

A rare case of constrictive pericarditis with Budd–Chiari syndrome due to right atrial thrombosis

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Abstract

Patients living with HIV (PLWH) with previous pulmonary tuberculosis, presenting with disproportionate ascites to peripheral congestion, should alert the clinician to consider constrictive pericarditis and Budd–Chiari syndrome (BCS). Constrictive pericarditis is the scarring and loss of the pericardial sac elasticity. The aetiology of constrictive pericarditis varies between developed and developing countries, with infective causes like tuberculosis being significant in South Africa. Budd–Chiari syndrome is a group of disorders characterised by hepatic venous outflow obstruction. The level of obstruction in Budd–Chiari syndrome varies globally. In Asia, South Africa, India, and China, obstruction is predominantly found in the inferior vena cava while in Western countries, hepatic vein obstruction occurs. Patients living with HIV are at increased risk of arterial and venous thromboembolism. The clinician must consider Budd–Chiari syndrome in patients living with HIV presenting with ascites. In patients living with HIV, tuberculosis co-infection has been associated with a higher risk of pericarditis. Both constrictive pericarditis and Budd–Chiari syndrome share a remarkably similar clinical presentation, with ascites and hepatomegaly. There is a dearth of literature on co-existent constrictive pericarditis and Budd–Chiari syndrome. We describe a 31-year-old HIV-infected female, on anti-retroviral therapy (CD4 count 208 cells/uL, undetected viral load), with previous pulmonary tuberculosis, who presented with a 2-month history of abdominal swelling, peripheral oedema, and New York Heart Association grade 4 dyspnoea. Examination revealed an elevated jugular venous pulsation with CV waves, atrial fibrillation, right-sided S3 gallop, pansystolic murmur (3/6) at the left sternal border, tender hepatomegaly, and massive ascites with minimal peripheral oedema. The discordant size of ascites prompted investigations, namely, ultrasound abdomen, echocardiogram, and computed tomography (chest and abdomen). These revealed constrictive pericarditis and Budd–Chiari syndrome with thrombus formation in the right atrium, hepatic vein, and inferior vena cava. She was initiated onto anti-coagulation, anti-tuberculosis therapy and referred for pericardiectomy. Clinicians must maintain a suspicion for constrictive pericarditis and Budd–Chiari syndrome in HIV-infected patients, especially in those with a previous tuberculosis, presenting with features of right heart failure.

Keywords

Infectious diseases, cardiovascular, gastroenterology/hepatology, tuberculosis, Budd–Chiari syndrome, HIV, constrictive pericarditis

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Introduction

Constrictive pericarditis (CP) is the scarring and consequent loss of normal elasticity of the pericardial sac.¹ The aetiology of CP varies between developed and developing countries, with infective causes (such as tuberculosis (TB)) being highly significant in a country like South Africa. Patients with CP present with symptoms related to fluid overload or decreased cardiac output, fatigability, and dyspnoea.¹

Budd–Chiari syndrome (BCS) is a heterogeneous group of disorders characterised by hepatic venous outflow

obstruction.^{2,3} Primary BCS involves obstruction primarily due to a venous process – thrombosis or phlebitis – an influence on Virchow’s triad. Secondary BCS arises from the compression or invasion of the hepatic vein (HV) and/or the

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inferior vena cava (IVC) by a lesion outside of the HV, for example, malignancy.² The level of obstruction found in BCS varies globally. In Asia, South Africa, India, and China, IVC obstruction remains the most common level of obstruction. In contrast, in Western countries, HV obstruction remains the most common site of obstruction.⁴⁻⁶

HIV-infected patients are at increased risk of both arterial and venous thromboembolism. This occurs more frequently in patients with a low cluster of differentiation (CD4) count and those who are virologically unsuppressed,⁷ making BCS a differential that the attending clinician must consider in any HIV-infected patient presenting with ascites. Co-infection with TB has also been associated with a higher risk of pericarditis in patients living with HIV (PLWH) infection.^{8,9}

Both CP and BCS share a remarkably similar, often indistinguishable, clinical presentation, with ascites and hepatomegaly being the standard features.¹⁰ Chest radiograph may or may not display pericardial calcifications (only about a quarter of chest radiographs illustrate this feature).¹¹ Hence, correct diagnosis of these conditions will require a high index of clinical suspicion. We describe a patient who presented with features of disproportionate fluid overload in the setting of HIV infection and a history of previous TB and the subsequent diagnosis of CP with BCS due to right atrial thrombosis.

Written informed consent and permission for this case report was obtained from the patient. Medical manager of King Edward VIII Hospital and the Department of Health Research Unit.

Case

A 31-year-old female, a mother of 2, originally from Mtubatuba in KwaZulu-Natal, presented to the King Edward VIII Hospital Acute Medical Unit with a 2-month history of progressive dyspnoea (New York Heart Association (NYHA) grade 4)¹² and generalised body swelling. She reported that her abdomen became enlarged within a few days before her presentation.

She reported no chest pain, palpitations, constitutional symptoms, nor rheumatic heart disease as a child. She was HIV-infected, diagnosed in 2019, and was currently on a tenofovir, emtricitabine, and efavirenz fixed-dose regimen of anti-retroviral therapy (ART). She also reported a previous history of pulmonary TB infection diagnosed on sputum testing in 2012, for which she had completed 6 months of anti-TB therapy. She reported no prior hospital admissions and related that she had regular menses, no history of pregnancy losses, nor a current or previous hormonal contraceptive history. She is of sober habits with no use of traditional or herbal treatment.

On examination, she was clinically fluid overloaded. She had mild pallor, no jaundice, no clubbing, and was acyanotic. She had minimal lower limb swelling – grade 1. She had no stigmata of advanced HIV infection or had any features consistent with chronic liver disease. She had distended pulsatile

neck veins, with a markedly raised JVP (confirmed by a positive abdominal-jugular reflex) to the angle of the jaw – with notable CV waves. She was tachycardic (112 beats/min) with a pulse that was irregularly irregular in rhythm, consistent with atrial fibrillation (AF). Her blood pressure was within normal limits. She had a right-sided S3 gallop (possible pericardial knock) with a grade 3/6 pansystolic murmur of tricuspid regurgitation at the left sternal border with no clinical signs of pulmonary hypertension (no loud pulmonic component of second heart sound (P2), no parasternal heave and no loud P2). She was in respiratory distress with a tachypnoea (24 breaths/min); however, maintaining an oxygen saturation of 96% on room air. No crepitations were heard on auscultation of her chest. Her abdomen was grossly distended with ascites (notably out of proportion to the lower limb peripheral oedema) together with a tender right upper quadrant. At this point, we entertained a differential diagnosis that explored the presentation of disproportional ascites to peripheral oedema.

Our considerations therefore included the following:

- right heart failure (RHF) secondary to post-TB bronchiectasis with cor pulmonale,
- CP,
- intra-abdominal malignancy or disseminated TB,
- restrictive cardiomyopathy,
- hepatic outflow obstruction,
- chronic liver disease,
- superior vena cava syndrome, and
- right atrial myxoma.

Laboratory investigations (Table 1) revealed liver function tests, suggesting a predominantly infiltrative pattern.

Her chest radiograph showed patchy lower lobe pulmonary opacification with small bilateral pleural effusions – not amenable to pleurocentesis, with no pericardial calcifications being noted (Figure 1).

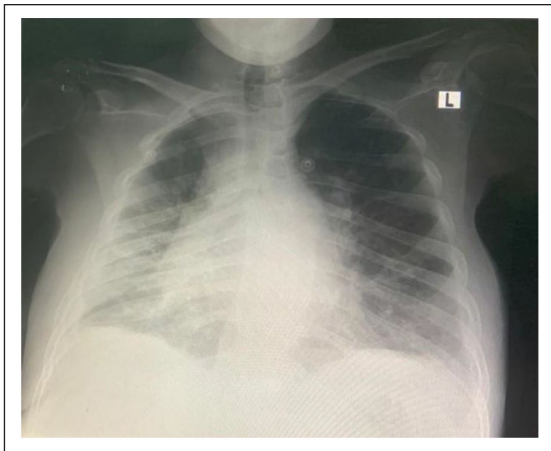
Her echocardiogram showed right atrial dilation with evidence of a thrombus, with mild to moderate tricuspid regurgitation. The cardiac contractility was mildly impaired with a significant septal bounce. The IVC appeared to be dilated with a spontaneous echo-contrast noted. There was respiratory variation, with a preserved left ventricular ejection fraction of 47%.

The abdominal ultrasound confirmed gross ascites with hepatomegaly measuring 16 cm with no focal lesions. The portal vein diameter was 1.2 cm (reference range=0.7–1.5 cm). A Doppler ultrasound of the IVC and HV showed notable sludging, suggesting a thrombus within the aforementioned vessels.

A computed tomography (CT) scan of the abdomen and chest confirmed CP – pericardial thickening and calcifications were demonstrated. The right atrium was enlarged with early reflux into the IVC which was attributed to the features of RHF (arrows in Figure 2).

Table 1. Laboratory investigations.

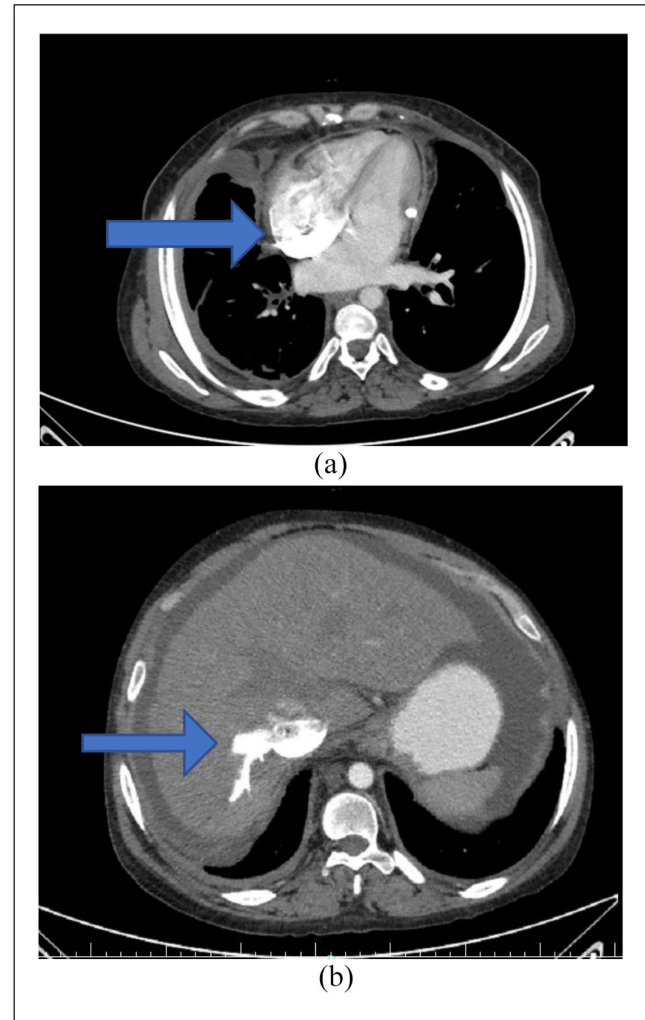
	Value	Range
Total protein	87	60–78 g/dL
Albumin	33	35–52 g/dL
Total bilirubin	17	5–21 $\mu\text{mol/L}$
Conjugated bilirubin	14	0–3 $\mu\text{mol/L}$
Alanine transaminase	14	7–35 U/L
Alkaline phosphatase	283	42–98 U/L
Gamma-glutamyl transferase	429	<40 U/L
Serum albumin to albumin gradient (SAAG)	12 g/L	
Mycobacterium tuberculosis polymerase chain reaction (PCR)	Negative	
Random blood glucose	5 mmol/L	
International normalised ratio (INR)	1.2	<1.3
Full blood count	Normal	
Renal functions	Normal	
Auto-immune screen	Negative	
CD4 count	208 cells/uL	
HIV viral load	Lower than detectable	
Urine LAM	Negative	
Urine dipstick	Negative	
Child–Pugh score	8	

**Figure 1.** Chest radiograph.

The final diagnosis made was that of an HIV-infected patient on ART with a previous history of treated pulmonary TB now presenting with features suggestive of:

- CP with a possible BCS (acute 2-week history of rapidly enlarging abdomen with sludge/thrombus in both HV and IVC) associated with AF.

The patient was continued on ART and started on anti-coagulation in the form of Clexane® 60 mg BD subcutaneous and warfarin 5 mg daily orally once the AF was detected. She

**Figure 2.** Features of constrictive pericarditis with right atrial enlargement (a) and markedly distended intrahepatic IVC (b).

was also commenced on anti-TB treatment before referral to the cardiothoracic department.

Discussion

RHF and cor pulmonale is a common diagnosis in low-to-middle income countries which have a high burden of TB resulting in post-TB bronchiectasis.^{13,14} This case serves to highlight the importance of maintaining a high level of clinical suspicion in patients presenting with ascites that is out of proportion in size to peripheral oedema.

CP can often be diagnosed by an astute clinical examination and must be suspected in any patient who has had a previous history of TB. This condition can be treated surgically, and it is imperative that the diagnosis is actively searched for in patients presenting with disproportionate ascites.

BCS is also a diagnosis that must be sought after in patients presenting with disproportionate ascites, here mainly for the evidence of thrombus occluding the HV and/or IVC,

which will require anti-coagulation. The majority of patients diagnosed with BCS have been shown to either have an obstruction in the IVC (in developed countries) or the HV (Western countries).^{4–6} We, however, showed in our patient that the venous obstruction was noted in both the HV and the IVC. Combined HV and IVC obstruction was found in 10% and 21% of BCS patients in studies conducted in Egypt by Ahmed et al.¹⁵ and Sakr et al.,¹⁶ respectively. Darwish Murad et al.¹⁷ in their study of patients treated at tertiary hospitals in France and the Netherlands found that 31% of patients with BCS had combined HV and IVC obstruction.

It is unusual for both these conditions to co-occur in the same patient. There does not exist much literature that compares cases of co-existent CP and BCS. A case series published in 1991 in the *Journal of Clinical Gastroenterology* cited three case reports in which patients had presented clinically with what initially appeared to be BCS, presenting with clinical evidence of fluid overload with hepatomegaly.¹⁰ The abdominal ultrasound findings showed evidence of HV and IVC dilation, while their chest radiographs showed no pericardial calcifications. All patients underwent an invasive liver biopsy which showed centrilobular haemorrhagic necrosis with sinusoidal dilatation – which is a histological feature of BCS but not specific to the condition. Any condition which results in increased pressure to the IVC/HV – for example, congestive heart failure, will cause this histological feature. Ultimately, the diagnosis of CP in these patients was made on echocardiographic and CT findings, and in the case of one of these patients, cardiac catheterisation. The take-home message of that case series was evidently to caution against an invasive procedure that ultimately does not point to a definitive diagnosis. These case reports also serve to allude to the clinical similarity between CP and BCS.

South Africa has the highest prevalence of HIV infection globally with more than 7 million positive individuals.¹⁸ The presence of HIV infection, especially those with low CD4 and unsuppressed viral load, should alert the attending clinician to search for increased thrombo-embolic events either in the form of pulmonary embolism or veno-thromboembolism like BCS. HIV infection increases the risk for developing TB by 10-fold and TB pericarditis by 40%–75%.⁹ Here again, the attending clinician should always have a high index of suspicion for CP in HIV-infected patients.

Caudate lobe hypertrophy can be seen in 80%–91% of cases of Budd–Chiari, especially later in the disease process.³ We postulate that the absence of caudate enlargement, elevated bilirubin and clinical jaundice in our patient indicated that she presented early in the process of the BCS. The presence of thrombosis may be related to the CP and back-flow into the liver; however, the overall picture of thrombosis in the right atrium or IVC is not a typical pattern found in CP and RHF.¹⁹

AF can be present in RHF, cor pulmonale, and CP but less so in liver diseases.²⁰ This makes it challenging to use the

presence or absence of AF as a differentiating factor between RHF, CP and cor pulmonale.

Both CP and BCS can be easily missed if the attending clinician treats the patient-reported in our presentation as a cor pulmonale secondary to post-TB bronchiectasis. This would delay the surgical pericardiectomy and/or anti-coagulation required for the patient.

Conclusion

This case serves to highlight the importance of having a high index of clinical suspicion for CP and BCS in HIV-infected patients, and to allude to the clinical similarity between CP and BCS, especially in those patients presenting with disproportional ascites.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Ethical approval

Ethical approval to report this case was obtained from the King Edward VIII Hospital Management and from the Department of Health KwaZulu-Natal Provincial Health Research and Ethics Committee.

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Informed consent

Written informed consent was obtained from the patient for their anonymised information to be published in this article.

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