Reminder of important clinical lesson

Shortness of breath in a CF patient: not always the chest

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The authors report the case of an adult with cystic fibrosis (CF) presenting with an episode of acute shortness of breath during an inpatient stay for infective exacerbation of CF. The treatment of bronchodilators and steroids for presumed airways disease was not effective and on review of the chest x-ray, it showed some evidence of pulmonary oedema. An echo showed a dilated left ventricle and globally reduced function. Serology came back positive for enterovirus which was in keeping with a diagnosis of myocarditis.

BACKGROUND

I think this case illustrates that when patients have a chronic condition, it is often easy to focus on that as the cause of a clinical presentation. In most cases, this may be the case but if things do not seem to fit then you must always think of differentials.

CASE PRESENTATION

This is the case of a middle aged patient with cystic fibrosis (CF) with co-existing diabetes, pancreatic insufficiency, asthma and vocal cord dysfunction.

She was admitted to the CF unit with a 1 week history of cough productive of thicker sputum, increased shortness of breath on exertion (with increased use of her home oxygen), and chest discomfort that she put down to sputum retention. Her abdomen was also distended, and with her

previous history of distal intestinal obstruction syndrome, she had commenced gastrografin at home. No fevers were reported.

These symptoms were typical of her infective exacerbations of CF and she was commenced on intravenous antibiotics and had intensive physiotherapy.

Blood tests revealed: C reactive protein 1, white cell count 8.1, neutrophils 4.11, haemoglobin 12.5, Na 136, K 5.1, Ur 2.8, Cr 59

Chest x-ray (CXR) (figure 1) showed apical fibrocystic changes unchanged from previous x-rays. Abdominal x-ray showed no dilated bowel loops.

Over the next 2 days, her symptoms started to resolve and she felt much better. She did think that her ankles had become more swollen and she had some pitting oedema to both ankles.

She was given a day leave, but returned later that day very unwell. She was profoundly short of breath, with severe paroxysms of coughing and oxygen saturations of 69% on air. She complained of no chest pain.

Figure 1 CXR on admission.

Figure 2 CXR at time of worsening symptoms.

Figure 3 ECG after episode during the night.

Figure 4 ECG a few hours later.

These symptoms were similar to her previous attacks of asthma with mucous plugging.

Auscultation revealed widespread wheeze with scattered crepitations and treatment for bronchospasm was started: she was given hydrocortisone and back-to-back nebulised bronchodilators to which she seemed to respond.

CXR showed no pneumothorax but there were some increased vascular marking throughout the lung field (figure 2). No ECG was done at this time.

She had two further attacks of acute shortness of breath the following morning which were only partially resolved by nebulisers. The crepitations in the chest were more pronounced than those heard on admission, and there was evidence of pitting oedema to lower one of three legs. This combined with the increased interstitial lung markings the differentials of pulmonary oedema and pneumonitis were considered. The CXR was re-reviewed and Kerly B lines were identified.

ECG at this time showed a sinus tachycardia, biphasic T waves in V2 and V3 and some lateral lead ST flattening. Serial ECGs performed throughout the day showed progression to widespread T wave inversion in all chest leads (figures 3 and 4).

Bedside echocardiography showed a dilated heart with severe left ventricular systolic dysfunction. Troponin T came back at 0.06 and rose to a peak of 0.11 the next day. A creatinine kinase was not requested.

She was reviewed by the cardiologists who commenced perindopril and furosemide. Although her breathing dramatically improved over the next few days, she complained of central chest pain, diffuse in nature, worse on lying flat and relieved by sitting forward. The pain was not exertional and was minimally relieved by glyceryl trinitrate spray. It was felt that this was pericarditic in nature, but she went on to have a coronary angiogram which showed normal coronary arteries.

The acute onset of the symptoms with a preceding viral prodrome seemed in keeping with a myocarditis. Serology subsequently came back positive for enterovirus (commonest cause of viral myocarditis).

DIFFERENTIAL DIAGNOSIS

- Main differentials are respiratory when a CF patient presents with shortness of breath
- Respiratory
- Exacerbation of CF
- Exacerbation of asthma

- ▶ Mucous plugging
- ▶ Pneumothorax (prior to CXR in this case)
- ▶ Pneumonia
- ▶ Pneumonitis
- ▶ Pulmonary embolus (rare in CF)
- ▶ Cardiac
- ▶ Heart failure

OUTCOME AND FOLLOW-UP

Her clinical symptoms further improved over the next 7 days. Her ECG normalised and on echocardiography 4 months later her left ventricular function had dramatically improved.

On follow-up echocardiography 1 year later the left ventricular function had normalised.

The patient in this case report has subsequently passed away of a CF related complication. At the time her cardiac function was normal and was unrelated to the cause of death.

DISCUSSION

There is a known association between CF and the development of right ventricular dysfunction.¹ The aetiology of this is thought to be due to raised intrapulmonary artery pressure.² Although some studies have shown abnormal left ventricular systolic function in patients with CF,^{3 4} there is no direct association between CF and myocarditis.

Enteroviruses are a group of single-stranded sense RNA viruses, very common in infants and children. They are responsible for a myriad of clinical syndromes, including hand-foot-and-mouth disease, herpangina, myocarditis and pleurodynia. The incubation period is usually 3–10 days.

The enterovirus enters the human host through the gastrointestinal or respiratory tract. The cell surfaces of the gastrointestinal tract serve as viral receptors, and initial replication begins in the local lymphatic gastrointestinal tissue. The virus seeds into the bloodstream, causing a minor viremia on the third day of infection. The virus then invades organ systems, causing a second viremic episode on days 3–7.

People of all ages, including adults, older people and young people, are at risk of manifesting symptoms of enteroviruses.

Unfortunately, no specific antiviral medication or treatment is available for an enteroviral infection. The best care is provided through supportive measures. Fluid hydration and antipyretics are the mainstays of care for a viral syndrome.

More than 90% of enteroviral infections are either asymptomatic or cause a non-specific febrile illness. A wide range of symptoms is observed, but most cases include fever, a viral prodrome and gastrointestinal symptoms.

Patients with myocarditis or pericarditis report chest pain, fatigue and dyspnea on exertion. These symptoms can progress to dysrhythmia and heart failure. The most common cause of cardiac involvement is coxsackievirus group B5 infection.

Exacerbations of CF can be infective in origin. One recent study in Leeds showed the annual incidence of admissions per patient associated with viral infection was 4.9%.⁵

However, enterovirus has not been identified as a major source of exacerbations. There are no case reports found in the literature concerning myocarditis in a patient with CF. This case illustrated that CF patients like all others can present with any condition.

The fact that she already had the diagnoses of asthma and CF did in part delay the diagnosis of myocarditis due to their similarity in signs and symptoms. It illustrates the importance of thinking laterally when a patient with a known chronic condition does not respond to the usual medication that is, there another pathology.

At the time of this admission, the hospital had no cardiac MRI facility on site, but if available this would have been a useful investigation. Given the atypical nature of the presentation for coronary artery disease, a cardiac MRI would have been preferable to angiography. Diffuse changes are seen in myocarditis on MRI and would have helped confirm the diagnosis.

That said, one can usually diagnose myocarditis with a good history and examination with collaborative ECG and echo findings.

Learning points

- ▶ Always consider a list of differentials in patients with chronic illnesses.
- ▶ If the treatment is not working always reconsider the differentials, it maybe you are treating the wrong thing.

Competing interests None.

Patient consent Not obtained.

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