Complete heart block in a 40-year-old man with anti-SSA/Ro autoantibodies

Raja Ezman Raja Shariff, Chiao Wen Lim and Sazzli Kasim

Abstract
Atrio-ventricular dissociation (AVD), including complete heart block (CHB), are far more common in the elderly. We report a rare case of CHB in a 40-year-old man, who tested positive for anti-Ro autoantibodies without systemic features. He had been suffering for giddiness over the previous two months. On arrival, his electrocardiogram revealed high-degree AVD. Upon further history, he mentioned that his 68-year-old mother with systemic lupus erythematosus (SLE) had suffered from similar episodes, requiring a permanent pacemaker implantation. On further investigation, he tested positive for antinuclear antibodies (ANA), anti-SSA/Ro and anti-RNP antibodies. However, from history and clinical examination, he had not manifested any articular, extra-articular or extra-glandular features suspicious of rheumatological conditions. Following a failed trial of intravenous hydrocortisone, he subsequently had a permanent pacemaker implanted himself. Although difficult to ascertain whether our patient suffered from a congenital form of anti-SSA/Ro-related CHB, there is evidence to suggest delayed presentation of CHB in those with anti-SSA/Ro and neonatal lupus syndrome. Anti-SSA/Ro antibodies without systemic features can be present in 3% of the population, although this occurs more commonly in the presence of a confirm diagnosis of SLE, Sjögren’s syndrome or poly- and dermatomyositis. Despite the scarcity of evidence, a trial of steroid-based treatment was attempted prior to subjecting the young patient to a permanent pacemaker and its associated complications. To our knowledge, this is only the second case of isolated anti-SSA/Ro syndrome presenting with CHB reported in the literature.

Keywords
Atrio-ventricular dissociation, complete heart block, autoimmune, anti-Ro, anti-SSA, case report

Introduction
Atrio-ventricular dissociation (AVD), including complete heart block (CHB), are far more common in the elderly. Causes include degenerative disease, myocardial infarctions, myocarditis and medication-induced CHB, among others. We report a rare case of CHB in a middle-aged man, who tested positive for anti-Ro autoantibodies without any other clinical manifestation of autoimmune diseases or vasculitis.

Case report
A 40-year-old man presented to the emergency department following episodes of giddiness and having suffered a fall. He had been suffering for giddiness over the previous two months, but did not seek medical attention prior to presentation. He had no known medical illnesses and was a smoker of 20 pack years. He denied any use of prescription medication or substance misuse. On arrival, his vital signs included a blood pressure of 133/69 mmHg and heart rate of 38 bpm. His cardiorespiratory examination was unremarkable, including for any murmurs or carotid bruits. His electrocardiogram on arrival revealed evidence of high-degree AVD, likely due to CHB (Figure 1).

Upon further history, he mentioned that his 68-year-old mother, who was known to have systemic lupus erythematosus (SLE) for 30 years, suffered from similar ‘spells’, which required a permanent pacemaker implantation a year ago. Due to his relatively young age and significant family history, further investigations were performed (Table 1). He tested positive for antinuclear antibodies (ANA), anti-SSA/Ro and anti-RNP antibodies. However, from history and clinical examination, he

Universiti Teknologi MARA (UITM), Malaysia

Corresponding author:
Raja Ezman Raja Shariff, Universiti Teknologi MARA, Jalan Hospital, Sungai Buloh, Selangor 47000, Malaysia.
Email: rajaezman@gmail.com

Creative Commons Non Commercial CC BY-NC: This article is distributed under the terms of the Creative Commons Attribution-NonCommercial 4.0 License (https://creativecommons.org/licenses/by-nc/4.0/) which permits non-commercial use, reproduction and distribution of the work without further permission provided the original work is attributed as specified on the SAGE and Open Access pages (https://us.sagepub.com/en-us/nam/open-access-at-sage).
had not manifested any articular, extra-articular or extra-glandular features to fulfil the diagnosis of SLE or Sjögren’s syndrome. The patient also underwent coronary angiography to exclude coronary artery disease, which was unremarkable.

Following a rheumatology consult, a trial of intravenous hydrocortisone was commenced for several days to no avail. He subsequently had a permanent pacemaker implanted himself, which was uneventful, and he remains under regular follow-up.

**Discussion**

Anti-SSA/Ro antibodies are often associated with congenital CHB. Despite this known association, the exact mechanism

---

**Table 1. Blood investigations**

| Test                          | Results | Test                          | Results               |
|-------------------------------|---------|-------------------------------|-----------------------|
| Haemoglobin ro/T              | 122     | Hepatitis B serology          | Negative              |
| White cell count (10⁹/L)      | 5.4     | Hepatitis B serology          | Negative              |
| Neutrophil count (10⁹/L)      | 2.7     | Human immunodeficiency virus serology | Negative |
| Lymphocyte count (10⁹/L)      | 2.1     | o-ANCA titre                  | Negative              |
| Eosinophil count (10⁹/L)      | 0.04    | c-ANCE titre                  | Negative              |
| Platelet (10⁹/L)              | 221     | ANA titre                      | Positive 1:320        |
| C-reactive protein (mg/L)     | < 0.5   | ANA pattern                    | Speckled appearance   |
| Erythrocyte sedimentation rate (mm/h) | 12 | SSA/Ro                         | Strongly positive 1:2560 |
| Free thyroxine (pmol/L) (reference 12.0–22.0) | 13.2 | SSB                            | Negative              |
| Thyrotropin (mIU/L) (reference 0.55–4.78) | 3.54 | RNP                            | Positive 1:1280       |
| Sodium (mmol/L)               | 138     | Ro52                           | Negative              |
| Potassium (mmol/L)            | 4.5     | Sm                             | Negative              |
| Corrected calcium (mmol/L)    | 2.32    | Scl-70                         | Negative              |
| Phosphate (mmol/L)            | 0.78    | Jo-1                           | Negative              |
| Magnesium (mmol/L)            | 0.8     | Lupus anticoagulant            | Negative              |
| Urea (mmol/L)                 | 11.3    | Anti-cardiolipin               | Negative              |
| Creatinine (mmol/L)           | 1.14    | Anti-beta2 glycoprotein-I      | Negative              |
| Angiotensin-converting enzyme (<40 nmol/mL/min) | 12 | Protein C                      | Negative              |
| Serum immunoglobulin A (g/L) (reference 0.8–3.0) | 0.9 | Protein S                      | Negative              |
| Serum immunoglobulin G (g/L) (reference 6.0–16.0) | 2.6 | Factor V                       | Negative              |
| Serum immunoglobulin M (g/L) (reference 0.4–2.5) | 0.8 | Complement C3 (mg/dL)          | 1.21 (within range)  |
| Rheumatoid factor (IU/mL) (reference 0–20) | 8 | Complement C4 (mg/dL)          | 0.32 (within range)  |

Positive results are shown in bold.

ANA: antinuclear antibodies
leading to conduction delay remains unknown. Placental passage of maternal autoantibodies damage foetal atrioventricular conduction tissue via various postulated mechanisms, including inflammation and direct channel action, with eventual fibrosis. Although difficult to ascertain if our patient suffered from a congenital form of anti-SSA/Ro-related CHB, there is evidence to suggest delayed presentation of CHB in those with anti-SSA/Ro and neonatal lupus syndrome. Anti-SSA/Ro antibodies without systemic features can be present in 3% of the population, although this occurs more commonly in the presence of a confirmed diagnosis of SLE, Sjögren’s syndrome or poly- and dermatomyositis. However, it remains pertinent to exclude more common causes of CHB in the young, including Lyme disease, sarcoidosis, thyroid disorders, myocarditis, electrolyte imbalances and autoimmune diseases, before concluding anti-SSA/Ro syndrome.

By excluding other causes (especially infection-based differentials), a trial of steroid-based therapy could be administered, as seen in our patient. However, the evidence for immunosuppression in reversing CHB is scarce, with only one other case of anti-Ro syndrome having trialled such therapy to success. In our case, despite failing, we felt it was worthwhile prescribing steroid-based treatment to our young patient prior to subjecting him to a permanent pacemaker and its associated immediate and long-term complications.

Conclusion

Anti-SSA/Ro antibody-related CHB remains a unique entity and a challenge to treat, as evidence and guidelines to management remains limited. To our knowledge, this is only the second case of isolated anti-SSA/Ro syndrome presenting with CHB reported in the literature. A multidisciplinary approach to management remains key, as commitment to standard lifelong therapy such as a permanent pacemaker needs to be heavily evaluated in these often young patients.

Acknowledgements

The authors would like to acknowledge Universiti Teknologi MARA (UiTM) for supporting the submission of the following article.

Authors’ contributions

R.E.F.R.S.: data collection and analysis, drafting of manuscript. L.C.W.: data collection and analysis, drafting of manuscript. S.S.: drafting or manuscript, revision of manuscript.

Availability of data and materials

The data that support the findings of this study are available from UiTM Sungai Buloh, but restrictions apply to the availability of these data, which were used under license for the current study and so are not publicly available. Data are, however, available from the authors upon reasonable request and with permission of UiTM Sungai Buloh.

Conflict of interest

The authors declared no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

Ethical approval

Approved by the Universiti Teknologi MARA (UiTM) Ethics Committee. The manuscript does not report on any animal data or tissue.

Funding

The authors received no financial support for the research, authorship and/or publication of this article.

Informed consent

Written informed consent was obtained from the patients for their anonymised information to be published in this article.

ORCID iD

Raja Ezman Raja Shariff https://orcid.org/0000-0002-5167-5863

References

1. Kusumoto FM, Schoenfeld MH, Barrett C, et al. 2018 ACC/AHA/HRS guideline on the evaluation and management of patients with bradycardia and cardiac conduction delay: executive summary: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines, and the Heart Rhythm Society. J Am Coll Cardiol 2019; 74: 932–987.
2. Ambrosi A and Wahren-Herlenius M. Congenital heart block: evidence for a pathogenic role of maternal autoantibodies. Arthritis Res Ther 2012; 14: 208.
3. Puri S, Pooni P, Mohan B, et al. Pregnancy with SLE and fetal congenital heart block: a case report. Cardiol Res 2013; 4: 126–128.
4. Tam WK, Hsu HC, Hsieh MH, et al. Association of anti-Ro/Sjögren’s syndrome type A antibodies and complete atrioventricular block in an adult with Sjögren’s syndrome. Arch Rheumatol 2017; 33: 225–229.
5. Yang YC, Pata RK and Aung TT. A case of complete heart block with diagnostic challenge and therapeutic dilemma. J Investig Med High Impact Case Rep 2018; 6: 2324709618788110.
6. Jobling K, Rajabally H and Ng WF. Anti-Ro antibodies and complete heart block in adults with Sjögren’s syndrome. Eur J Rheumatol 2018; 5: 194–196.
7. Santos-Pardo I, Martinez-Morillo M, Villuendas R, et al. Anti-Ro antibodies and reversible atrioventricular block. N Engl J Med 2013; 368: 2335–2337.