19. FEVER, DYSPNOEA AND A RAISED CRP: JUST ANOTHER CHEST SEPSIS?

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Introduction: Autoinflammatory conditions can arise in patients presenting on the medical take. Patients presenting with an inflammatory response, fever and organ dysfunction are usually (and appropriately) managed as sepsis until proven otherwise. However, a suspicion should be maintained for alternative diagnoses when there is no improvement with conventional antimicrobial therapy. We present a case of a young woman presenting with polyserositis with a wide differential diagnosis including infection, autoimmunity and malignancy. She was treated successfully with colchicine. Subsequent genetic analysis at the Royal Free hospital for fever syndromes has not found a pre-described genetic polymorphism.

Case description: A 39-year-old woman presented to A&E with dyspnoea, night sweats, lethargy and right upper quadrant pain. Her past medical history included hypothyroidism and two years of intermittent diarrhoea. This diarrhoea was associated with a raised faecal calprotectin. Subsequent investigation with colonoscopy and MRI small bowel were normal. Other past history included pleurisy felt to be secondary to a lower respiratory tract infection aged 38 and a right knee effusion aged 15. She was born in the UK but had a Greek mother.

Examination revealed reduced breath sounds in the bases which were dull to percuss, normal heart sounds and mild abdominal distension. There was no peripheral oedema. Observations showed oxygen saturations of 96% on air, BP 90/50, HR 120 and a pyrexia of 38˚C. Blood results showed a raised ALT of 100 and CRP 40. She was initially treated with antibiotics for a working diagnosis of atypical pneumonia. Her respiratory failure deteriorated and a CTPA two days later showed bilateral pleural effusions, a massive pericardial effusion with impending tamponade and a right upper lobe PE. A CT CAP show moderate ascites but no evidence of malignancy or lymphoma.

Pericardiocentesis showed a reactive picture with no organisms grown and normal cytology and microbiology of the fluid. Pleural and ascitic fluid were also drained and she was commenced on LMWH for the PE. Virology, microbiological and autoimmune screens were unremarkable. CRP at this stage had risen to 78 as had the ALT to 1400. 6 days after admission, antibiotics were stopped she was commenced on colchicine 1mg bd. Within 48 hours she rapidly improved both clinically and biochemically.

Discussion: Since the index admission genetic analysis has shown she is HLA B51 negative and she has no recognised genetic polymorphism associated with a periodic fever syndrome. 20 months since admission she has made a good recovery and is stable on colchicine 1mg od with no recurrence of the pleural and pericardial effusions. She has had one further episode of diarrhoea for 2 weeks associated with a raised faecal calprotectin and red macular rashes on her torso. Anticoagulation for the PE has been stopped.

Polyserositis in the absence of infection or malignancy is uncommon and autoinflammatory and autoimmune aetiologies should be considered. This case highlights the importance of keeping a broad mind in patients presenting in an unusual way or when they do not respond to initial treatment.

Autoinflammatory conditions are characterised by marked inflammation affecting the skin, serosal surfaces and synovium among others. Her history of colitis is intriguing. There are case reports of patients with familial Mediterranean fever presenting with serositis but she was negative for FMF on further genetic testing. Similarly she did not prove to have any of the genes that have so far been recognised as causing a periodic fever syndrome.

We did consider Behçet’s but she denied a history of recurrent oral or genital ulceration or iritis and there was no family history. However Behçet’s can lead to inflammatory bowel disease and this remains an intriguing possibility.
The dramatically improved response to colchicine was remarkable. Within 48 hours a re-accumulated pericardial effusion causing right ventricular strain had disappeared as had clinically apparent ascites. Most autoimmune, infective or malignant aetiologies would not be expected to respond in such a way and a yet undiscovered autoinflammatory syndrome remains a tantalising possibility.

**Key learning points:** Main learning points so far: Maintain a wide differential in patients presenting with polyserositis, especially when there is a poor response to typical management approaches. Consider autoinflammatory disease and take a detailed past medical and family history. Colchicine is first line therapy for recurrent pericarditis. Learning points from the conference: What’s her risk of developing secondary amyloid or other secondary sequelae? How long will she need treatment for and how/when would you wean the colchicine? Is her partial Greek genetic heritage relevant? How common are wild type genetic polymorphisms occurring in autoinflammatory conditions? How frequently is colitis a feature of autoinflammatory diseases? If she becomes unwell again what would your second line therapy be?

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