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CASE REPORT: CHOREA SYDENHAM IN YOUNG ADULT FEMALE AS SYMPTOM OF RHEUMATIC FEVER

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ABSTRACT

Background: Sydenham chorea is an involuntary movement disorder due to an autoimmune process. It may be part of rheumatic fever symptoms. A thorough and complete examination is needed to create the right diagnose. It will support the success of therapy and prevent other complications.

Case: An 18-year-old woman went to the hospital with complaints of involuntary movement on the left side accompanied by fever, breathing problems, and nausea. There were no motoric weakness, tremor or another involuntary movement. Laboratory blood results for Anti Streptolysin Titer O (ASTO) and C-Reactive Protein (CRP) results increased. The cardiologist confirmed she had mild mitral valve regurgitation. Those exams results were part of the criteria for diagnosing rheumatic fever, and the patient can get the right therapy immediately.

Discussion: Sydenham Chorea can't threaten the patient's life directly, but incorrect examination and treatment will cause other severe complications. That is because Chorea Sydenham is part of the autoimmune process, which can cause the other organs complication.

Conclusion: Sydenham Chorea may relate to rheumatic fever. Early diagnostic is essential and it will support the provision of therapy and accelerate the recovery of patients.

Keywords: sydenham chorea, rheumatic fever, symptoms, diagnostic criteria

Background

Involuntary movement is a spontaneous movement that is uncontrollable, unconscious, aimless, unpredictable at any time and not controlled or stopped when the person moves and disappears during sleep time. There are several types of involuntary movement disorders. Chorea is one type of involuntary movement disorder caused by a disturbance in the part of the brain called the basal ganglia, precisely in the caudate nucleus of the corpus striatum. Chorea comes from the Greek "Chorea" which means dancing. As the name implies, the characteristics of the chorea movement resemble in a dancing. The movements are not synchronizing to each other; they are strong, fast, and jerked, while the direction of movement changes quickly. There are several types of chorea including Huntington chorea (major chorea) caused by hereditary factors, chorea due to metabolic disorders such as Wilson disease, idiopathic chorea such as physiological chorea in infants, and Sydenham chorea (chorea minor) caused by autoimmune disorders of the body.[1]

In Sydenham chorea, the involuntary movements symptoms started by bacterial infection of Haemolytic Streptococcus Beta A Group. The bacterial infection manifested around six months earlier before the involuntary movement symptom appears on the patient. Some data show that post-streptococcal infection might manifest into other extrapyramidal movement disorders such as dystonia, tic, tremor, but Sydenham chorea is specifically found most commonly on the patient with rheumatic fever. This symptom is part of rheumatic fever disease. Sydenham chorea attacks women more often than men with a ratio of 3:1 and more often appears on children around 5-15 years old.[2]

Unfortunately, there is no exact number about the Sydenham chorea incident in Indonesia yet. There is some proof that in developing countries, the incidence of rheumatic fever is higher due to poor sanitation compared to developed countries.[3] Those involuntary movements can appear on the face, arms, or lower limbs. Involuntary movements that arise during an attack can be repeated from several minutes to hours, either with a resting interval between movements or continuously. It is common for patients to experience pain or fatigue due to repeated movements uncontrollably. Aside from chorea, rheumatic fever can also manifest in ballistic movement due to damage of other parts in basal ganglia, meningeal irritation, encephalomyelitis, and optic neuritis.[4]

Other symptoms commonly complained include fever, shortness of breath due to abnormalities in the heart valves, weakness and pain in the limbs. Those are mainly caused by the inflammation of the organs due to the infection process caused by Streptococcus. To deal with abnormalities of Sydenham chorea, a comprehensive and complete examination is required. This step is needed to create a proper diagnosis and will support the success of therapy. The goal of therapy is not only to overcome the symptoms of involuntary motion disorders but also to eliminate the main causes, overcome additional symptoms, and prevent other severe complications. Therefore, sometimes treatment and teamwork of several specialists are needed so the diagnosis can be properly made. When those are accomplished, the therapy can be administered to the patients as soon as possible.[5]

Some laboratory blood tests such as erythrocyte sedimentation rate and C-reactive protein are needed to determine whether there is an inflammatory process and to know the existence of the previous infection. Another useful examination to determine the presence of Haemolytic Streptococcus Beta A Group bacterial infection previously was the Anti Streptolysin O (ASTO) blood examination. There is a precaution that 20% of Sydenham chorea patients have no serological evidence of previous Streptococcus infection.[3]

ECG or echocardiography is needed to check if there is a rheumatic fever disorder that affects the part of the heart, although not every patient might experience it. The principle of therapy in Sydenham chorea itself is by giving penicillin group antibiotics, or erythromycin and anti-inflammatory groups to eliminate the etiology of streptococcal bacterial infection. If complications are found in the heart valve, it is recommended to immediately request further treatment from the cardiologist. Whereas the involuntary movement abnormalities can be medicated by antipsychotic drugs which function as dopamine antagonists and have the

effect of being anti-spasmodic to control abnormal movements.

This case report provides information about the symptoms of Sydenham chorea, which is sometimes forgotten as part of the symptoms of rheumatic fever. Understanding the symptoms of Sydenham chorea and rheumatic fever can lead to an earlier establishment of the diagnosis. This information will be useful to provide appropriate management in patients with Sydenham chorea and can avoid further complications from rheumatic fever.

Case Illustration

An 18-year-old young woman was sent to the ER of Wangaya Regional Hospital with the main complaint of the left side of the body moved uncontrollably and can't be stopped for the last three days. In the beginning, these movements only appear a few minutes every few hours. As time goes, the duration is getting longer, and the frequency increases more often. Those symptoms were causing discomfort and disrupting the patient's activity. When the patient came to the hospital for treatment, she was still experiencing involuntary movements that occur in parts of the body covering the neck area, up to the left arm. The movements are slow, repetitive, swinging, similar to the movement of a dancer, which occur unnoticed and cannot be controlled. The duration of movement happening from 5-10 minutes to each attack and reappears several times. There is a break between several attacks when no involuntary movement is happening. The patient also experienced other symptoms such as fever, shortness of breath, nausea, and muscle pain (arthralgia) due to repetitive involuntary movement. In the past nine months, there was a history of repetitive rhinitis and pharyngitis. Patients claimed there had been no previous history of a similar disease and no history of seizures in either the patient or family. The patient also denied using other drugs as medication. During the examination, the patient had full awareness. The consciousness level was Compos Mentis with GCS score E4V5M6, and had adequate contact. The patient was moderately ill, with a body temperature of 38.50C, blood pressure 130/80 mmHg, respiratory rate 30x / minute, pulse frequency 110x / minute, 99% oxygen saturation. The neurological examination results consisted of isochore pupils with positive bilateral light reflex, no meningeal signs were found. There were no weaknesses and lateralization. The results of motor and muscle tone examination were within normal limits. No pathological reflex was found. No other involuntary movements such as hemiballismus, athetosis or tremor were found. Laboratory tests carried out in the form of routine blood with leukocyte results: 15.46 / ul, neutrophils: 71, 5%, lymphocytes: 15.1%, monocytes: 13.1%. Examination of the chest x-ray on anterior-posterior position showed the result of the heart, lung, pleura, and diaphragm were in normal condition. On Echocardiography examination, there was a sinus tachycardia rhythm. During the patient stay at the hospital, other blood tests were being examined. Those tests were Anti-Streptolysin Titer O (ASTO), and C-Reactive Protein (CRP) with ASTO results: 450 IU / mL and CRP: 4.5 mg / L. The neurologist gave medication of antipsychotic therapy which functions as an anti-spasm to reduce involuntary movements. The chosen drug was haloperidol tablet 2x5 mg, and diazepam tablet 2x5 mg if the involuntary movement symptoms not showing any improvement. The neurologist then consults the patient to get further examination and treatment from a cardiologist. Echocardiography was performed by a cardiologist, and mild mitral regurgitation was found. Treatments for this abnormality are acetosal tablet 3x500mg, and benzathine Penicillin 1.2 million IU IM per injection for two days. For other additional symptoms, the patient has to take paracetamol tablets 3x500 mg to treat the fever. The injection of Proton Pump Inhibitor (PPI) class drug was administered to prevent stomach irritation and reduce nausea. After the third day of treatment, the patient condition had improved, and she experienced less involuntary movements in the left part of her. The patient also experienced fewer symptoms of fever, nausea and shortness of breath. The dose of haloperidol is reduced to 3x2.5 mg (1/2 tablet) orally and maintained until home. The patient returned home on the 6th day and had planned to control her condition at the neurology clinic five days after leaving the hospital. The

patient was also got advice to take head CT-scan examination when she visited the neurology clinic if there were going to be any other neurological symptoms during her stay at home.

Discussion

Sydenham chorea is a neurological manifestation of rheumatic fever and occurs 20-40% in patient with rheumatic fever. It was caused by the infection of Haemolytic Streptococcus Group A bacteria. During the infection, the patient's body forms immunoglobulin G antibodies which reacting to cross the Streptococcus membrane antigens and cells in the neuronal cytoplasm of the caudate nucleus and subthalamic nucleus (intracellular tubulin and extracellular lysoganglioside). This process happens because the antigens from Streptococcus bacteria molecularly in the patient's body disguise themselves so that they are similar to the cells found in the cytoplasmic wall of the basal ganglia, especially the caudate nucleus (this process is also called mimicry). This cause the host antibody not only attack antigens from Streptococcus bacteria but also contributes to damage to basal ganglia cells because the host antibodies cannot distinguish between the original bacterial antigen and the basal ganglia cell wall which has been modified by Streptococcus bacterial antigens. As a result of damage to the basal ganglia cell wall, its function is to control and change the movements of rough movements to be smooth and regular also damaged. This movement abnormality is a neurological manifestation of rheumatic fever.[6]

It is only one part of an autoimmune disease caused by bacterial infection with Haemolytic Streptococcus Group A. Rheumatic fever is a collection of some abnormality that appears due to this autoimmune process. The Streptococcus antigen is also reported to have similarities to the structure on the muscle fibers of the heart wall organ. That causes the abnormalities in the heart is also a frequent manifestation.[7]

Diagnosing Sydenham chorea is not enough just by having a physical examination or a brief history. A thorough anamnesis is needed, including the history of the patient's illness to the symptoms of other diseases and the accompanying complications. To be able to diagnose rheumatic fever, the latest 2015 Jones criteria used by the AHA are used. It is said that the patient meets the criteria for rheumatic fever when there are two major criteria or one major criterion plus two minor criteria from the classification of rheumatic fever by Jones. There are three major criteria, and three minor criteria of Jones diagnosis criteria can be found on the patient which are appropriate for rheumatic fever diagnosis. The major criteria found in this patient are the presence of subclinical carditis where cardiac abnormalities are found on echocardiography in the form of mild mitral regurgitation. Physical examination revealed tachycardia with shortness of breath in the initial complaint. And for the second major criteria is the presence of Sydenham chorea itself. Whereas for the two minor criteria in this patient were the symptoms of fever with the results of ASTO blood laboratory tests: 400 IU / mL and CRP patients with 4.5 mg/dl where both values experienced an increase. The patient also experienced muscle pain (polyarthralgia) caused by the rapid involuntary movement. These symptoms have met the diagnostic criteria for rheumatic fever according to the latest revised Jones classification in 2015. (table 1). Another supporting factor is the female gender of the patient, where the female sex has a high incidence of Sydenham chorea in rheumatic fever. Even though the patient's age is over 15 years, it does not rule out the diagnosis possibility of rheumatic fever. The diagnose of rheumatic fever for this patient is supported by the symptoms and previous history of respiratory tract infection, which are matching the Jones criteria for rheumatic fever.[8] The symptoms of cardiac abnormalities in the patient are only mild, which is the presence of tachycardia and mild tightness. But to ensure there is no further complication in the heart organ, the patient must go through several examinations on the second day of treatment. Those are among others an echocardiography, electrocardiography (ECG), and chest X-ray. From the chest X-ray, no abnormality was found, and there was only a sinus rhythm tachycardia on the ECG results. On echocardiography examination, there was mild mitral regurgitation due to an autoimmune process caused by bacterial infection of Haemolytic Streptococcus Group A which caused inflammation

in the heart area.[9] Other supportive examinations such as CT scan or MRI were not performed because they usually did not produce significant results in Sydenham chorea patient with rheumatic fever and the patient was not showing any other neurological abnormalities.[10] The patient is still given the advice to take head CT-Scan examination during her visit to the neurology policlinic if she had any other neurological symptoms.

The medication she took was for eliminating the infection of the Haemolytic Streptococcus group A which is the etiology of the autoimmune process itself. The therapy was in the form of penicillin class antibiotics, namely benzathine penicillin at a dose of 1.2 million IU IM injection/day, administered for two days. Other antibiotic choices to treat Haemolytic Streptococcus Group A infections is azithromycin with the dose of 500 mg once daily for 5-7 days or clarithromycin 2x250 mg each day for 10 days. The treatment for the symptoms of involuntary movement was neuroleptic drugs. The chosen drug therapy is haloperidol. It's working against chorea by inhibiting the effect of dopamine and increasing its turnover. Dopamine is a neurotransmitter, a signaling molecule in the brain which is involved in motor control. The increased activity of dopamine is thought to cause chorea. The exact mechanism is not yet fully understood but it is thought to block so-called dopamine D2 receptors. This prevents dopamine from binding to receptors, which interfering with dopamine's action in the result, and it will control abnormal movements in the end. The dose given on the therapy is 2x5mg tablets. In addition, the patient was also given diazepam with a dose of 2x5 mg given which was only used on the first day of treatment because the patient already shows improvement after given the previous drug.

Diazepam is a benzodiazepine class of drugs that used to increase levels of GABA concentration in cauda, putamen, substantia nigra, and globus pallidus. By analogy with increased GABA activity can reduce chorea movement. Usually given as adjuvant therapy. The mild manifestations of the patient's heart were treated by using aspirin. The use of aspirin is intended as a non-steroid anti-inflammatory to deal with damage to the heart organs due to streptococcus infections. The recommended dose is 100 mg/kg which is divided into 4 doses per day, but the high dose is feared to give negative side effects to the patient's gastrointestinal tract so that the administration drug dose is adjusted to 3 x 500 mg tablet. Non-steroid anti-inflammation drugs are a better option for treating carditis rather than a steroid one. That's because non-steroid anti-inflammation drugs can be used continuously without having to taper off the dose when the therapy has to stop. Anti-inflammation drugs can be used for 2-4 weeks, depending on the clinical progress. The patient was also medicated by using Proton Pump Inhibitor (PPI) drugs to reduce nausea and reduce the risk of stomach irritation. Paracetamol 3x500mg tablet was given to reduce fever. Prophylaxis treatment can be given for young patients around 3-15 years old who have resistant symptoms or recurrent infection of Streptococcus.

The chosen prevention therapy is the continuous injection of benzathine G penicillin 1.2 million units every 3-4 weeks. The other oral antibiotics used as prophylaxis isn't as effective.

But this prevention might different for each patient because of the inconvenience of frequent injection.[1]

The early diagnosis is very important because it will affect the administration of therapy. Sydenham chorea does not directly threaten life immediately, but other symptoms of rheumatic fever can have a fatal effect on the patient. The complication which might appear due to mistreated and misdiagnosis are permanent damage to basal ganglia which can cause untreated involuntary movement and worsening the muscle spasm. Another severe complication is heart failure due to further damage to heart tissue which can cause death. Sydenham chorea in this patient appears because it is part of rheumatic fever symptoms so that the treatment is not separated only from medication to stop the symptoms of involuntary motion alone. With the right diagnosis and early treatment, the symptoms of the patient's illness gradually reduced and other severe complications can be avoided. The patient is taking advice to maintain hygiene for preventing recurrent upper respiratory tract infections which can cause repetitive infections and can manifest again into rheumatic fever.[12]

Table 1. Jones criteria for rheumatic fever (revised 2015) and patient's condition and symptoms

Symptoms	Low-risk population (ARF incidence ≤2 per 100.000 school-aged children or all-age RHD prevalence of ≤1 per 1000 population year	Moderate/high-risk population (Children not clearly from a low-risk population)	Patient's Case
	Major Criteria		
Carditis	Clinic or subclinic	Clinic or subclinic	Subclinic
Arthritis	Only polyarthritis	Monoarthritis, polyarthritis and or polyarthralgia	Polyarthralgia (muscle pain due to involuntary movement)
Chorea	Yes	Yes	Yes
Erythema Marginatum	Yes	Yes	No
Subcutaneus Nodules	Yes	Yes	No
	Minor Criteria		
Fever	≥ 38,5°C	≥ 38°C	≥ 38,5°C
Athralgia	Polyarthralgia	Monoarthralgia	Polyarthralgia
ESR	Peak ESR ≥ 60 mm in 1 hour	Peak ESR ≥ 30 mm in 1 hour	-
CRP	≥ 3,0 mg/dl	≥ 3,0 mg/dl	4.5mg/dl
PR interval (duration depend on age, unless carditis is found)	Prolonged	Prolonged	-

Conclusion

Sydenham Chorea is an involuntary movement disorder that can be part of rheumatic fever symptoms. It may attack young adult who has recurrent upper respiratory tract infections in the last six months, the presence of fever, and the appearance of abnormalities in the heart. When the patient can get treatment immediately, it will reduce the risk of other severe complications such as permanent damage to basal ganglia and cardiac heart muscle.

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