Total AV Block in Amyotrophic Lateral Sclerosis

Yudhie Tanta[1](✉), Ali Ghanie[1], Andika Okparasta[1], Taufik Indrajaya[1], and Erwin Sukandi[2]

[1] Internal Medicine Department, Sriwijaya University, Palembang, Indonesia
tanta7an7a@yahoo.com
[2] Neurology Department, Sriwijaya University, Palembang, Indonesia

Abstract. Bradyarrhythmia can be a life-threatening condition that mandates quick and proper responses and acts from a clinician. Furthermore, bradyarrhythmia management should not stop only in increasing the patient heart rate, but also to find the etiologies that caused this condition. Etiologies of bradyarrhythmia range from pathological conditions that solely affect the heart to systemic and multi-organ processes. Permanent pacemaker implantation is generally needed when the condition underlying the bradyarrhythmia is irreversible. On the other hand, correction of reversible causes might avoid patients from unnecessary permanent pacemaker implantation. This study aims to present a unique case of bradyarrhythmia with rare etiology. Case summary: a young man with total AV block accompanied by hemodynamic disturbance. Other symptoms include general weakness and significant loss of body weight. Magnetic Resonance Imaging can't be performed in this patient since early transvenous temporary pacemaker implantation was mandated. ENMG examination was performed as an alternative. Results: The ENMG results showed a decrease in motoric wave amplitude, pathological wave was observed in the deltoid muscle, dorsal interosseous, and in the right part of the tongue, while sensory wave showed a normal amplitude. These findings are consistent with the ENMG pattern of definite Amyotrophic Lateral Sclerosis. Since the underlying disease of the total AV block is considered irreversible, permanent pacemaker implantation was performed in this patient. The bradyarrhythmia management must not stop only in the measure of increasing heart rate, but also should include the efforts of searching the underlying etiology thoroughly.

Keywords: Amyotrophic Lateral Sclerosis · AV block · bradyarrhythmia

1 Introduction

Bradyarrhythmia can pose a life -threatening condition that should be responded fast and accurately by clinicians. After acute stabilization, the next step of bradyarrhythmia management is to find the etiologies that caused this condition. Etiologies of bradyarrhythmia range from pathological conditions that solely affect the heart to systemic and multi-organ processes. Permanent pacemaker implantation is needed when the bradyarrhythmia is irreversible despite correction measurements of the underlying condition. On the other hand, correction of reversible causes might avoid patients from unnecessary permanent pacemaker implantation.

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2 Case Report

A 30-year-old man, Mr. OS, was referred from another hospital with complaints of 1 day of vision loss. Complaints of dizzy vision worsened when the patient changed position from lying to standing. Since 5 years ago the patient complained of difficulty in lifting the eyelids and difficulty swallowing. The patient's weight loss was +10 kg compared to 1 year ago. For 2 weeks before being admitted the patient also complained of weakness in the patient's arms and legs.

Vital sign examinations showed that blood pressure was 80/50 mmHg, pulse rate: 30 bpm-regular, respiratory rate was 28 bpm, consciousness was somnolence. Physical examination revealed generalized muscle atrophy, bilateral palpebral ptosis, bilateral ophthalmoplegia, and atrophy of the papillae of the tongue. Also obtained wrist and thumb sign, cruris length 45 cm, and the ratio of the upper segment: lower body segment 0.77 (<0.9).

The ECG showed total AV block rhythm. On laboratory examination, it was found that Hb 13.3 g/dl, Leukocytes 9930/mm^3, Platelets 236000/mm^3, Hematocrit 36%, Albumin 4.1 g/dl, Sodium 138 mEq/l, Potassium 4.7 mEq/l, Rapid AntiHIV non-reactive, Troponin T < 40. Examination of the patient's chest x-ray showed a normal impression.

The patient was diagnosed with unstable total AV block and suspected intracranial mass with myasthenia gravis as a differential diagnosis. Patients were treated with atropine sulfate 0.5 mg iv evaluated and repeated every 5 min. The pulse rate after administration of up to 3 mg (6 times iv bolus) was 30x/minute with a total AV block rhythm. The patient was planned for the implantation of the Temporary Transvenous Pacemaker (TPM) immediately. The patient was also managed with a 4 mg/h dopamine drip while waiting for TPM preparation.

Implantation of TPM is done through the access of the right femoral vein. After the lead pacemaker reaches the apex of the right ventricle, the pacemaker is activated with an output of 3 mV, sensing of 2 mV, and a heart rate of 60 beats/min.

After insertion of TPM, the patient had hemodynamic improvement. The patient's blood pressure is 100/60, heart rate is 60x/min, respiratory rate is 22x/min, consciousness is compos mentis.

The patient underwent an echocardiographic examination. The result was good wall motion, with 57.1% ejection fraction, normal heart chamber dimensions, normal valves, with normal echocardiography impression.

The patient was consulted to the Neurology department. The Ice Pack test was negative, physiological reflexes decreased and pathological reflexes increased. Apart from Myasthenia Gravis which is a Lower Motor Neuron disease, the patient was suspected of having an Upper Motor Neuron lesion.

The patient was diagnosed as intracranial SOL, differentially diagnosed with Wallenberg syndrome, amyotrophic lateral sclerosis (ALS), Spinal Muscular Atrophy, and Myasthenia Gravis. Patients were advised to do a cranial MRI or a cranial CT scan with contrast and an ENMG examination.

From the CT scan of the cranial with contrast, no abnormalities were found, while the MRI and ENMG examinations could not be performed because the patient had TPM installed. In addition, the patient also underwent a Digital Subtraction Angiography

(DSA) examination to evaluate the possibility of posterior cerebral artery infarction, but the results were normal.

The patient then underwent the installation of a Single Chamber Permanent Pacemaker. Installation of PPM is done by using the Seldinger technique through the left subclavian vein. PPM is activated by VVIR (Ventricular sense, Ventricular inhibit, Rate Responsive) mode. The heart rate is set at a rate of 60 bpm. The minimum pacing threshold obtained is 0.6 V.

The patient was then tested for acetylcholine receptor antibodies as an alternative to the ENMG examination. A negative result was obtained.

Finally, it was decided to do an ENMG examination with close rhythm monitoring. There was a decrease in the amplitude of the motor waves, and pathological waves of the deltoid, dorsal interosseous, and right tongue were also found. While the amplitude of the sensory wave is normal. These results are in agreement with definite amyotrophic lateral sclerosis.

3 Result and Discussion

Presyncope symptoms are a common manifestation in cases of bradyarrhythmia, so they are often the entry point in detecting bradyarrhythmia abnormalities. In this patient, the symptoms of blurred vision were found since 1 day of SMRS. The ECG showed a total AV block rhythm with a heart rate of 30 beats per minute.

The most commonly used approach to managing bradyarrhythmias is the Advanced CardiovascularLifeSupport (ACLS) guidelines. In ACLS, the heart rate is categorized as bradyarrhythmia if the value is <50 bpm.

The next step is to determine whether this bradyarrhythmia condition is classified as stable or unstable. Stable bradyarrhythmias do not require special treatment, but the patient's condition must be monitored closely so that if he falls in an unstable condition, prompt action can be taken. On the other hand, unstable bradyarrhythmias require special medical treatment to the implantation of an emergency Temporary Transvenous Pacemaker (TPM) [1].

In this patient, there was a decrease in consciousness, where the patient's level of consciousness was somnolence. The patient's blood pressure is 80/50 mmHg. So it can be concluded that the patient is in an unstable condition. The patient received initial medical treatment, because the response was inadequate, it was decided to install TPM on the patient.

After the patient's condition has been stabilized, the next step is to find the etiology of the patient's bradyarrhythmia. Findings and good treatment of reversible causes can prevent patients from unneeded Permanent Pacemaker (PPM) implantation. The etiology of reversible bradyarrhythmias include; ischemic conditions or infarction, hyperkalemia, hypothyroidism, drug toxicity, and Lyme infection [2].

In this patient, there were no complaints of chest pain, biomarkers of heart damage Troponin T < 40, plasma potassium values within normal limits, previous routine drug use, and narcotic use were denied. There were no specific findings suggestive of a reversible cause in this patient.

Besides the problem of bradyarrhythmias, the patient also found general weakness, and generalized muscular atrophy, bilateral palpebral ptosis, and difficulty swallowing. There is a need to explore the causes of bradyarrhythmias associated with neuromuscular disease.

Some clinical findings obtained in patients tend to point to Myasthenia Gravis. These clinical findings include weakness that starts in the small muscles first and then progresses to weakness in the large muscles. Besides that, it was also found in the weakness of the legs that was felt, especially after walking for a while [4]. Meanwhile, tongue papillary atrophy and tongue fasciculations are more likely to lead to the diagnosis of Amyotrophic Lateral Sclerosis or Spinal Muscular Atrophy [5, 6]. Examination of acetylcholine receptor antibodies which showed negative results left Amyotrophic Lateral Sclerosis or Spinal Muscular Atrophy type 3 as the etiology of neuromuscular disorders in the patient [7, 8].

As no reversible etiology was found and monitoring of the original rhythm of the heart remained inadequate, it was decided to implant a permanent pacemaker in the patient. The selected device is the single chamber pacing with VVIR mode and passive lead fixation [9].

Then an ENMG examination was performed with preparation in case of pacemaker failure [10]. On ENMG examination, Upper Motor Neuron and Lower Motor Neuron abnormalities were found at two levels, and bulbar abnormalities were also found in this case, pathological waves were found on the right tongue. Meanwhile, there was no sensory disturbance. With these ENMG findings the diagnosis of Amyotrophic Lateral Sclerosis can be established [11].

4 Conclusion

Amyotrophic Lateral Sclerosis is an idiopathic neurodegenerative disease that affects both Upper Motor Neurons and Lower Motor Neurons [8]. This disease generally affects men in the 5th and 6th decades of life, but can also occur at a younger age. This disease has a poor prognosis, where the average life expectancy of patients from first onset is 3 years [11].

Cardiovascular manifestations of amyotrophic lateral sclerosis are caused by dysfunction of the autonomic nervous system [8]. Early autonomic dysfunction is characterized by sympathetic nerve predominance, whereas later stages are characterized by sympathetic nerve failure and vagus nerve predominance. These cardiovascular manifestations appear in baroreceptor disorders, and impaired heart rate regulation [8].

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