Acute rheumatic fever: a surprising diagnosis

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Introduction

Acute rheumatic fever has varying presentations which include arthritis, carditis, chorea, subcutaneous nodules or erythema marginatum. Initially these signs may lead to a range of differential diagnoses and varying clinical management especially when presenting in uncommon risk groups. Such was the case in examining a middle-aged man presenting with polyarthritis, as the majority of cases of acute rheumatic fever occur in children aged 5–15 years and in low to middle income countries. Thus, comprehensive history taking followed by appropriate testing for group A streptococcus is essential in confirming diagnosis and guiding treatment.

Discussion

The diagnosis of acute rheumatic fever is guided by the modified Jones criteria, which aim to direct clinicians in this diagnosis and to help minimise overdiagnosis. The criteria require the presence of a preceding group A streptococcal infection, in addition to the presence of two major criteria or one major and two minor criteria, supported by evidence of a group A streptococcal infection by throat culture, rapid streptococcal antigen test, or elevated or rising streptococcal antibody titres (Dajani et al, 1992).

Case Report

A 52-year-old man, with a previous medical history of gout, hypertension and alcohol misuse, presented with a 9-day history of gradually worsening ascending arthralgia of his right wrist, right elbow and bilateral toes, ankles and knees. These joints were hot and swollen, making it difficult and painful to mobilise. Despite his history of gout attacks, he continued to drink more than 4 litres of beer per day and eat red meat. Before presentation, his primary care physician had discontinued his regularly prescribed colchicine and treated this as an acute flare up of gout with methylprednisolone intramuscularly, with minimal benefit.

On presentation, he was treated with indomethacin and colchicine for a possible gout flare, with no improvement. He also received vancomycin for 2 days because there was concern that he may have septic arthritis, having an elevated temperature of 38.8 °C (101.9 °F). He was subsequently started on prednisolone 60 mg tapered over 10 days. Differentials considered included gout, pseudogout, septic arthritis, rheumatoid arthritis, reactive arthritis, myeloma and human immunodeficiency virus (HIV). Joint aspiration revealed no crystals and was negative for bacterial growth. Further analysis revealed absent leukocytosis, elevated erythrocyte sedimentation rate 98 mm/hr (normal range 0–20 mm/hr) and elevated C-reactive protein level 17.7 mg/dl (≤0.5 mg/dl), normal levels of uric acid 5.6 mg/dl (2.6–7.2 mg/dl), negative antibodies (anti-CCP, rheumatoid factor, HLA B-27, ANA, anti SS-A, anti SS-B), and non-reactive hepatitis B and C and HIV.

Upon further history taking, the patient reported a possible tick bite with an upper right tight rash that resolved 2 weeks before the arthralgias. Subsequent testing was negative for Lyme disease, Rocky Mountain spotted fever and *Ehrlichia chaffeensis*. He then reported that his grandson had had pharyngitis, following which the patient and his wife had developed severe malaise, fatigue and myalgias that resolved after a few days. This prompted further investigation, which revealed an elevated anti-streptolysin O titre of 880.3 IU/ml (0.0–200.0 IU/ml) and a normal anti-DNase B titre <78 U/ml (0–120 U/ml). Given these findings, acute rheumatic fever was diagnosed, as the patient had fulfilled the Jones criteria with one major feature (polyarthritis) and two minor features (documented fever and an elevated erythrocyte sedimentation rate and C-reactive protein level). The patient was started on a 10-day course of oral penicillin V for treatment of group A streptococcal infection (Shulman et al, 2012). The patient's symptoms gradually improved, at which point he was discharged to follow up.

How to cite this article:
Feghaly J, Mooradian A. Acute rheumatic fever: a surprising diagnosis. Br J Hosp Med. 2020. https://doi.org/10.12968/hmed.2019.0272

Learning points

- Acute rheumatic fever may have varying presentation and needs to be considered when assessing patients with polyarthritis.
- Various tests can be used for diagnosis of group A streptococcal infection.
- The combined measurement of anti-streptolysin O and anti-DNase B titres demonstrates the highest sensitivity.

Throat culture is the standard test for the diagnosis of group A streptococcal pharyngitis, but only a quarter of patients with acute rheumatic fever will have a positive throat culture (Dajani et al, 1992). This could be because of previous antibiotic use or latency between initial infection and onset of acute rheumatic fever (Burke and Chang, 2014). A second, more practical test is rapid streptococcal antigen testing that allows diagnosis of group A streptococcus within minutes in the clinic, demonstrating sensitivity and specificity >95% (Rogo et al, 2011). Yet despite these advantages, throat culture and rapid antigen testing may be inconclusive as they do not differentiate between chronic pharyngeal colonisation and acute rheumatic fever, thus serological evidence of elevated or rising anti-streptolysin O and anti-DNase B titres is needed (Dajani et al, 1992).

Anti-streptolysin O has been the most widely used antibody test for group A streptococcal having a sensitivity of 72.7% and specificity of 93.2% post-streptococcal disease, whereas anti-DNase B titres had lower sensitivity of 70.5% and similar specificity to anti-streptolysin O of 93.2%. However, combined measurement of anti-streptolysin O and anti-DNase B titres exhibits a sensitivity of 95.5% in identifying post-streptococcal disease (Blyth and Robertson, 2006). Despite their utility, a low titre of either antibody does not exclude a diagnosis of acute rheumatic fever, so high clinical suspicion should prompt repeat testing to observe for rising titres (Dajani et al, 1992).

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