International Journal of Research in Medical Sciences
Mishra V et al. Int J Res Med Sci. 2023 Oct;11(10):3855-3858
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20233045

Case Report

Complete heart block: an atypical presentation with an atypical diagnosis

Vikas Mishra[1], Trisha Chatterjee[2]*, Chandramouli M.[3]

[1]Department of Cardiology, [2]Department of Medicine, Netaji Subhash Chandra Bose Super Speciality Hospital, Jabalpur, Madhya Pradesh, India
[3]Department of Cardiology, CMC Vellore, Vellore, Tamil Nadu, India

Received: 02 September 2023
Accepted: 18 September 2023

***Correspondence:**
Dr. Trisha Chatterjee,
E-mail: chatterjeetrisha75@gmail.com

ABSTRACT

Congenital heart block is frequently associated with underlying structural congenital heart disease or connective tissue disorders. Neonatal lupus is considered a form of passively acquired autoimmune disease in which maternal autoantibodies, namely, anti-Ro and anti-La, cross the placenta and injure the previously normal fetal heart. Here, a young female, 30 years of age, who presented with atypical cardiac manifestations, was diagnosed with complete heart block. She was evaluated thoroughly for its etiology, including structural, medication, infective, thyroid and autoimmune panel. Antinuclear antibody (ANA) was positive by immunofluorescence method with 1:160 titre and with fine speckled pattern. Anti-SSA/Ro antibody was positive with a titre of 30.20 units. Anti-Smith antibody was also reported to be positive. On further investigations, her mother also had positive ANA by Immunofluorescence method with a titre of 1:80, with anti dsDNA, anti Ro/SSA, anti La/SSB positivity. The patient was thus diagnosed as a case of lupus causing her to present with complete heart block in adulthood. Patient was managed with TPI followed by PPI and was discharged with no complications. Neonatal lupus can cause heart block of varying degrees that may be noted in utero presenting as congenital heart block. SLE presenting as congenital heart block in young adults is a rare finding. This case report reviews an adult female of SLE presenting to the emergency for the first time with atypical cardiac manifestations and diagnosed as a case of congenital heart block with laboratory investigations revealing lupus as the primary etiology.

Keywords: Complete heart block, Congenital, Adult, SLE, Anti-Ro

INTRODUCTION

An atrioventricular block is a loss of the regular functioning of the cardiac electroconductive system connecting the sinoatrial node (SA node) and the ventricles via conduction through the atrioventricular node (AV node). Third-degree AV block indicates a complete loss of communication between the atria and the ventricles.[1] The cause of AV blocks can be including idiopathic fibrosis, structural heart disease, acute ischemic heart disease, medication toxicity, nodal ablation, electrolyte abnormalities, and post-operative heart block such as after surgical or transcatheter aortic valve replacement, drugs, lyme disease and some systemic diseases such as collagen vascular disorders, amyloidosis, sarcoidosis, and systemic lupus erythematosus.[1,2]

The incidence of complete (third degree) congenital heart block is one in approximately 20,000 to 25,000 live births.[3] Congenital heart block is frequently associated with underlying structural congenital heart disease or connective tissue disorders. Neonatal lupus is considered a form of passively acquired autoimmune disease in which maternal autoantibodies to the intracellular ribonucleo-proteins Ro (SS-A) and La (SS-B), cross the placenta and injure the previously normal fetal heart.[4]

The prognosis of congenital complete atrioventricular block (CCHB) is usually considered favorable in adults.[5] Conduction abnormalities associated with lupus, however, uncommonly present in adulthood.

In this study, we will review a young female with no previous symptom, who presented with atypical manifestations and was diagnosed with congenital heart block as the first manifestation of lupus at 30 years of age. The diagnostic criteria of congenital heart block proposed by Yater were applied, namely: heart block established in a young patient by graphic records.[6] There must be some evidence of the existence of the slow pulse at a fairly early age and absence of a history of any infection which might cause the condition after birth: notably diphtheria, rheumatic fever, chorea and congenital syphilis." The ECG criteria used are: the atria and ventricles should be completely dissociated, the ventricular rate (VR) should be slower than the atrial rate, and no captured beats should be present.

CASE REPORT

A 30-year-old Asian female with no known past medical history presented to the emergency department with shortness of breath (NYHA grade III) since 1 week and cough since 3 days. Review of systems was otherwise negative denying chest pain, dizziness, palpitation, or syncope. The patient was not taking any medications. However, she had history of oral ulcers. She had no history of recent travel. On physical examination, the patient appeared comfortable. She was afebrile with blood pressure of 124/68 mm Hg, heart rate of 36/minute, and oxygen saturation of 97% on ambient air. The patient had no jugular venous distension. Auscultation of the heart revealed slow heart rate, but it was regular with normal first and second heart sounds having no murmurs. Auscultation of bilateral lungs revealed clear breath sounds. There were neither skin rash, clubbing, pedal edema.

The patient was admitted to the cardiac intensive care unit in the diagnosis of CHB with profound bradycardia at rest. Her average systolic blood pressure was around 110 mm Hg, and her average heart rate was 30 to 40 beats per minute. She occasionally switched to apparent 2:1 heart block. After 2 days of her hospital stay, with ongoing intermittent complete heart block, patient had an episode of presyncope.

Investigations

Admission electrocardiogram (ECG) showed CHB characterized by AV dissociation with wide QRS (duration=120 ms) escape rhythm, atrial rate of 88/minute, and ventricular rate of 38/minute, with inverted T-waves in anteroseptal leads, poor R-wave progression and LBBB pattern (Figure 1). Chest X-ray was unremarkable. Complete blood count was suggestive of leukopenia. CPKMB was 53 IU/l. Biochemistry panel, Troponin, erythrocyte sedimentation rate, and thyroid function tests were also within normal limits.

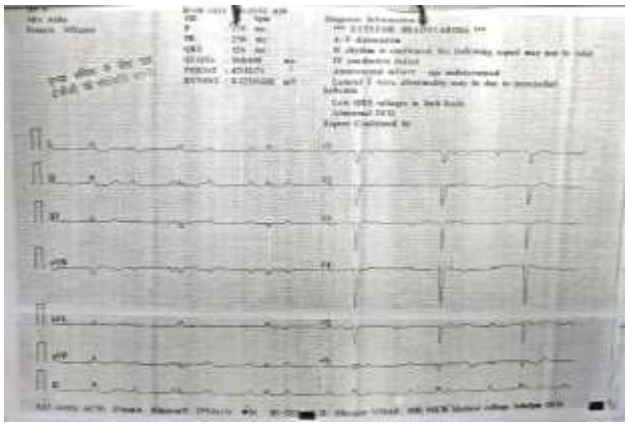

Figure 1: ECG on admission showing complete heart block with heart rate of 38/min.

Transthoracic echocardiogram revealed mitral valve prolapse (anterior mitral leaflet) with mild tricuspid regurgitation. mild eccentric mitral regurgitation, no regional wall motion abnormality and normal LV systolic function with left ventricular ejection fraction 55%.

Antinuclear antibody (ANA) was positive by immunofluorescence method with 1:160 titre with fine speckled pattern. Anti-SSA/Ro antibody was positive with a titre of 30.20 units. Anti-Smith antibody was also reported to be positive. Serum ACE level was in normal limits. Patient was tested for Borrelia Burgdorferi IgM and IgG antibodies by ELISA which turned out to be negative. Patient has normal ESR, CRP, complement levels with negative Anti dsDNA.

On further investigations, her mother also had positive ANA by immunofluorescence method with a titre of 1:80, with anti dsDNA, anti Ro/SSA, anti La/SSB.

Treatment and outcome

Patient was initially kept on oral deriphylline and isoprenaline. With ongoing intermittent CHB and following an episode of presyncope, under all aseptic conditions, right Internal jugular venous access was obtained using 6F introducer sheath and a bipolar temporary pacing catheter (6F from St Jude Medical) was advanced though this sheath from jugular vein to IVC to right atrium and finally into right ventricle at which point, temporary pacing was achieved. The pacer was then advanced additional for 2 cm and capture was achieved at 1 mAmp. The pacing rate was kept at 70/min (Figure 2). Temporary pacing was then followed by insertion of a bipolar permanent pacemaker lead positioned at Right ventricular apex through left subclavian vein puncture, with lead parameters kept at a heart rate of 70/min (Figure 3). The mode was kept as VVIR, with a threshold of 0.9 V, impedance 620 ohms and a pulse generator VVIR 297

PROMR1 was connected to the lead and implanted in the left pectoral pocket, with TPI lead removal. The patient was discharged with permanent pacemaker in situ, 5 days after insertion with no complications thereafter. Currently, patient is on follow up since past 2 years.

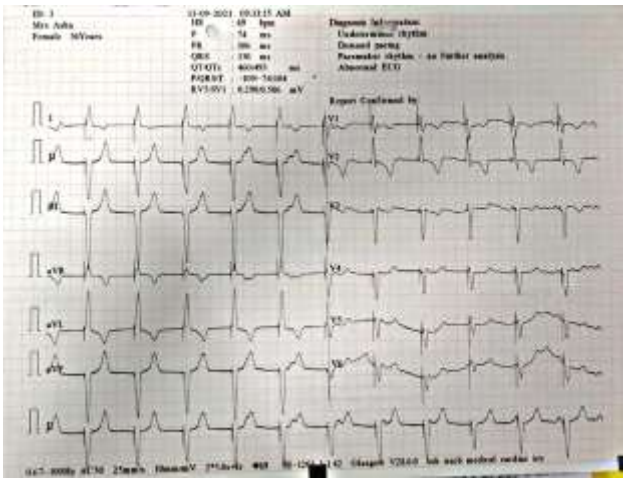

Figure 2: ECG post TPI-pacemaker rhythm with heart rate 70/min.

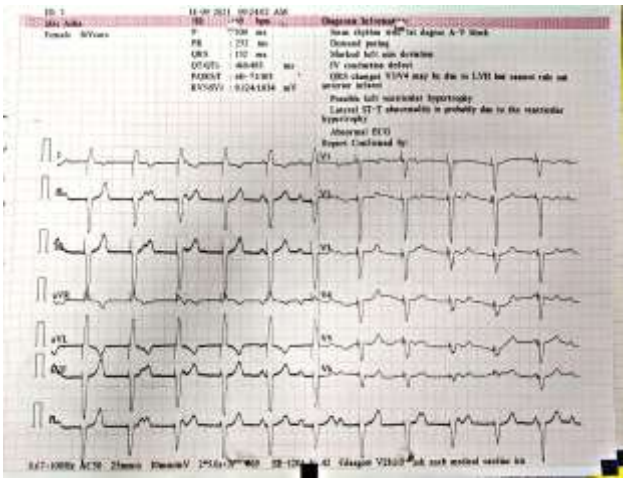

Figure 3: ECG post PPI-demand pacing with heart rate of 70/min.

DISCUSSION

The underlying cause of AV blocks is varied and the same for all degrees of blocks. These causes include idiopathic fibrosis and underlying chronic cardiac diseases such as structural heart disease, acute ischemic heart disease, medication toxicity, nodal ablation, electrolyte abnormalities, and post-operative heart block such as after surgical or transcatheter aortic valve replacement.[2] Additional causes of AV block include Lyme disease and some systemic diseases such as collagen vascular disorders, amyloidosis, sarcoidosis, and systemic lupus erythematosus.[1] Drugs associated with third-degree heart block include anti-arrhythmics from all four classes, and digoxin.[1]

Although AV blocks are fairly common, third-degree AV block is relatively rare.[7] The incidence in the general population appears to be low, approximately 0.02% to 0.04%.[8] Congenital heart block is a rare disorder. It has an incidence of about 1 in 22,000 live births. It may be associated with high mortality and morbidity.[9] The incidence of complete (third degree) congenital heart block is one in approximately 20,000 to 25,000 live births.[3] White and Eustis were the first to document congenital complete heart block (CCHB) with electrocardiogram in 1921. Until the 1950s, CCHB without structural heart disease was considered rare and benign.[10]

Congenital heart block is frequently associated with underlying structural congenital heart disease. The commonest forms of congenital heart disease associated with heart block include left atrial isomerism, often with an accompanying atrioventricular septal defect, as well as levo transposition of the great arteries.[4] The congenital heart block associated with neonatal lupus is considered a form of passively acquired autoimmune disease in which maternal autoantibodies to the intracellular ribonucleo-proteins Ro (SS-A) and La (SS-B), cross the placenta and injure the previously normal fetal heart. Women with serum titers of anti-Ro antibody carry a 3% risk of having a child with neonatal lupus syndrome. Recurrence rates are about 18%.[4]

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease of unknown cause that can affect virtually any organ of the body. Immunologic abnormalities, especially the production of a number of antinuclear antibodies (ANA), are a prominent feature of the disease.[11] Constitutional symptoms such as fatigue, fever, and weight loss are present in most patients with SLE at some point during the course of the disease. Arthritis and arthralgias occur in over 90 percent of patients with SLE and are often one of the earliest manifestations. Neonatal lupus being a rare entity, SLE presenting as congenital heart block is an even rare finding in the young adult population.

While CCHB in the absence of immunological exposure is recognised, patients with antibody-mediated CCHB have been found to require pacing earlier in life and follow a more malignant disease course than antibody-negative patients.[12] These antibody-positive infants experienced a higher risk of developing dilated cardiomyopathy (DCM) with clinical CHF with signs and symptoms. Several studies have attempted to elucidate the risk factors for the requirement of pacemaker implantation in patients with congenital heart block.[9] It is fairly well accepted that a mean resting heart rate below a determined number for the age group could be an indication to place a pacemaker. This is frequently quoted as a 55 bpm in the newborn period and gradually decreases with advancing age.[4]

However rare the presentation of congenital complete heart block is, the prognosis of congenital complete atrioventricular block (CCHB) is usually considered

favorable in adults.[5] This case report thus reviews such favourable outcome in an adult female of SLE presenting to the emergency for the first time with atypical cardiac manifestations and diagnosed as a case of congenital heart block with laboratory investigations revealing lupus as the primary etiology.

CONCLUSION

Complete heart block has a varied range of etiologies, including neonatal lupus presenting as congenital complete heart block. The primary presentation at a later age makes the diagnosis difficult, especially with an atypical presentation, making clinical suspicion all the more unlikely. However, with its favourable prognosis, it becomes essential for both the physician and cardiologist to direct towards the diagnosis with thorough history taking and early investigations.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Knabben V, Chhabra L, Slane M. Third-Degree Atrioventricular Block. In: StatPearls. StatPearls Publishing, Treasure Island (FL). 2022.
2. Kashou AH, Goyal A, Nguyen T, Chhabra L. Atrioventricular Block. In: StatPearls. StatPearls Publishing, Treasure Island (FL). 2022.
3. Friedman DM, Duncanson LJ, Glickstein J, Buyon JP. A review of congenital heart block. Images in paediatric cardiology. 2003;5(3):36.
4. National Organization for Rare Diseases. Congenital Heart Block, 2020. Available at https://rarediseases.org/rare-diseases/heart-block-congenital/. Accessed on 1 September 2023.
5. Michaëlsson M, Jonzon A, Riesenfeld T. Isolated congenital complete atrioventricular block in adult life: a prospective study. Circulation. 1995;92(3):442-9.
6. Yater WM. Congenital heart-block: review of the literature; report of a case with incomplete heterotaxy; the electrocardiogram in dextrocardia. Am J Dis Children. 1929;38(1):112-36.
7. Benjamin EJ, Blaha MJ, Chiuve SE, Cushman M, Das SR, Deo R, et al. Heart disease and stroke statistics—2017 update: a report from the American Heart Association. Circulation. 2017;135(10):e146-603.
8. Ostrander Jr LD, Brandt RL, Kjelsberg MO, Epstein FH. Electrocardiographic findings among the adult population of a total natural community, Tecumseh, Michigan. Circulation. 1965;31(6):888-98.
9. Kertesz NJ, Fenrich AL, Friedman RA. Congenital complete atrioventricular block. Texas Heart Institute J. 1997;24(4):301.
10. Wilkins E, Wilson L, Wickramasinghe K, Bhatnagar P, Rayner M, Townsend N. European Cardiovascular Disease: European Heart Network. 2007;3(2):111-2.
11. UpToDate. Clinical manifestations and diagnosis of systemic lupus erythematosus in adults, 2023. Available at: https://www.uptodate.com/contents/clinical-manifestations-and-diagnosis-of-systemic-lupus-erythematosus-in-adults. Accessed on 01 September 2023.
12. Villain E, Coastedoat-Chalumeau N, Marijon E, Boudjemline Y, Piette JC, Bonnet D. Presentation and prognosis of complete atrioventricular block in childhood, according to maternal antibody status. J Am Coll Cardiol. 2006;48(8):1682-7.

Cite this article as: Mishra V, Chatterjee T, Chandramouli M. Complete heart block – an atypical presentation with an atypical diagnosis. Int J Res Med Sci 2023;11:3855-8.