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CASE REPORT
Heart failure

An eye-opening case report of constrictive pericarditis

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Background Constrictive pericarditis is characterized by the encasement of the heart by a stiff pericardium leading to impaired diastolic function, which ultimately leads to congestive heart failure.

Case summary We report a case of a young woman, who first presented to the ophthalmologist with the sudden appearance of floaters and vision reduction. Eventually, invasive haemodynamic assessment led to the diagnosis of constrictive pericarditis leading to venous congestion.

Conclusion Understanding the pathophysiology and integrating the results of invasive and non-invasive diagnostic work up is important in making this challenging diagnosis.

Keywords Constrictive pericarditis • Retinal detachment • Congestive heart failure • Case report

ESC Curriculum 6.3 Heart failure with preserved ejection fraction • 6.6 Pericardial disease

Learning points

- Floaters and vision loss can be presenting symptoms of venous congestion.
- If a non-invasive evaluation is inconclusive or discordant with clinical findings suggesting constrictive pericarditis, haemodynamic assessment by cardiac catheterization should be performed.

Introduction

The normal pericardium is a double-layered sac that consists of fibroelastic tissue and contains a small amount of fluid. By exerting a restraining force, the pericardium prevents sudden dilatation of the cardiac chambers, restricts the anatomic position of the heart, and likely delays the spread of infection. Constrictive pericarditis (CP) is defined as an impedance to diastolic filling caused by loss of elasticity of the pericardium.[1] Idiopathic and/or viral pericarditis are the predominant causes in Western countries, followed by post-cardiac surgery and post-radiation.[2] The diagnosis is challenging because of non-specific symptoms and clinical signs as well as broad differential diagnosis [i.e. restrictive cardiomyopathy (RCM), cor pulmonale, and valve disease]. Most patients display elevated jugular venous pressure, and this pressure fails to decline during inspiration (*Kussmaul's sign*).[3] A stiff pericardium restricts inflow into the

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ventricles: in early diastole, there is initially a high driving pressure but when the elastic limit of the pericardium is reached, an abrupt reduction in the pressure leads to a restrictive diastolic filling pattern.[4] The result is an increase of diastolic pressure in all four chambers at similar levels with a dip and plateau pattern seen on the invasive pressure curves. Pericardiectomy with complete decortication is the treatment of choice.[1] Sodium restriction and diuretics are useful in the preoperative period.

Timeline

17/12/2019	Ophthalmologist: floaters and reduced vision • Bilateral exudative retinal detachment and dilated episcleral veins • Lumbar puncture: exclusion Vogt-Koyanagi-Harada syndrome • MR angiography brain: possible carotid cavernous fistula • Lab results: exclusion auto-immune disorders and viral diseases • Start corticosteroids
26/12/2019	Rheumatologist: exclusion of auto-immune disorders, normal chest X-ray
02/01/2020	Cerebral angiography: exclusion of carotid cavernous fistula
23/01/2020	Ophthalmologist follow-up: • Bilateral exudative retinal detachment and dilated episcleral veins: no changes • Stop corticosteroids
31/01/2020	Full body PET/CT scan: • Hepatomegaly, ascites: *nutmeg liver* • Slightly thickened pericardium
10/02/2020	CT pulmonary angiography showed no pulmonary embolism
10/02/2020	Gastroenterology: • Abdominal paracentesis: dry tap • Lab results: slightly elevated gamma-glutamyl transferase • Normal doppler sonography of the liver • Work up for liver failure: negative
15/02/2020	Cardiology: • No signs of congestion, minimal exertional shortness of breath • Rest ecg: sinus rhythm, incomplete right bundle branch block • Echocardiography: no restrictive filling pattern, dilated vena cava inferior with reduced collapsibility
09/03/2020	Cardiology and gastroenterology: planning invasive pressure curves to rule out shunts, pulmonary hypertension, or constrictive pericarditis
12/03/2020	Right heart catherization: • High end-diastolic pressures • Square root sign • Necking of the vena cava superior entering right atrium
05/05/2020	Cardiac surgery consultation: indication for pericardiectomy, low operative risk score
02/07/2020	Pericardiectomy, specimen examination showed fibrosis and further no abnormalities
01/07/2021	Cardiology and ophthalmologist • Normal cardiac follow-up • Regression of retinal detachment

Case description

A 35-year-old woman presented to the ophthalmologist because of the appearance of many specks that seem to drift through her eye field (floaters) as well as reduced vision. Her medical history was unremarkable. Examination revealed bilateral exudative retinal detachment and dilated episcleral veins for which she received a broad diagnostic work up, as shown in the timeline above, including chest X-ray (*Figure* 1), lumbar puncture, brain magnetic resonance imaging and cerebral angiography showing initially no causal disease. Extensive blood analysis was normal besides mild elevated gamma-glutamyl transferase (51 U/L, reference value 10–40 U/L). She had no shortness of breath; she only mentioned that exercising has always been difficult due to poor endurance. Clinical examination was normal, there were no clear signs of elevated central venous pressure and no peripheral oedema. Following parameters are noted: blood pressure 126/79 mmHg, heart rate regular at 75 bpm, body mass index of 26 kg/m^2. An additional PET/CT scan to rule out a paraneoplastic phenomenon showed congestive hepatomegaly, also referred to as a *nutmeg liver* and associated perihepatic fluid (*Figure* 2). The pericardium was slightly thickened and calcified (*Figure* 3). Although a pericardial thickness of more than 4 mm on CT scan is predictive of CP, up to 20% of patients with normal pericardial thickness can still develop CP so it yields low sensitivity.[5] Ultimately, the patient was referred to the cardiology department: transthoracic echocardiography documented dilatation of the inferior vena cava with reduced (<50%) respiration collapsibility, but no other direct or indirect signs of pulmonary hypertension and diastolic function was classified as normal based on mitral inflow pattern (E/A ratio of 2), normal deceleration time and normal peak early diastolic mitral annular velocity (*Figure* 4). Pulmonary embolism was excluded by CT pulmonary angiogram and there was a pulsatile flow in portal vein on Doppler echography. Owing to the findings of venous congestion throughout the body and the discordant non-invasive evaluation, the patient was scheduled for haemodynamic assessment by right heart catheterization. This showed high end-diastolic, equalized (within 5 mm Hg) pressures in all four chambers (21 mm Hg). The cardiac output was normal (5.8 L/min). The waveform of pressure measurements in diastole followed a dip and plateau pattern (square root sign) suggestive of pericardial constriction (*Figure* 5). Necking of the superior vena cava entering the right atrium through the pericardium was also present (*Figure* 5). The patient was planned for surgical pericardiectomy with the specimen examination revealing no abnormalities apart from some element of fibrosis. During the following months, the patient's symptoms of floaters and reduced vision resolved.

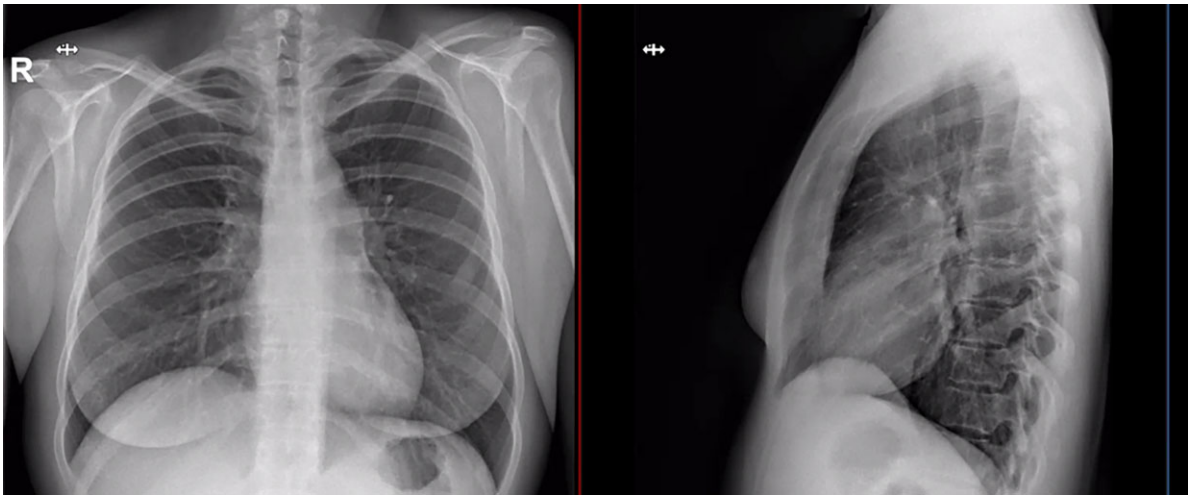

Figure 1 Chest X-ray.

Figure 2 Full body PET/CT scan: nutmeg liver with perihepatic fluid.

Discussion

Patients with CP typically present with symptoms related to fluid overload and/or reduced cardiac output.[6] Floaters and vision loss as presenting symptoms are rarely described in the current literature. The time between first presentation and pericardiectomy was 7 months, highlighting the challenging diagnostic process. The *nutmeg liver* seen on computed tomography pointed in the direction of congestive heart failure. This appearance is most pronounced in the periphery of the liver during the portovenous phase and returns to normal in the delayed phase. It is caused by perfusion abnormality; the areas of lower attenuation correspond to areas with decreased portal blood flow due to hepatic congestion.[7] Various conditions can give rise to a *nutmeg liver*, including right ventricular failure, CP, valvulopathy, pulmonary hypertension, congenital heart diseases, hepatic veno-occlusive disease, and Budd–Chiari syndrome. In 1896, the concept of *Pick's disease* was introduced, which represents patients with CP who had concomitant ascites and hepatomegaly.[3] Invasive hemodynamic evaluation is occasionally needed to confirm the diagnosis, particularly in patients with suboptimal or non-diagnostic echocardiographic findings in whom cross-sectional imaging is non-specific.[3] Typical haemodynamic findings in CP include the square root sign in the RV and LV diastolic pressure tracings, often with an absent a-wave and equalization of the diastolic plateau pressure tracings in the four heart chambers.[6] Differentiation with RCM can be challenging and is often based on pressure differences during respiration. On inspiration, the intrathoracic pressures decrease while LV diastolic pressure in CP remains high, the net effect is a reduction in the pressure gradient for ventricular filling. Decreased left ventricular filling during diastole allows more room for right ventricular filling, which leads to a septal shift/bounce and an increase in right-sided inflow.[6] The opposite sequence occurs in expiration. These findings are not present in RCM in whom the left ventricular and right ventricular pressures are concordant during the respiratory cycle. Unfortunately, our patient was anxious during the procedure for which she received sedatives leading to impaired Valsalva testing, ventricular interdependence could thus not be tested. Serum (NT-pro)BNP levels could also be useful as a differentiating biomarker. In CP, there is no stretch on the myocardium because of the pericardial constrictive effect. In contrary, in RCM, the restrictive effect is in ventricular myocardium which produces significant atrial stretch and higher (NT-pro)BNP levels.[8] The cause of CP is undetermined in the presented case although, an asymptomatic or forgotten bout of viral pericarditis may have been the inciting event. The risk of progression to CP is related to the aetiology: low (<1%) in viral and idiopathic pericarditis, intermediate

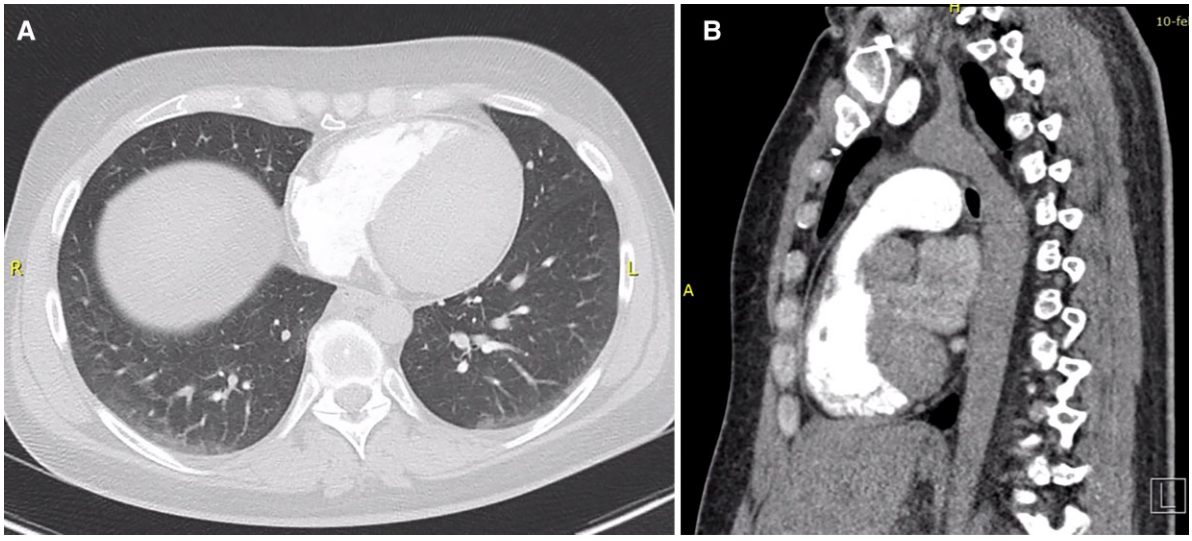

Figure 3 Full body PET/CT scan: pericardium is slightly thickened and calcified.

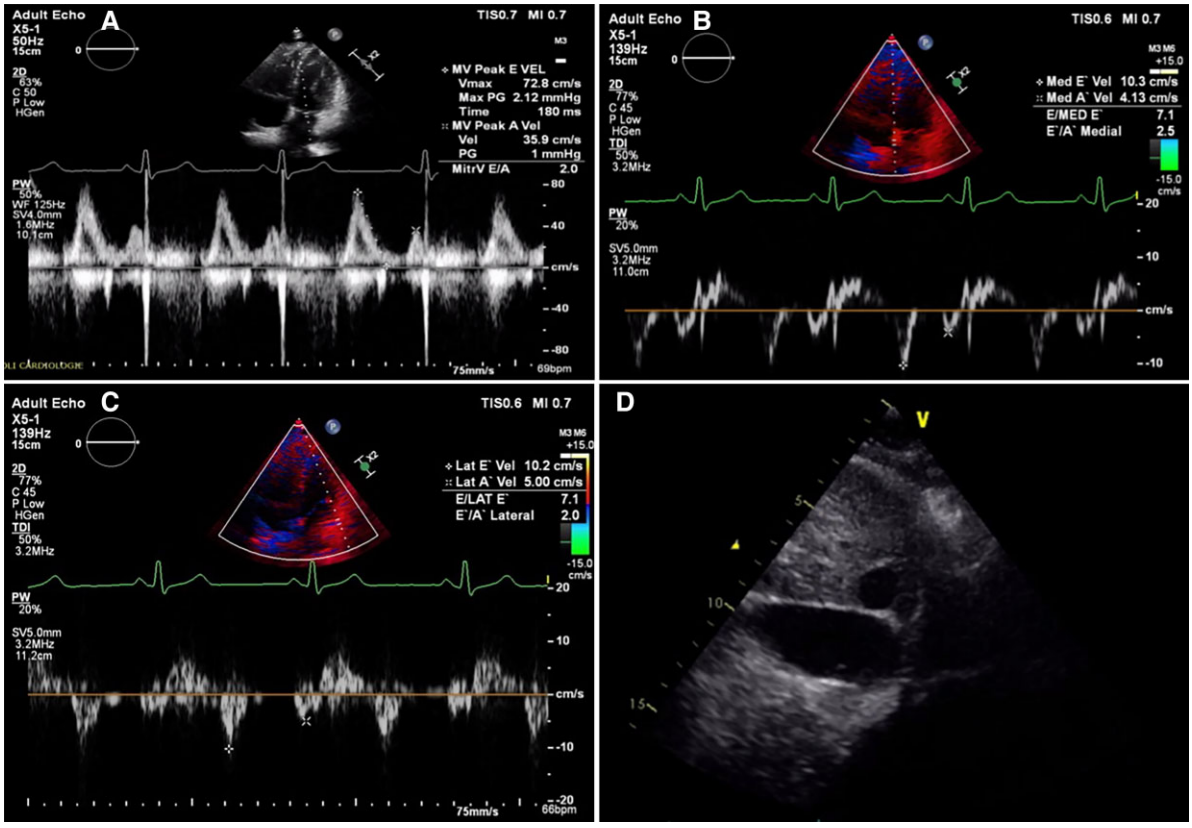

Figure 4 Echocardiography showing slightly (supra)normal diastolic function (E/A ratio 2, normal deceleration time end normal peak early diastolic mitral annular velocity (medial and lateral e'), dilated inferior vena cava.

(2–5%) in immune-mediated pericarditis and neoplastic pericardial diseases and high (20–30%) in bacterial pericarditis, especially purulent pericarditis.[2,6] The aetiology of CP has an important impact on long-term survival after successful pericardiectomy and is best for patients with idiopathic constriction. In one series, the 7-year survival rate after surgery for patients with idiopathic CP was 88%.[9]

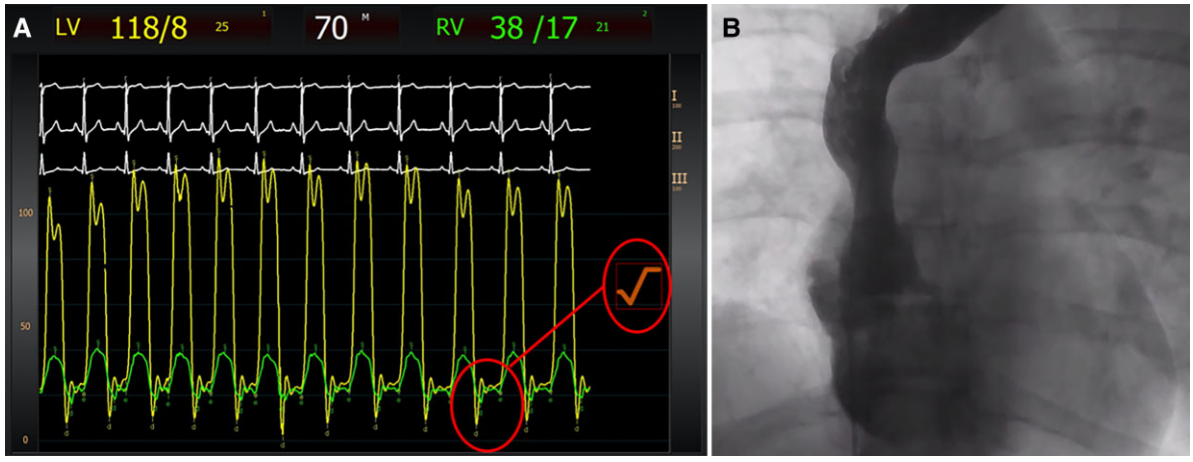

Figure 5 Waveform of pressure measurements showing square root sign (circle) left, necking of the superior vena cava entering right atrium right.

Conclusions

Constrictive pericarditis should always be considered in patients with signs of chronic fluid congestion. Because of the atypical presentation, there is often a significant delay in diagnosis and treatment. In equivocal cases, where non-invasive evaluation is inconclusive or discordant with clinical findings, haemodynamic assessment by cardiac catheterization should be performed.

Lead author biography

Sebastiaan Dhont graduated in Medicine from Ghent University in 2008. He is currently a cardiology fellow with special interest in heart failure, echocardiography, and cardiac devices.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

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

Consent: The patient gave her consent for images or other clinical information relating to this case to be reported in a medical publication.

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Ethics approval and consent to participate available.

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