (Scl-70)	0.8	<7 EliA U/mL
Jo-1 Ab	0.3	<7 EliA U/mL
IL-6	619	<7.0 pg/mL
Microbiology		
Babesia	Not detected	
Treponema pallidum, serum	Not detected	
Respiratory viral panel	Not detected	
Cytomegalovirus	Not detected	
Enterovirus and parechovirus	Not detected	
Lyme antibodies	Not detected	
Anaplasma, DNA PCR	Not detected	
Epstein–Barr Virus	Not detected	
Cryptococcal Ag	Negative	
Fungitell (1-3)-B-D-glucan	Negative	
Aspergillus galactomannan antigen	Negative	
Histoplasma Ag, urine	Negative	
Legionella and S. Pneumo	Negative	

TSH, thyroid-stimulating hormone; ACTH, adrenocorticotropic hormone; Ab, antibody; Ag, antigen; IGF-1, insulin-like growth factor; ANA, antinuclear antibody; ANCA, antineutrophil cytoplasmic antibody; RF, rheumatoid factor; SS, Sjögren's syndrome; IL-6, interleukin-6

Discussion

Autoimmune polyglandular syndrome Type 2 is a rare immunoendocrinopathy affecting the adrenal glands and the endocrine pancreas and/or the thyroid. Although APS 2 can affect individuals across the lifespan, onset occurs most commonly between the age of 30 and 40 with a female predominance (female:male ratio ranging from 1.8 to 4.0).[1–5]

Specific HLA haplotypes have been found to be associated with autoimmune adrenal insufficiency in APS 2.[1,2,6] However, the pathogenesis remains poorly elucidated. One theory involves the development of autoantibodies against antigens from the same embryologic germ layer, but this would not explain the autoimmune disease in organs from different germ layers or why endocrine organs seem preferentially affected in these syndromes while other organs from within the same germ layer would be unaffected.[1] Loss of regulatory T-cell function could help explain autoimmunity against multiple targets and organ systems.[6] It also remains unclear if the loss of tolerance to multiple antigens occurs simultaneously or sequentially.[1]

Patients often present with symptoms of thyroid dysfunction (either hypothyroid or hyperthyroid) and non-specific symptoms of adrenal insufficiency (including fatigue, weight loss, decreased appetite, and abdominal pain). In addition, patients may present with other autoimmune conditions including vitiligo, hypogonatropic hypogonadism, autoimmune hepatitis, alopecia, pernicious anaemia, myasthenia gravis, or Sjögren syndrome.[1–3,7] Owing to cortisol and aldosterone deficiencies, general laboratory findings in patients with primary adrenal insufficiency include hyponatraemia, hyperkalaemia, Type IV renal tubular metabolic acidosis, hypoglycaemia or decreasing insulin requirements, hypercalcaemia, mild normocytic anaemia, lymphocytosis, and mild eosinophilia.[7,8]

Diagnosis involves serologies and organ function tests. Autoantibodies against the adrenal glands (21 hydroxylase antibodies, adrenal cortex antibodies), thyroid (thyroid peroxidase antibodies, thyroglobulin anitbodies), and pancreas (glutamic acid decarboxylase autoantibodies, insulin autoantibodies, islet cell cytoplasmic autoantibodies, tyrosine phosphatase-like autoantibodies, and insulinoma-associated 2 autoantibodies) are often present. There are no specific imaging findings in APS 2; the adrenal glands often appear normal but may become atrophied later in the course.[1]

Pericardial effusions have been reported in primary adrenal insufficiency, and in recent years, there have been multiple case reports of patients with APS 2 initially presenting with pericardial effusion and/or cardiac tamponade (*Table 2*).[9–15] The pathogenesis of the effusion in APS 2 may result from autoimmune inflammation of the pericardium with resulting inflammatory reaction and fluid accumulation.

Tamponade physiology may develop more easily in the setting of primary adrenal insufficiency due to several factors. First, aldosterone deficiency results in mild intravascular volume depletion, which decreases right-sided filling and allows right atrial and ventricular collapse.[9,11,13] Second, baseline cortisol deficiency results in decreased vascular tone, and therefore propensity towards hypotension.[10] Finally, these patients lack a stress response and are therefore at higher risk for haemodynamic instability and shock in the acute setting.[10,14] In our patient, the aetiology of cardiac arrest was likely multifactorial, including distributive shock secondary to adrenal crisis, worsening severe acidosis, and tamponade physiology. The persistence of refractory shock at our tertiary care hospital reflected continued adrenal crisis and worsening tamponade physiology, as removal of the pericardial fluid resulted in immediate, although partial, improvement in haemodynamics.

Table 2: Characteristics of previously reported cases of cardiac tamponade in Autoimmune polyglandular syndrome, type II

	Alkaabi et al. 2008[9]	Alkaabi et al. 2008[9]	Alkaabi et al. 2008[9]	Palmer et al. 2014[10]	Khalid et al. 2015[11]	McNamara et al. 2017[12]	Vryonidou et al. 2017[13]	Bacal et al. 2018[14]	Marinho et al. 2020[15]
Age/Sex	34 y/o F	58 y/o M	35 y/o M	54 y/o M	48 y/o F	29 y/o M	40 y/o F	21 y/o M	32 y/o M
History of present illness	Breathlessness, central chest pain, and long-standing lethargy with weight loss	Long-standing lethargy, weight loss, nausea, and excessive tiredness on minimal exertion	Long-standing breathlessness, Unusual gum hyperpigmentation noted during a dental visit	Four days of worsening weakness, subjective fevers, nausea, and malaise leading to decreased oral intake, Two days of non-radiating substernal, dull, pleuritic chest pain	Fatigue, malaise, sudden onset of pleuritic left-sided chest pressure, and associated shortness of breath.	Positional chest pain	Admitted to ICU for rapid onset dyspnea and orthopnea in setting of progression weakness, 10kg weight loss, and amenorrhea during last year with intermittent fever in previous two months	Flu-like symptoms 1-2 days, chest pain	Three days of retrosternal chest pain and low-grade fevers
Vitals and Exam	Afebrile, severely hypotensive, hyperpigmented, tachycardiac, distant heart sounds	Hyperpigmented and had pulsus paradoxus	Admitted to medical ICU with hypotension and hypoxia Striking hyperpigmentation	Somnolent SBP 60s following 4L NS JVD to angle of the jaw Distant heart sounds Diminished peripheral pulses	BP 90-100/40-60 HR 110-120 Decreased breath sounds, dullness to percussion, decreased tactile fremitus JVP 12cm	BP 70/40 HR 130 JVD to the angle of the mandible Soft heart sounds Friction rub	Febrile Tachycardic Hypotensive Slight hyperpigmentation	BP 108/64 (on pressors) HR 121	Friction rub
Past medical history	Hashimoto thyroiditis on levothyroxine	No reported significant past medical history	Autoimmune thyroiditis on levothyroxine	Longstanding primary adrenal insufficiency (no longer on fludrocortisone but prednisone) and primary hypothyroidism on levothyroxine	Known APS II with Addison's disease on predisone and fludrocortisone and autoimmune primary hypothyroidism (no comment on levothyroxine dose)	Hypothyroidism	Hashimoto thyroiditis on levothyroxine	Raynuad's	Childhood asthma Nonallergic rhinitis Idiopathic episcleritis Recent admission for tonsillitis c/b septic shock
General laboratory studies	Hyponatremia; Hyperkalemia; Acidosis TSH: 8 mIU/L ACTH: 355 pg/mL Cortisol: undetectable 250-mcg cosyntropin stimulation test confirmed adrenal insufficiency	TSH: 25 mIU/L ACTH: 261 pg/mL Cortisol: undetectable 250-mcg cosyntropin stimulation test confirmed adrenal insufficiency	Na 132 mEq/L K 4.9 mEq/L Anemia Albumin 1.8 TSH: 12 mIU/L ACTH: 137 pg/mL Cortisol: 1.6 µg/dL before and 1.7 µg/dL 1 hour after 250-mcg cosyntropin stimulation testing	Na 131 mEq/L HCO3 17 mmom/L Cr 1.7 mg/dL INR 1.2 ALT 56 u/l AST 61 u/L Total bilirubin 2 mg/dL CRP: 87.5 mg/L TSH wnl Cortisol: undetectable	WBC 16,3000 /uL with 85% PMN Na 131 Phos 1.7 mg/dL Mg 1.4 mg/dL Ionized Ca 1.04 mg/dL	Morning and random cortisol: undetectable	Na 131 mEq/L Hb 11.3 mg/dL CRP 26.4 mg/D C4 9ng/dL AM cortisol 5 µg/dL, ACTH: >1400pg/mL	Troponin 2.19 ng/mL TSH 17.9 uL/mL T4: 1.0 Cortisol 0.5ug/dL (midday on pressors, intubated, balloon pump) Coxsackie B type 5 antibodies: 1:160	Cortisol: undetectable ACTH: 172 pg/mL

Continued

Table 2: ***Continued***

	Alkaabi et al. 2008[9]	Alkaabi et al. 2008[9]	Alkaabi et al. 2008[9]	Palmer et al. 2014[10]	Khalid et al. 2015[11]	McNamara et al. 2017[12]	Vryonidou et al. 2017[13]	Bacal et al. 2018[14]	Marinho et al. 2020[15]
Auto-antibodies	Thyroperoxidase "Adrenal"	Thyroperoxidase	Thyroperoxidase Transglutaminase Endomysial	None reported	None reported	Thyroid peroxidase 21-hydroxylase Glutamic acid decarboxylase	Parietal cell	Thyroid peroxidase Thyroglobulin Glutamic acid decarboxylase 21-hydroxylase	Anti-intrinsic factor Hypogonadism
Electrocardiogram	Not reported	Note reported	Not reported	NSR with diffuse ST-segment elevation and PR depression	NSR with low volage	Diffuse PR depressions	None reported	None reported	Sinus tachycardia with widespread STE and PR depression
Echocardiography	Large pericardial effusion	Notable pericardial fluid and cardiac tamponade	Cardiac tamponade (Bilateral pleural effusions on lung ultrasound)	Moderately sized loculated pericardial effusion with right ventricular collapse and phasic respiratory hyperdynamic motion of the interventricular septum	Moderate-sized pericardial effusion, and impaired diastolic filling of the right atrium and right ventricle	Low-volume, circumferential pericardial effusion, with diastolic right atrial and right ventricular collapse, and >25% respiratory flow variation across the mitral valve septal to lateral E' ratio >1.0 and elevated absolute septal E' velocity of 8.7 cm/s	Significant pericardial effusion (Pleural effusions on lung ultrasound)	RV diastolic flattening, significant respiratory variation of mitral and tricuspid inflow, diastolic septal bounce, and plethora of the IVC	Mild, circumferential pericardial effusion; abnormal rapid motion of interventricular septum (notching in early diastole), lateral e' velocity lower than the medial e' velocity, exacerbated respiratory variance of mitral and plethoric IVC, expiratory reversal of diastolic wave
Pericardiocentesis and Cardiac Catheterization	190 mL yellow and cloudy fluid WBC 1500 with 89% PMN LDH: 1748 U/L Bacterial, acid-fast, and fungal negative	Volume not reported Fluid: Yellow and +Cloudy WBC 10, 200 with 93% PMN LDH: 295 U/L Bacterial, acid-fast, and fungal negative	Volume not reported Pericardial fluid: Yellow and cloudy WBC 12 438 with 94% PMN LDH: 540 U/L Bacterial, acid-fast, and fungal negative	250mL straw colored fluid Mean RAP 14 mmHg Mean PCWP 14mmHg Arterial SBP 40-70mmHg	300 mL of thin yellow pericardial fluid by subxiphoid pericardial window Pericardial fluid cultures negative	150 mL straw-colored fluid Negative cultures WBC 29,375/mL with 81% PMN RAP and RVDP 25mmHg PCWP 26mmHg	Volume not reported Cultures negative	400mL amber fluid drained before referral to author's reference hospital	350mL serous fluid positive for *Streptococcus mitis*
Recurrence	7 documented attacks of pericarditis over 28 months	1 episode left-sided pleuritis and 5 episodes of pericarditis	2 episodes pleural effusions (1 unilateral, 1 bilateral)	Similar presentation requiring repeat pericardiocentesis of 140mL blood tinged fluid	None reported	Patient had presented with idiopathic pericarditis with tamponade requiring pericardiocentesis 3 months earlier	ENA autoantibodies positive on repeat analysis and malar rash, arthritis, polyserositis present so diagnosis of SLE also made	Episode of recurrent pericarditis with mild to moderate effusion that grew to large effusion within 24 as patient became hypotensive and tachycardic. Managed with pericardial window	Episode of recurrent pericarditis and tamponade managed with pericardiocentesis and pleuro-pericardial window

TSH: Thyroid stimulating hormone; WBC: White blood cells; PMN: Polymorphonuclear neutrophils; LDH: lactate dehydrogenase; JVD: jugular venous distension; NSR: normal sinus rhythm; RAP: right atrial pressure; PCWP: pulmonary capillary wedge pressure; RVDP: right ventricular diastolic pressure; IVC: inferior vena cava; ENA: extractable nuclear antigen; SLE: Systemic Lupus erythematosus

Nine reported cases of tamponade in APS 2 are summarized in *Table 2*. Several themes emerge from these case reports, and the current case reflects each of these themes. First, patients were relatively young (aged 21–58 years old) and healthy, aside of pre-existing autoimmune disease. Although there is a female predominance for APS 2, two-thirds of the published tamponade cases occurred in males. In addition, these patients presented with non-specific symptoms, but physical examination revealed signs of tamponade physiology. Interestingly, the amount of fluid responsible for haemodynamic instability in these patients may be relatively small. From the six cases in which the volume of pericardial removed was reported, two had <200 mL removed. Finally, pericardial effusion often recurs, necessitating frequent adjustment of hormone replacement and often pericardial window; among the nine cases, four had recurrent tamponade, two had recurrent pericarditis, and one had recurrent pleural effusions. In our case, the patient was a young healthy male who presented with non-specific symptoms, atrophic adrenal glands, accumulation of 200 mL of pericardial fluid causing haemodynamic instability, and has had at least one episode of recurrent pericarditis following initial diagnosis.

Conclusion

Cardiac tamponade is a rare but serious manifestation of APS 2. Although this disease is often difficult to diagnose given its vague symptoms, it should be considered in the differential diagnosis, especially in young patients presenting with cardiac tamponade of unknown origin. Early diagnosis and management are critical and often result in rapid improvement after the initiation of corticosteroids.

Lead author biography

Laura Glick, MD, is a currently a third-year resident in internal medicine at Yale New Haven Hospital. She received her Bachelor's degree from Tufts University and her MD degree from the University of Chicago Pritzker School of Medicine. She has published more than 30 peer-reviewed articles, and she has presented her research nationally and internationally. In addition, she was featured in the "Diagnosis" column of the the New York Times and on the Clinical Problem Solvers Podcast for her work with this case.

Acknowledgements

The authors would like to acknowledge Erica Herzog, Sanket Thakore, and Avinainder Singh for their help with the diagnosis and treatment of this patient.

Slide sets: A fully edited slide set detailing these cases and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared..

Funding: None declared.

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