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

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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 auto-antibodies 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.
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 cosynotropin stimulation test revealed an elevated adrenocorticotropin hormone (ACTH) and undetectable cortisol. Started on high-dose corticosteroids and levothyroxine
Day 4: Extubated and weaned off of vasopressors
Day 6: Transferred out of the intensive care unit
Day 12: Discharged home

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 × 10^9/L (reference range: 4.0–10.0 × 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 acidic 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 airlifed 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 etiology of his tamponade, a broad infectious and autoimmune work up was sent (Table 1). His thyroid-stimulating hormone (TSH) was elevated to 30 μU/mL (reference range: 0.27–0.42 μU/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 cosynotropin stimulation test revealed an elevated adrenocorticotropin hormone (ACTH) and undetectable cortisol, leading to a diagnosis of primary adrenal insufficiency. Computed tomography of the abdomen
showed slightly atrophic adrenal glands bilaterally. Further auto-
immune 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 corticoster-
oids and levothyroxine were initiated on hospital Day 3, and the pa-
tient showed marked improvement; he was extubated and weaned
off all vasopressors by hospital Day 4. The combination of auto-
immune adrenal insufficiency and autoimmune thyroid disease con-
firmed 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 μU/mL |
| T3, total                     | 67.3       | 72–153 ng/dL    |
| Free T4                       | 0.34       | 72–153 ng/dL    |
| ACTH                          | 560        | 72–63.3 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    |
| Sclerodema (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 galactomanannan   | Negative   |                  |
| antibogen                    |            |                 |
| Histoplasma Ab, 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 immunendocrinopathy 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 2.1,2,4 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.1,4–7 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 antibodies), 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).9–12 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

| Study          | Age/Sex | History of present illness | Vitals and Exam | Past medical history | General laboratory studies |
|----------------|---------|----------------------------|-----------------|----------------------|--------------------------|
| Alkaabi et al. 2008 | 34 y/o F | Breathlessness, central chest pain, and long-standing lethargy with weight loss | Afebrile, severely hypotensive, hyperpigmented, tachycardic, distant heart sounds | Hashimoto thyroiditis on levothyroxine | Hyponatremia; Hyperkalemia; Acidosis |
| Alkaabi et al. 2008 | 58 y/o M | Long-standing lethargy, nausea, and excessive tiredness on minimal exertion | Hyperpigmented and had pulsus paradoxus | No reported significant past medical history | TSH: 25 mIU/mL ACTH: 261 pg/mL Cortisol: undetectable |
| Alkaabi et al. 2008 | 35 y/o M | Long-standing breathlessness, Unusual gum hyperpigmentation noted during a dental visit | Admitted to medical ICU with hypotension and hypoxia | Autoimmune thyroiditis on levothyroxine | ACTH: 137 pg/mL Cortisol: 1.6 µg/dL before and 1.7 µg/dL 1 hour after |
| Palmer et al. 2014 | 54 y/o M | Four days of worsening weakness, subjective fevers, nausea, and malaise leading to decreased oral intake, Two days of non-radiating substernal, dul, pleuritic chest pain | Somnolent SBP 60s following 4L NS | Kown APS II with Addison’s disease on prednisone and prednisolone and autoimmune primary hypothyroidism on levothyroxine | TSH: 13 mEq/L K 4.9 mEq/L Anemia Albumin 1.8 |
| Khalid et al. 2015 | 48 y/o F | Positional chest pain | BP 90-100/40-60 HR 110-120 | Hashimoto thyroiditis on levothyroxine | Na 132 mEq/L K 4.9 mEq/L Anemia Albumin 1.8 |
| McNamara et al. 2017 | 29 y/o M | Admitted to ICU for rapid onset dyspnea and orthopnea in setting of progression weakness, 10 kg weight loss, and amenorrhea during last year with intermittent fever in previous two months | BP 70/40 HR 130 JVD to the angle of the mandible Soft heart sounds Friction rub | Raynaud’s | Na 131 mEq/L HCO3 17 mmol/L Cr 1.7 mg/dL INR 1.2 ALT 56 uL AST 61 uL Total bilirubin 2 mg/dL CRP 87.5 mg/mL |
| Vryonidou et al. 2017 | 40 y/o F | Admitted to ICU for rapid onset dyspnea and orthopnea in setting of progression weakness, 10 kg weight loss, and amenorrhea during last year with intermittent fever in previous two months | BP 70/40 HR 130 JVD to the angle of the mandible Soft heart sounds Friction rub | Hashimoto thyroiditis on levothyroxine | Na 131 mEq/L HCO3 17 mmol/L Cr 1.7 mg/dL INR 1.2 ALT 56 uL AST 61 uL Total bilirubin 2 mg/dL CRP 87.5 mg/mL |
| Bacal et al. 2018 | 21 y/o M | Three days of retrosternal chest pain and low-grade fevers | BP 108/64 (on pressors) HR 121 | Childhood asthma | Na 131 mEq/L HCO3 17 mmol/L Cr 1.7 mg/dL INR 1.2 ALT 56 uL AST 61 uL Total bilirubin 2 mg/dL CRP 87.5 mg/mL |
| Marinho et al. 2020 | 32 y/o M | | Friction rub | Nonallergic rhinitis | Na 131 mEq/L HCO3 17 mmol/L Cr 1.7 mg/dL INR 1.2 ALT 56 uL AST 61 uL Total bilirubin 2 mg/dL CRP 87.5 mg/mL |

Continued
### Table 2: Continued

| Auto-antibodies | Electrocardiogram | Echocardiography | Pericardiocentesis and Cardiac Catheterization | Recurrence |
|-----------------|-------------------|------------------|-----------------------------------------------|------------|
| Thyroperoxidase | Not reported | Large pericardial effusion | 190 mL yellow and cloudy fluid/WBC 1500 with 89% PMN, LDH: 1748 U/L | 7 documented attacks of pericarditis over 28 months |
| Thyroperoxidase | Not reported | Notable pericardial fluid and cardiac tamponade | Volume not reported | 1 episode left-sided pleuritis and 5 episodes of pericarditis |
| Thyroperoxidase “Adrenal” | Not reported | Cardiac tamponade | Fluid: Yellow and cloudy WBC 10, 200 with 93% PMN LDH: 295 U/L, Bacterial, acid-fast, and fungal negative | 2 episodes pleural effusions (1 unilateral, 1 bilateral) |
| Transglutaminase Endomysial | Not reported | Moderately sized loculated pericardial effusion with right ventricular collapse and phasic respiratory hyperdynamic motion of the interventricular septum | Pericardial fluid, and cardiac tamponade (Bilateral pleural effusions on lung ultrasound) | Similar presentation requiring repeat pericardiocentesis of 140 mL blood tinged fluid |
| None reported | NSR with diffuse ST-segment elevation and PR depression | 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 | Volume not reported | None reported |
| None reported | NSR with low voltage | Moderate-sized pericardial effusion, and impaired diastolic filling of the right atrium and right ventricle | Pericardial fluid cultures negative | 2 episodes of pericarditis requiring pericardiocentesis 3 months earlier |
| None reported | Diffuse PR depressions | 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 | None reported | Patient had presented with idiopathic pericarditis with tamponade requiring pericardiocentesis 3 months earlier |
| Thyroid peroxidase | None reported | Significant pericardial effusion | 150 mL straw-colored fluid | ENA autoantibodies positive on repeat analysis and malar rash, arthritis, polyserositis present so diagnosis of SLE also made |
| 21-hydroxylase | None reported | (Pleural effusions on lung ultrasound) | Volume not reported | 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 |
| Glutamic acid decarboxylase | None reported | RV diastolic flattening, significant respiratory variation of mitral and tricuspid inflow, diastolic septal bounce, and plethora of the IVC | Cultures negative | Episode of recurrent pericarditis and tamponade managed with pericardiocentesis and pleuro-pericardial window |
| Non reported | None reported | MD, 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 | 400 mL amber fluid drained before referral to author’s reference hospital | |
| 21-hydroxylase |

**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

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*Initial presentation of APS 2*
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.

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**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.

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