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Neurologically Disabling Features of Rheumatic Fever

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Abstract

Acute Rheumatic Fever (ARF) is rare multisystem, immune-mediated sequelae of an untreated group-A streptococcal infection. Due to its immune-mediated pathogenesis, ARF can present with a variety of different tissue and organ manifestations like arthritis, carditis, chorea, subcutaneous nodules, and erythema marginatum. The large number of differential diagnosis associated with any one manifestation can make the diagnosis of ARF extremely difficult. It is essential to recognize and treat ARF in order to avoid the long-term morbidity. One of the rarest components of ARF is the manifestation of Sydenham's chorea. It is an autoimmune, neuropsychiatric movement disorder that makes up the major criteria alone to diagnose ARF.

An 8-year-old female presents with a two-week history of worsening involuntary movements, difficulty with speech, and functional decline. On physical examination, the child displays non-rhythmic movements, an abnormal gait, and incomprehensible speech. Her oral pharynx is non-erythematous and no exudates are visualized. A new systolic murmur is appreciated and her Anti-Streptolysin O (ASO) was positive. ARF was diagnosed based on the chorea, new murmur, and also the positive ASO. The patient was administered with one dose of IM penicillin benzathine to treat her ARF and commenced on risperidone to help manage the chorea.

ARF is still present in developed nations and hospitalist should be familiar with the varying ways it can present. Sydenham chorea is a disabling feature of ARF that requires early recognition and treatment.

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Introduction

Sydenham chorea is a complication of Acute Rheumatic Fever (ARF) and is one of the leading acquired causes of acute onset chorea in children [1,2]. The Neurotransmitter (NT) dopamine has been shown to be the main NT attributing to the hyperkinetic movements seen in Sydenham's chorea, which is very similar to the presentation in Huntington's disease [3].

In developing countries, the incidence of ARF is about 470,000 cases per year and around 275,000 lead to mortality due to a cardiovascular pathology [2]. This can all be prevented if ARF is treated and diagnosed in time. The official criteria to diagnose ARF are defined by understanding the major and minor criteria that rule the disease. Officially known as the Jones criteria, it was originally described in 1944 by Dr. Jones. The Jones criteria have been constantly revised since 1944 in order to properly diagnose ARF. In order to have ARF, there must be strict evidence of a previous Group A Beta-Hemolytic Streptococcal (GABS) infection with days [4]. This is the first initial step in diagnosing ARF, and without a GABS infection; the probability of ARF is unlikely. A GABS infection can be diagnosed with elevated or rising anti-streptolysin O titers, or other antibodies such as anti-DNase B antibodies (there are multiple that are specific to GABS infections), a positive GABS throat culture, or a positive group A streptococcal carbohydrate antigen test individuals with suspected streptococcus pharyngitis [3].

According to the American Heart Association (AHA), to make the diagnosis of ARF in a low risk area, one individual must satisfy the two out of five major manifestations or one major and two minor manifestations. While in a moderate/high risk area the criteria can also be met with the presence of three minor manifestations. Chorea is the only exception when ARF can be considered without having strict evidence of GAS infection. Chorea, by itself is sufficient criterion to diagnose ARF given that other neurological disorders are ruled out [4] (Table 1).

Case Presentation

An 8-year-old female with no past medical history presents with a two-week history of

Table 1: Jones criteria in low risk populations.

Jones criteria in low risk populations	
Major Criteria	**Minor Criteria**
Subclinical carditis	Polyarthralgia**
Polyarthritis*	Fever (>38.5°C)
Chorea	ESR>60 mm in the first hour***
Erythema marginatum	CRP>3 mg/dL
Subcutaneous nodules	Prolonged PR interval
Medium/High Risk Population:	
*monoarthritis or polyarthralgia	
**monoarthralgia	
* **ESR of >30 mm in the first hour	

worsening involuntary movements, difficulty with speech, and functional decline. The patient recently returned from a three-week trip to the Dominican Republic (DR), where she first started to develop symptoms after 7 days of arriving. Initially the patient began to have some small involuntary tics in her extremities and was noted to be more anxious. Over the course of one week, the tics began to become more of an involuntary jerking movement that affected the entire body including the face and torso. The movements were so severe that the child was no longer able to feed herself, dress herself, or even ambulate without assistance. She also began to have difficulty with speaking and was only able to communicate with one-word sentences, body gestures, and facial expressions.

During the onset of her symptoms, the patient developed two skin abscesses that were noted to drain minimal yellow pus, one located in the gluteal region and one on the scalp. The mother denies any sick contacts, fevers, cough, sore throat, abdominal pain, diarrhea, or head trauma.

The child is noted to be afebrile, alert, and active. She displays non-rhythmic movements, an abnormal gait, and incomprehensible speech. Her oral pharynx is non-erythematous, and no exudates are visualized. Heart sounds are regular rate and rhythm, a 2/6 systolic murmur is appreciated most prominently over the left lower sternal border. Her lungs are clear to auscultation bilaterally. No lymphadenopathy appreciated. Two small abscesses with scabs are noted, one on right buttocks and second on the frontotemporal region of scalp. Cultures were collected from the patient's throat, urine, blood, and cerebral spinal fluids.

Laboratory results

- ASO: Positive
- Rapid Strep Antigen Test: Negative
- White blood cell count: 9.9 × 10^3/uL (5.2-12.4)
- Urine Toxicology: Negative

Findings on imaging

Head MRI showed leptomeningeal enhancements which cannot rule out meningitis, but no masses were visualized. Her electrocardiogram showed normal sinus rhythm and did not show an increase in the PR Interval or signs of atrioventricular blocks. While her echocardiogram showed a left ventricle ejection fraction of 65%, the septum was intact, mild mitral regurgitation, and some trace of tricuspid regurgitation.

Culture results

The patient's cultures of throat, urine, blood, and cerebral spinal fluids did not have any growth after 72 h. Testing for other sources of infection such as Measles, Respiratory Syncytial virus, West Nile virus, Herpes Simplex Virus (HSV), Lyme's disease, and Influenza all came back negative. NMDA antibodies were also negative.

The patient's mother does recall the child have a self-resolving cold a couple weeks prior to the presentation of the acute chorea, but she does not recall the child complaining of any throat pain. On physical exam, the patient's gait was severely affected by the sporadic jerking movements. She was also noted to have a new murmur in the mitral region. Her laboratory results came back negative for rapid strep antigen, but her ASO levels came back elevated. Thus, ARF must be highly considered as the diagnoses.

The patient was administered with one dose of IM penicillin benzathine to treat her ARF and commenced on risperidone 1mg to help manage the chorea. Her symptoms began to improve during her hospital stay. Her chorea movements began to decrease in intensity. She was able to verbalize more phonetic sounds, and slowly began to form complete words. She continued to have elevated DNase B antibody and was started on oral penicillin V twice a day for prophylaxis.

Discussion

The average worldwide incidence of ARF is 19 cases per 100,000 school aged children, while the incidence rate in the USA is only <2 cases per 100,000 annually. Having Sydenham's chorea as the sole manifestation is even rarer of a phenomenon, with only 5% of those 2 per 100,000 reporting in this manner. In these cases, females were affected at a 2:1 ratio compared to males, and most often in the age group of 5 to 16 years old with a peak around the ages of 8 and 9 years old [2].

ARF is a condition that can be prevented with proper treatments and recognition, making it a rarity of a diagnosis in the United States [2]. Primary prevention of ARF requires adequate treatment of Group A Strep (GAS) pharyngitis with either Amoxicillin (50 mg per kg orally once for 10 day with a maximum of 1 gram daily), Penicillin G benzathine (patients weighing 27 kg or less should receive 600,000 units IM once or patients weighing more than 27 kg should receive 1,200,000 units IM once), Penicillin V Potassium (250 mg orally BID or TID for 10 days for <27 kg or 500 mg orally BID or TID for 10 days >27 kg). For patients allergic to penicillin, narrow spectrum Cephalosporins (Cephalexin), Azithromycin, or Clarithromycin are recommended [4].

Secondary prophylaxis of treatment for ARF is aiming at preventing recurrent ARF in those has been previously diagnosed with ARF. According to the American Heart Association (AHA), patients who have ARF with carditis and residual heart disease should receive antibiotic prophylaxis for ten years or until the age of forty years (whichever is longer); some patients need lifetime prophylaxis. Patients with ARF without any evidence of carditis should be treated with antibiotics for at least five years or until age twenty one (whichever is longer). Furthermore, AHA no longer recommends prophylaxis for infective endocarditis in most patients with rheumatic heart disease unless the patients have prosthetic valves, valves repaired with prosthetic materials, heart transplant patients, previous cases of endocarditis, and specific forms of congenital heart disease [4].

Sydenham's chorea is manifestation that is very rare in the world, therefore there is little evidence to support that one drug is proffered over another to treat the patient's symptoms. According to the US Food and Drug Administration (FDA), there is no clear evidence that one antipsychotic, benzodiazepines, or anti-epileptic is preferred over others for the treatment of Sydenham's chorea. However, the FDA does verify the pediatric doses of antipsychotics specifically Haloperidol as well as another antipsychotic called pimozide. Furthermore, the FDA warns that the cardiotoxic side effects of these drugs should be kept in mind when addressing treatment and an electrocardiogram is a standard before treatment [5]. In conclusion, the world is in need for double-blinded, placebo- and comparative-controlled trials to establish evidence based worldwide treatment for Sydenham's chorea.

One study performed in sixty five children in the country of Turkey has shown that haloperidol is superior to pimozide for controlling chorea in terms of recovery and time to remission. Haloperidol is a First-Generation Antipsychotic (FGAs) and has greater potential for toxicities and adverse effects compared to Second-Generation Antipsychotics (SGAs). While both sets of antipsychotics are used in motor disorders as they are all essentially dopamine antagonists, the SGA have proven to have less side effects and better in treating specific disorders rather than simply controlling motor symptoms of a disorder. In a study of haloperidol *vs.* risperidone, it shows that haloperidol is associated with greater severity of extrapyramidal symptoms, but there is no statistical difference in prolactin related measures or sedation [5].

Conclusion

No antipsychotics like all medications are free of adverse effects and the choice of antipsychotic in our case was risperidone due to its lower side effect profile as compared to FGAs as well as other SGAs.

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