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Case report

An Unusual Case of Systemic Sarcoidosis with Hepatic Involvement Presenting as Complete Heart Block

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Abstract

Sarcoidosis is a systemic disease of non-caseating granulomas in any organ of the body. Diagnosis is challenging with a wide variety of patterns of involvement and no unifying diagnostic test. In this unusual case we came face to face with these issues when a lady in her mid-forties presented win complete heart block, and shortly after developed significantly deranged liver function tests. After several investigations liver biopsy results showed non caseating granuloma and an DG PET-CT showed active myocarditis in keeping with systemic sarcoid. She had a pacemaker inserted and was started on oral corticosteroids with a weekly reducing dose plus azathioprine. Her liver function deteriorated on reducing the steroids so her dose was re-escalated and a slower wean plan initiated. In this case we discovered the complexities in diagnosing a systemic sarcoid with no pulmonary involvement. And, when planning treatment for this lady it was unveiled that the evidence behind management of hepatic and cardiac sarcoid is extremely limited, but there are some consistent findings from systematic reviews available.

Background

Sarcoidosis is a systemic disease of noncaseating granulomas in any organ. Diagnosis is challenging without one reliable diagnostic test and a wide variety of symptoms as such it is important to consider in the undifferentiated patient [1]. Only around 5% of patients with systemic sarcoid have clinically manifest cardiac involvement, although around 20% of those referred for cardiac imaging have cardiac involvement [2]. Conversely, there can be hepatic involvement in up to 80% of cases of sarcoid. While many of these will be asymptomatic, they can progress to severe liver disease [3].

This case report demonstrates: the challenges of diagnosis of sarcoid and importance of considering this diagnosis in young patients presenting with complete heart block even if they have relatively non-specific cardiac imaging. It also highlights that sarcoidosis can present in a wide variety of organs and even in those presenting with clinically significant sarcoid in specific organs they do not necessarily have pulmonary involvement (although this is the most common presentation affecting > 90% of those with the condition [1]).

Case

A patient in her mid forties presented to the emergency department with a 5 day history of central pressure type chest pain. She had associated shortness of breath and palpitations over the same 5 day period but none preceding this. She had no history of collapse or prior cardiac disease; and no significant family history for sudden unexplained or cardiac death.

Her past medical history was only significant for type 2 diabetes mellitus for which she took just glimepiride (although her last HbA1c was poorly controlled). An ECG demonstrated complete heart block at a rate of 38 beats per minute with a narrow ‘QRS’ complex. She was haemodynamically stable with no clinical syndrome of heart failure.

She had a full panel of bloods requested for electrolytes which demonstrated a raised calcium at 2.73mmol/L along with slightly deranged liver function tests (bilirubin 11Umol/L, ALT 54unit/L, ALP 535 unit/L).

As the patient was clinically stable, she was admitted to the coronary care unit for ongoing monitoring and subsequently underwent a routine pacemaker insertion (within 48 hours of ad-

mission). Her echocardiogram demonstrated a normal left ventricle size and function.

She was subsequently discharged home with plan for an outpatient cardiac MRI to investigate the cause of her complete heart block further in the context of an unclear underlying aetiology. Regarding her deranged liver function tests these were due to be repeated on discharge but the team were not able to contact her to arrange these.

Three months after her initial admission under Cardiology she then re-presented with a two week history of feeling generally unwell with dark urine, pale stools, vomiting, generalised itch and abdominal pain. Her liver tests at this point were significantly deranged demonstrating: bilirubin 148Umol/L, ALT 88unit/L, ALP 998unit/L.

A full non-invasive liver screen including acute hepatitis was sent and a detailed history of travel and over the counter drugs was taken. These did not explain the aetiology of her presentation. She underwent a magnetic resonance cholangiopancreatography (MRCP) which was reported as: intrahepatic biliary tree shows impression of subtle multifocal stricturing affecting both right and left sided intrahepatic ducts. A subsequent endoscopic ultrasound demonstrated: non dilated CBD (common bile duct) with no choledocholithiasis.

She then underwent a liver biopsy. This was reported 6 days later and the pathology showed: portal expansion with fibrosis and prominent non-necrotising granulomas which appear quite florid and coalescing in areas. These contain multinucleate giant cells and scattered lymphocytes. More commonly seen in sarcoidosis, other causes of a granulomatous hepatopathy should also be considered (including tuberculosis).

Following this report the admitting team underwent discussions with local hepatology, respiratory and cardiology in an attempt to confirm the suspected diagnosis of sarcoidosis and agree a treatment regimen.

The respiratory physicians advised a CT thorax and checking the liver biopsy for biochemical evidence of Mycobacterium tuberculosis. The CT demonstrated: multiple tiny nodules throughout both lungs without any convincing distribution pattern. Would be compatible with sarcoidosis in this context. They felt her pulmonary disease did not necessitate treatment for sarcoid.

The pathology team were contacted and a Ziehl Neeson stain was done on the sample followed by a Mycobacterium tuberculosis polymerase chain reaction (PCR). An Angiotensin Converting Enzyme (ACE) blood level was sent on three occasions to the laboratories but was unable to be tested as the sample was too icteric on each occasion.

The patient's liver function tests remained markedly deranged and she had persistent malaise throughout this period. The decision was made by the gastroenterology team to initiate prednisolone at 30mg once daily in the context of highly likely multisystem sarcoid with significant hepatic involvement. She was subsequently discharged home with virtual ward monitoring of bloods and outpatient follow up.

The patient's subsequent liver function tests showed a steady improvement. She underwent a cardiac magnetic resonance imaging (MRI) and fluorodeoxyglucose positron emission tomography scan (FDG-PET) as discussed below.

Investigations

The diagnosis of cardiac sarcoidosis can be challenging to confirm, an initial electrocardiogram and echocardiogram are low cost and easily accessible although can be normal and have non-specific findings [2]. The clinical presentation depends on the location, extent and activity of the granulomatous infiltration. Cardiac MRI and FDG-PET are more sensitive and specific. In a systematic review and meta analysis of diagnostic accuracy [4]: cardiac MRI has a sensitivity of 95% and a specificity of 84%, while FDG-PET has 84% sensitivity and a specificity of 82% for diagnosis of sarcoidosis. Despite this, the absence of late gadolinium enhancement on cardiac MRI does not exclude cardiac sarcoid as early cardiac involvement can be present before any imaging abnormalities [2].

The patient underwent a cardiac MRI scan which had been planned following the diagnosis of complete heart block. The cardiac MRI was reported as: no convincing evidence of cardiac sarcoid. There is possible faint midwall late gadolinium enhancement inferolaterally but this is a pattern more seen in historic myocarditis. If there is concern about cardiac sarcoid it would be useful to correlate with BNP, troponin and consider an FDG-PET.

A subsequent FDG PET CT reported: appearances are compatible with active myocarditis in the context of suspected cardiac sarcoidosis. Probable mildly increased FDG activity in the liver and equivocal right hilar lymph node. Neither of these investigations gave a conclusive diagnosis but both supported the overall clinical picture of sarcoidosis.

The liver biopsy in this case demonstrated: portal expansion with fibrosis and prominent non-necrotising granulomas. A subsequent Ziehl-Neeson (ZN) stain was negative.

In a study of twenty-two patients with symptomatic hepatic sarcoidosis [5] in all cases non-caseating granulomas were present on liver biopsy. In all cases the ZN stain was negative and no fungal hyphae were evident. Damage to the bile ducts and portal tract vessels were evident in some cases. However, it can be challenging to discern sarcoid from primary biliary cirrhosis (PBC) and primary sclerosis cholangitis (PSC); especially as in rare cases sarcoidosis can also cause extrahepatic biliary structuring. There have also been reported cases of caseating granulomas on biopsy which are more commonly associated with a diagnosis of tuberculosis [6].

Overall, the diagnosis of sarcoid is not able to be made with one diagnostic test. It is dependent on: presence of a compatible clinical and radiological presentation, non-caseating granulomas on pathology and exclusion of other diseases with similar findings [1].

Differential Diagnoses

When this patient initially presented there was concern for an infiltrative cardiac disease causing her complete heart block in the context of her young age. This included: sarcoidosis, amyloidosis and haemochromatosis.

When she re-presented three months later with markedly de-

ranged liver function tests she was initially admitted to the surgical unit at which point it was thought her liver function derangement could be associated with her known gallstones but this was subsequently excluded with ultrasound imaging. She was then reviewed by the gastroenterology team and underwent a full non-invasive liver screen. At this point the diagnosis of sarcoid was not immediately considered. She underwent an endoscopic retrograde cholangiopancreatography (ERCP) which did not show choledocholithiasis. In the context that her liver function tests were not improving, she underwent an ultrasound guided liver biopsy. The results of the liver biopsy demonstrated diffuse granulomatous disease concerning for sarcoidosis, and leaving tuberculosis as a possibility albeit a less likely one.

This finding on liver biopsy led to the team re-considering her history and piecing together the clinical situation and investigations we had to that point led to the final diagnosis.

Treatment

On initial presentation the patient was in complete heart block and underwent urgent pacemaker insertion.

The first medical treatment she received for her liver function was ursodeoxycholic acid (UDCA) which she has continued on discharge. We saw no improvement in her symptoms or LFTs on this medication alone. Upon diagnosis of high likely multisystemic sarcoidosis with significant hepatic sarcoid she was initiated on prednisolone 30mg once daily, weaning by 5mg every 7 days to stop. Prior to being seen in outpatient hepatology clinic she was also started on azathioprine at the recommendation of our local hepatology team. As per NICE guidelines indications for active treatment of sarcoidosis are 'dangerous disease and/or unacceptable loss of quality of life'. Oral corticosteroids are the first line treatment. Withdrawal from steroids is advised after 6-12 months if disease is controlled [7].

Outcome and Follow up

Plans were made for this lady to be followed up by our local hepatology and cardiology teams. Unfortunately, her bloods were worsening at the time of her first hepatology clinic appointment 4 weeks post discharge which was attributed to her steroids being reduced too quickly. As such her steroids have been increased again and a much slower wean implemented to allow her azathioprine time to have therapeutic effect. She will be subsequently followed up at our nearest tertiary centre from a hepatology perspective.

Discussion

Pulmonary sarcoidosis is a well-known disease process with steroids being used confidently in its management. However, in terms of evidence based medicine the management of hepatic sarcoid remains an unclear issue. Sarcoid UK quotes that up to 70% of patients with sarcoid have liver involvement, but the vast majority of these will never reach a severity needing treatment or self-resolve [8]. In those who do need treatment oral steroid is again the first line treatment. However, evidence for this is based mainly on clinical experience and retrospective case studies [9]. Sinnanaidu at et performed a systematic review of Case Studies and case reports and proposed a treatment ladder of initiating with oral steroid or UDCA. Though UDCA only seemed to have been effective when patients had cholestasis or pruritus. The recommendation for steroid was to wean over 3-6 months, rather than an immediate dose reduction after the first week. If this first line failed then azathioprine and methotrexate were shown to have the most consistent benefit, with the acknowledgement that azathioprine would be preferred being it less renowned for hepatotoxicity. TNF-α biologics were the third line recommendation [10]. Severe refractory cases may lead to transplantation. There is very little literature on outcomes post this procedure but cases of recurrence have been described [11].

The evidence around cardiac sarcoid is derived from similar level studies. Giblin G T et al quote that The HRS consensus statement makes recommendations for immunosuppression in the setting of myocardial inflammation in the following scenarios: high-grade AV block; frequent ventricular ectopy or non-sustained ventricular tachycardia (VT) and evidence of myocardial inflammation; and sustained ventricular arrhythmias and evidence of myocardial inflammation [12]. Pharmacotherapy escalation follows a similar pattern to the recommendations in hepatic sarcoidosis, with oral steroids being front line and TNF-α agents being the last line. Cheng R, et al [2] recommend consideration of initiating a steroid sparing agent at the same time as commencing corticosteroids.

All in all this was a very interesting case. Her diagnosis of heart block at a young age immediately raised concerns for a multisystemic disease process. There were delays in a solid diagnosis, mainly due to non-specific investigation results. Despite this our patient has left hospital on treatment and is living with a good quality of life at the time of this article being written. The lack of high level evidence for treatment of multisystemic sarcoidosis is surprising, but there seems to be a consistent consensus for treatment escalation no matter which organ is affected.

Learning points

1. Based on current evidence and experiences hepatic sarcoidosis steroid regimes should be tapered over a prolonged period of time (3-6 months)
2. The majority of hepatic sarcoid will never reach a level that require treatment, or resolve spontaneously
3. Cardiac MRI is not always diagnostic for myocardial sarcoidosis with FDG-PET being more specific for diagnosis

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