

problems.⁵ The excessive unilateral growth of the mandibular condyle can lead to facial asymmetry, occlusal disturbance, and joint dysfunction.⁶ In 1986, Obwegeser and Makek classified the asymmetry associated with CH into 3 categories: hemimandibular hyperplasia, causing asymmetry in the vertical plane; hemimandibular elongation, resulting in asymmetry in the transverse plane; and a combination of the 2 entities called as hybrid variant.

The aetiology and pathogenesis of condylar hyperplasia is unknown. A plethora of presumed causes has been proposed in the course of time. Some of them include previous trauma, true neoplasia, hormonal disturbances, partial hemihypertrophy, arthrosis, osteochondromatosis, local circulatory disturbances, and neurotrophic disturbances.⁷ Other condition which can cause challenges in diagnosing this condition includes hemifacial hyperplasia and synovial chondromatosis. In the former condition, the associated soft tissues and teeth also will be enlarged and in the latter, there will be preauricular swelling with pain and limitation of joint function.⁸ Condylar hyperplasia usually occurs after puberty and is completed by 18 to 25 years.⁹ Prominent clinical features of condylar hyperplasia include an enlarged mandibular condyle, elongated condylar neck, outward bowing and downward growth of the body, and ramus of the mandible of the mandible on the affected side, causing fullness of the face on that side and flattening of the face on the contralateral side.¹⁰ The prominence of the chin is shifted to the unaffected side. An open bite might exist on the abnormal side. This depends, on one hand, on the rate of increasing enlargement of condyle and, on the other hand, on the downward growth of the maxillary alveolus and teeth. The unilateral asymmetric increase in length of the face, gives rise to a sloping rima oris. The mouth can be opened without restriction.¹¹ At an early stage occlusal contact is maintained by increased vertical height of the dentoalveolar structures in both the upper and lower jaws. This results in slanting of the occlusal plane towards the affected side. When the downward growth of mandible continues further it exceeds the dento-alveolar growth potential and produces an open bite in the premolar and molar regions.¹²

Radiographically, the condyle may appear relatively normal but symmetrically enlarged, or it may be altered in shape (e.g., conical, spherical, elongated, lobulated) or irregular in outline. It may appear more radiopaque because of additional bone present. A morphologic variation like elongation of the condylar head and neck may be seen. The ramus and mandibular body on the affected side also may be enlarged.¹³ Condylectomy on the affected side is the accepted method of treatment. It gives the best possible result with little post-operative discomfort to the patient.¹²

Unilateral condylar hyperplasia is one of the rare condition and challenging for both orthodontists and oral and maxillofacial surgeons. Its various types and variants can be assessed by imaging including photographs, postero-anterior cephalograms and 3D computed tomography (CT) scans. Early diagnosis by 3D computed tomography (CT) scans is mandatory in order to obtain optimal functional and aesthetics results.

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CASE REPORT

COMPLETE HEART BLOCK LACKING FUNCTIONAL ANATOMICAL IMPAIRMENT IN THE CONDUCTION SYSTEM

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ABSTRACT

Complete heart block also known as third-degree atrioventricular block (AV block) is a condition in which there is no conduction of the impulse produce in the sinoatrial node (SA node) in the atrium to the ventricle.¹ Complete heart block may be congenital or acquired. There are certain conditions which can lead to third-degree heart block, commonest being the coronary ischemia. Initially there may be first degree atrioventricular block (AV block), second degree atrioventricular block (AV block), bundle branch block or bifasicular block ultimately leading to complete heart block. In most cases third-degree AV block presents with acute myocardial infarction.^{2,3} A 45 years old male with no known comorbidities and without any risk factors for coronary heart disease coming with a short duration of complaints of dizziness and vertigo was diagnosed as a case of complete heart block without any functional and anatomical impairment in conduction system.

KEY WORDS: Complete Heart Block, Atrioventricular Block (AV block), Sinoatrial Node (SA Node), Coronary Ischemia, Bifasicular Block, Myocardial Infarction, Conduction System.

INTRODUCTION

Complete heart block is a condition in which there is inability in propagation of impulse from Sinoatrial node (SA node) to atrioventricular node. It has been identified that many patients with third-degree heart block endure from a bilateral bundle branch block. Pathophysiology and clinical aspects of heart block have shown that myocardial infarction is the common cause of complete heart block.⁴

Myocardial ischemia can produce broad range of conduction disturbances involving both the atrioventricular node and infranodal structures. Complete heart block is more common with inferior/posterior infarctions. Complete heart block occurring in association with anterior myocardial infarction implies extensive myocardial damage and has a worse prognosis.

Complete heart block complicating either inferior or anterior wall myocardial infarction is independently associated with mortality and in hospital complications. We report an uncommon pattern of complete heart block in which myocardial ischemia is absent and any other causes are not present.⁵⁻⁷

CASE

A 45 years old male presented to emergency department

with complaints of dizziness and vertigo for two hours. There were no risk factors for coronary artery disease. He was afibrile, alert, oriented with time place and person. His pulse rate was 35beats/min, blood pressure was 130/80mmHg, respiratory rate was 16breaths/min and oxygen saturation was 97% on room air. There was no raised jugular venous pulsation. He had normal respiratory, cardiac, abdominal and neurologic examination. Serum electrolytes, creatinine, complete blood count, blood culture and sensitivity and chest x-ray were normal. TSH is 9.709uIU/ml (0.36-4.94uIU/ml), FREE T3 and FREE T4 were in normal limits, tropinin I was 0.04 rests of cardiac enzymes were within normal ranges. His electrocardiogram at the time of presentation showed complete heart block and right bundle branch block (Figure 1).

He was admitted with the diagnosis of complete heart-block in critical care unit. A temporary pacemaker was placed (Figure 2).

All the possible causes for complete heart block were investigated. Coronary angiography was performed which showed insignificant coronary artery disease with preserved systolic function. Patient stayed in the hospital for 3 days.

Anti atheromas and anti coagulants were given to the patient. Patients escape heart rate stayed 35beats/min whenever rate of temporary pacemaker was decreased to 30beats/min (Figure 3).

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Figure 1. Complete Heart Block

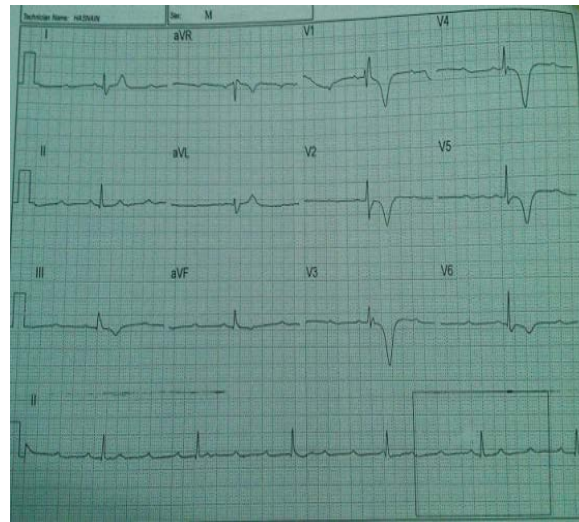


Figure 2. After Placement of Temporary Pacemaker

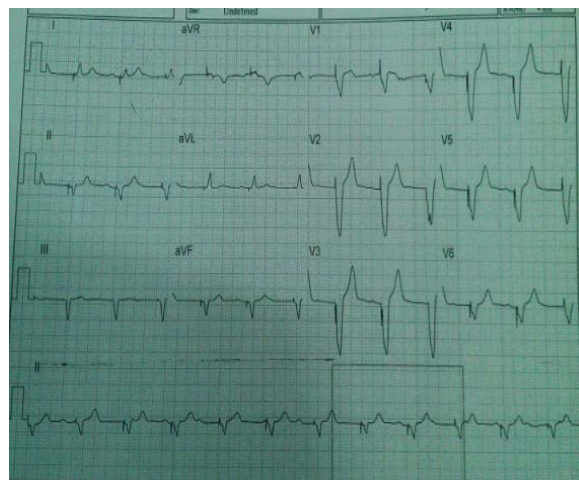
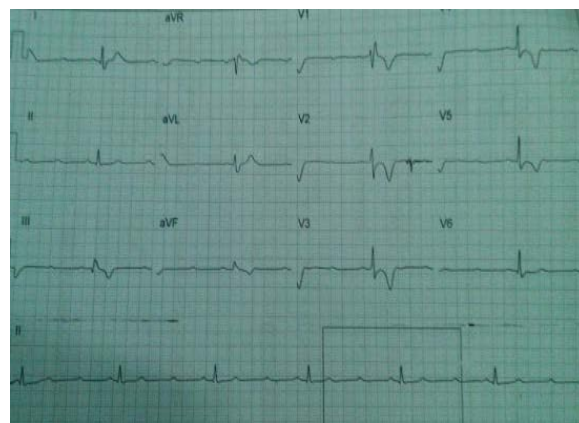


Figure 3. ECG showing HR of 30 when TPM rate is set to 30 /min.



DISCUSSION

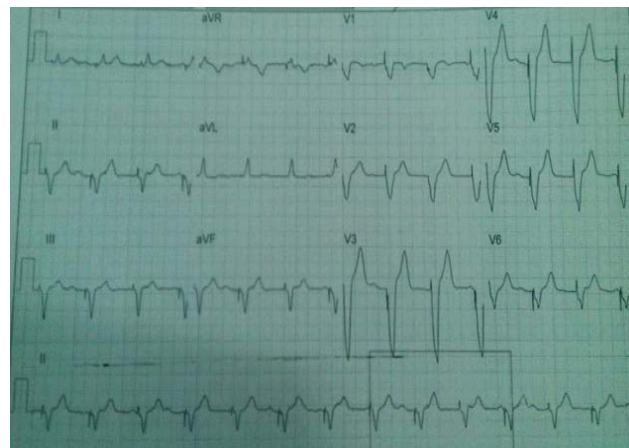
In Atrioventricular block or complete heart block there is impairment in the impulse propagation from the atria to the ventricle. This can be transient or permanent. Complete third-degree heart block can be congenital or acquired. Congenital form of complete or third-degree block commonly occurs at the level of atrioventricular node. Initially patients are asymptomatic relatively and at rest but later on develop symptoms with advancing age due to exertion because of inability of the body to adjust accordingly.

Most common cause is coronary ischemia an inferior wall myocardial infarction can cause harm to the atrioventricular block (AV block), causing third-degree heart block. An anterior wall myocardial infarction may damage the distal conduction system, causing third-degree heart block. This usually is extensive, permanent damage to the conduction system of heart; require a permanent pacemaker to be set.⁸ The escape rhythm typically generate in the ventricles, fabricate a wide complex escape rhythm. Congenital heart block is often related with maternal antibodies to SSA (Ro) and SSB (La) in the absence of any structural abnormality.

Atrioventricular block or third-degree heart block resulted from the various pathologic states causing fibrosis, infiltration or loss of connection in the process of the healthy conduction system. The common causes of acquired third-degree heart block are as follows:⁹

- Degenerative diseases Lev disease, Lenegre disease sclerodegenerative process and non compaction cardiomyopathy, nail-patella syndrome, mitochondrial myopathy
- Infectious causes cruzi infection, myocarditis, Chagas disease, rheumatic fever, varicella-zoster virus infection, Aspergillus myocarditis, valve ring abscess.
- Rheumatic diseases - Reiter syndrome, Ankylosing spondylitis, relapsing polychondritis, rheumatoid arthritis. Scleroderma
- Infiltrative processes sarcoidosis, multiple myeloma, Hodgkin disease, tumors.
- Neuromuscular disorders – myotonic muscular

Figure 4. ECG after implanting permanent pacemaker



dystrophy, becker muscular dystrophy

- Ischemic infarctive, causes - AVN block related with inferior wall myocardial (MI), anterior wall myocardial infarction associated with His-Purkinje block.
- Metabolic causes - hypothyroidism, hyperkalemia, hypoxia
- Toxins - "Mad" honey (grayanotoxin), cardiac glycosides (eg, oleandrin),
- Phase IV block (also known as bradycardia-related block)
- Iatrogenic causes

Single agent overdose can lead to a complete heart block or as is often the case from combined or iatrogenic co administration of atrioventricular nodal, calcium channel blocking agents and beta-adrenergic. Drugs which are utilized in heart block comprise of the following:

- Class Ia antiarrhythmics (e.g. procainamide, quinidine, disopyramide)
- Class Ic antiarrhythmics (e.g. encainide, flecainide, propafenone)
- Class II antiarrhythmics (beta-blockers)
- Class III antiarrhythmics (e.g. sotalol, ibutilide, dofetilide, amiodarone.)
- Class IV antiarrhythmics (channel blockers - calcium)
- Digoxin or other cardiac glycosides; those patients who are using digoxin should be educated about early symptoms of digoxin

The decision to perform the laboratory investigations depends on the associated history and findings. Complete blood counts indicate concordant problems like anemia. Spikes of fever or increased white blood cell count should be assessed by blood cultures because endocarditis can lead to heart block. Any metabolic disproportion like hyperkalemia which shows renal insufficiency or failure should be evaluated by serum electrolyte.

Coagulation profile should be performed and digoxin level should be taken for all patients who are on digoxin. In patients with suspicion of myocarditis than lab studies should be performed which includes Lyme titers, HIV serologies enteroviruses polymerase chain reaction and chagas titers as clinically appropriate. In our there is no history of any illness, no history of recent travelling, no history of I/V drug abuse. No history of hospitalization. He is otherwise a healthy male who has his own business works 8 hours a day. The definitive treatment of complete heart block is implantable pacemaker device unless it is due to the medication that can be put an end to or an infectious process that can be completely and easily treated.

According to American College of Cardiology, American Heart Association and Heart Rhythm Society suggested for permanent pacing in get hold of atrioventricular block (AV block) in adults includes: Permanent pacemaker implantation is designated for advanced second degree and third-degree atrioventricular block (AV block) at any anatomic level in conscious, symptom free patients in sinus rhythm with accepted duration of asystole 3 seconds or greater or any rate less than 40 beats/min or with an escape rhythm that is below the atrioventricular node.^{10,11}

Permanent pacemaker implantations specified for second degree and third-degree atrioventricular block

(AV block) at any anatomic level in conscious, warning sign free patients that have atrial fibrillation and bradycardia with one or more pauses of least 5 seconds or greater. In our patient after looking and evaluating for the possible causes he has the escape rate of 30beats/min with spells syncope in between after which permanent pacemaker was implanted.^{12,13}

CONCLUSION

Complete heart block can present in an adult patient, solitary in which there is no anatomical or functional impairment in the conducting system.

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CASE REPORT

SCHMIDT'S SYNDROME

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ABSTRACT

Schmidt's syndrome is a rare disease which is a one of the types of autoimmune polyglandular autoimmune syndrome. In these polyglandular autoimmune syndrome autoimmunity with auto-antibodies directed against different endocrine organs suggest in the pathogenesis of the disease. In addition there is role of genetic and familial predisposition of the disease. Autoimmune thyroid disease in combination with Addison disease is the most common presentation. In addition Diabetes mellitus Hyperparathyroidism, Pernicious Anemia, Hypogonadism, Vitiligo, Chronic atrophic gastritis, Chronic autoimmune hepatitis, Alopecia, Myasthenia gravis, Rheumatoid arthritis, Sjögren's syndrome and Thrombocytic purpura may or may not be present. So in patients suffering from one endocrine hormone deficiency should be thoroughly looked for the deficiency of other hormones. We present a case of 23 year old man who present with symptoms of gastritis and was refractory to treatment and on further evaluation was diagnosed as a case of Schmidt syndrome.

KEY WORDS: Schmidt's Syndrome, Autoimmune Hypothyroid, Polyglandular.

INTRODUCTION

Schmidt's syndrome is one of the types of polyglandular autoimmune syndrome. These are uncommon diseases characterized by abnormal hormone production by more than one endocrine gland. Autoimmunity directed against more than one endocrine gland is considered the main pathological abnormality. Other organs apart from endocrine glands might be affected. These syndromes are further classified in to different types on the basis of the involvement of different endocrine glands:

- Autoimmune Polyendocrine Syndrome Type 1 (Autoimmune Polyendocrinopathy-Candidiasis-Ectodermal Dystrophy or Whitaker's syndrome)
- Autoimmune Polyendocrine Syndrome Type 2 (Schmidt's syndrome)⁴
- X-linked Polyendocrinopathy, Immunodeficiency and Diarrhea-syndrome, also called XLAAD (X-linked Autoimmunity and Allergic Dysregulation)

SCHMIDT'S SYNDROME is rare and usually presents in early adulthood with females affected three times more than men.³ It has been linked to the class II HLA haplotypes DR3 (DQB*0201) and DR4 (DQB1*0302).⁷ Autoimmune thyroid disease in combination with Addison disease is the most common presentation.¹

Diabetes mellitus in 20% of cases and rarely in addition to above symptoms hypogonadism, hyperparathyroidism, pernicious anemia and vitiligo is also present. Autoimmune thyroid disease can manifest in the form of hashimoto thyroiditis as well as Graves' disease but hashimoto thyroiditis is much more common as occurring in more than 90% cases of Schmidt's syndrome.

In addition to genetic predisposition there is also a role of autoimmunity behind the pathogenesis of the disease. The autoantibodies are considered to be the basis behind the destruction of these multiple endocrine organs.² Very few

cases of Schmidt's syndrome have been reported and here we present a case of 23 year old man who present with symptoms of gastritis and was refractory to treatment and on further evaluation was diagnosed as a case of Schmidt syndrome

CASE

This case was seen between 8th of march 2013 to 22nd of march 2013. A 23 year old male presented to the medical outdoor of military hospital Rawalpindi in the second week of march 2013 with a history of repeated episodes of tiredness, lethargy, lassitude, drowsiness of one year duration. He also complained of vague abdominal pain, indigestion, and reflux symptoms for the same duration for which he was taking Tablet Omeprazole.

His symptoms had aggravated over the past few weeks and he was unable to perform his daily routine work. He also complained of constipation, decrease apatite, forgetfulness, cold intolerance, joint and muscle pains, headache, hair loss, dryness and itching of skin. He has been married for the last 2 years but is not able to conceive. Family history was positive for diabetes mellitus.

On examination a pale young man with dull expressionless face and periorbital puffiness was seen (Figure 2). Vitals were normal but ankle reflex was delayed. Dry rough skin was seen all over the body especially on arms and legs. A midline neck swelling which moves with swallowing was seen. Swelling was 5*6cm in dimension with smooth margins and surface. Non pitting bilateral pedal edema was seen. He was admitted in the hospital for evaluation and on laboratory investigations it was found that his T3 (0.6nmol/l) and T4 (3.5pmol/l) levels were low and TSH (75 IU/L) levels are high. Antithyroid peroxidase antibodies were positive. Serum cholesterol (6.5mmol/l) was increased as well as serum ALT (266U/l) levels.

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