Complete Heart Block in a Case of Rheumatoid Arthritis: Uncommon Presentation of a Common Disease

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Abstract
We report a case of complete heart block (CHB) in a patient with rheumatoid arthritis (RA) because of its atypical presentation, negative Anti CCP and one of the uncommon causes of CHB. It occurs mainly in patients with established erosive nodular RA. It is usually sudden and permanent. It has several mechanisms, but the most common cause is infiltration in or near the AV node or bundle of His. If CHB develop the best option of treatment is the insertion of a permanent pacemaker. The prognosis is usually good provided no other cardiac lesions exist.

Key Words: Complete Heart Block, Rheumatoid Arthritis, Permanent Pacemaker

Introduction
Rheumatoid Arthritis (RA) is in itself, not fatal, but the associated complications may shorten the survival. The spectrum of cardiac involvement may vary from asymptomatic to sudden cardiac death.

The cardiac involvement in RA was first reported by Charcot in 1881¹ when he described pericarditis both as a clinical and as a necropsy finding in patients with ‘rheumatism articulaire chronique’.¹ No further observations were made for sixty years, until when Baggenstoss and Rosenberg in 1941² noted valvular lesions in association with RA. They attributed these two previous, unrecognized RA. However, in 1944 they identified granulomas in association with uncomplicated pericarditis and thought this a specific result of RA.³ Confirmation from other authors followed,⁴ and the first report of CHB due to a rheumatoid granuloma involving the conduction pathways was shared in 1959.⁴ Until 1983, about 20 cases were reported and Ahern et al. reported a series of 8 cases.

Implanting a permanent pacemaker (PPM) offers a reasonably good prognosis in the absence of other cardiac lesions.

Case Report:
Our Patient a 50-Year-old female of nondiabetic, normotensive, presented to us with vertigo two episodes of syncope for 2 weeks prior to our hospital admission. ECG in the cardiac emergency showed, CHB with junctional escape (Fig. 1). A detailed history was taken in which no obvious cause was found except the existing disease of RA. Through clinical examination revealed pulse-32/min with large volume, BP-110/70 mm of Hg. Raise jugular venous pressure of 11 mm of Hg. SPO2 was 96% on room air. Cardiovascular examination showed nothing abnormality except variable first heart sound. Respiratory examination revealed only bilateral basal crepitation otherwise normal. Musculoskeletal system examination revealed grade II tenderness of the proximal interphalangeal & metacarpophalangeal joints of the both hands. There were no deformities in the hand & feet.

Emergency temporary pacemaker was done. A detailed cardiac evaluation was done to find out the etiology of CHB. Troponin I, CK-MB was normal. Echocardiography (Fig. 3) revealed, no regional wall motion abnormality, good LV systolic function, mild mitral regurgitation, moderate tricuspid regurgitation with moderate pulmonary hypertension (pulmonary arterial systolic pressure of 55 mm of Hg). Diffuse pulmonary lung disease was confirmed by HRCT scan of chest. Coronary angiogram was done, which was absolutely normal.

On past history, she was diagnosed as a case of RA with extra articular involvement (scleritis) in Rheumatology Department, BSMMU, 10 years back on the basis of inflammatory joint pain involving multiple small & large joints of hand & feet, painful red eye, positive RA test (28.4IU/ml normal: less than 15 IU/ml). CRP was 128 mg/L, ANA, Anti dsDNA, c-ANCA, p-ANCA, Anti-CCP,
VDRL, TPHA, HLA B27 were negative. C3, C4 were normal. That time, she was treated with NSAID, weekly methotrexate 15 mg and oral methylprednisolone.

PPM was planned & subsequently dual chamber magnetic resonance compatible permanent pacemaker was implanted without any complications and mode was in DDDR (Fig.2). Since the pacemaker implantation, she had no episode of vertigo and dizziness.

**Fig.-1:** Complete Heart Block

**Fig.-2:** ECG-After PPM

**Fig.-3:** Echocardiogram-Good LV systolic function.

**Fig.-4:** Implanted Pacemaker

**Discussion:**
Rheumatoid arthritis (RA) is a chronic inflammatory disease of unknown etiology. It is a systemic disease, with extraarticular manifestations, including subcutaneous nodules, ocular, lung and cardiac involvement, peripheral neuropathy, vasculitis, and hematologic abnormalities. The spectrum of cardiac disease in rheumatoid arthritis includes pericardial effusion, cardiomyopathy, valvular involvement, conduction defects and coronary artery disease. Atrio-ventricular and intra ventricular conduction disturbances are described in patients with RA. These could be RBBB, hemiblocks or AV blocks of any degree. Complete AV block is rare, with an approximate incidence of 1 in 1000 patients with rheumatoid arthritis.\(^3,4\) It can develop suddenly, being discovered after syncope or found unexpectedly on routine physical and electrocardiographic examination.\(^4\) Lesser degrees of block may precede CHB for varying periods, sometimes with intervening periods of normal conduction.\(^4,5\) In a case series, M Ahern et al reported 4 out of 8 patients with CHB progressing from a lesser degree of AV block or bundle branch block in 24 hours to 7 years. It is possible that such changes are not uncommon but remain undiagnosed.\(^4\) The conduction disturbances are usually mild, asymptomatic, and incidentally diagnosed by electrocardiography. There is a female preponderance.\(^4\) The mean duration of RA prior to the development of the block is 10 to 12 years,\(^4,5\) although shorter duration is known. In the case series published by M Ahern et al, the average age for development of CHB was about 60 years.\(^4\) CHB in RA occurs generally in patients with established erosive nodular rheumatoid
Patients usually have other extra articular manifestations like our patient had eye and lung involvement. Most patients have a high titre of rheumatoid factor. CHB usually suggests active disease though it can occur in patients with well-controlled disease. The likely cause of CHB in patients with RA are (1) Direct involvement of the conductive system by granulomas and subsequent fibrosis, (2) Extension of the inflammatory process from the base of the aorta or mitral valves to the conduction pathways, (3) Secondary amyloidosis, (4) Hemorrhage into a rheumatoid nodule, (5) Coronary arteritis causing ischemia of the conduction tissue, (6) Focal myocarditis due to RA and (7) Premature coronary artery disease due to accelerated atherosclerosis in patients with RA. We consider the involvement of conduction system with granulomas and fibrosis as the likely etiology of CHB in our patient, although an endomyocardial biopsy would be required for confirmation, which was not done in our patient due to financial restrictions.

**Conclusion:**
Commonest causes of CHB in the cardiac department are usually ischemic and degenerative. But other secondary causes should be kept in mind, like in our case. Treating such case of CHB with RA needs both PPM and treatment of the primary disease. Spontaneous recovery is possible, but extremely rare. Conduction blocks once established in the disease do not respond to anti-inflammatory treatment. The indications for a permanent pacemaker are the same as any patient of CHB without RA. Once a pacemaker has been installed, the prognosis is usually good in the absence of other cardiac complications such as congestive cardiac failure.

**References**