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Case Report

Can PANDAS Swear? A Curious Case of Coprolalia in a 15 Year Old Girl Presenting to the Emergency Department

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ABSTRACT

Pediatric autoimmune neuropsychiatric disorders associated with Streptococcal infections, or PANDAS, is a diagnosis of children with an acute and fast onset of obsessions, compulsions or tics succeeding a Group A beta-hemolytic streptococcal infection. Coprolalia is a form of tics where the patient involuntarily says obscene and inappropriate words. We report a case of a 15-year old girl with a history of suspected PANDAS presenting to the emergency department with recurrent coprolalia without signs of a streptococcus infection. PANDAS and other neuropsychiatric syndromes can have different acute presentations. The ED physicians should be familiar with such disorders and presentations.

Key words: neuropsychiatric disorders, pediatric emergency, PANDAS

INTRODUCTION

Pediatric autoimmune neuropsychiatric disorders associated with Streptococcal infections, or PANDAS, is a diagnosis of children following the acute and dramatic onset of compulsions, obsessions or tics following a Group A beta-hemolytic streptococcal (GABHS) infection.[1] This disorder has gained wide attention and controversy since the last decade when Swedo et. al identified 50 pediatric patients in whom GABHS infection triggered acute onset obsessive compulsive disorder (OCD) symptoms and/or tics and therefore led to the development of 5 diagnostic criteria of PANDAS: the presence of OCD or tic disorder as per DSM criteria, pre-pubertal onset, a relapsing and remitting development of illness, presence of a chronological relation between symptoms and GABHS infection and finally, the presence of other neurological symptoms.[2,3]

The most widely accepted hypothesis explaining the pathophysiology of PANDAS is that of molecular mimicry whereby certain antigens on the bacterial surface trigger antibodies that cross react with the basal ganglia of susceptible individuals.[3] Conflicting data has been published however regarding this autoimmune theory, further contributing to the controversy of this entity and posing challenges regarding diagnosis and adequate treatment.[4] This, in addition to difficulty establishing a temporal relationship with GABHS infection, has led to the proposal of new criteria for a broader clinical diagnosis, pediatric acute-onset neuropsychiatric syndrome (PANS) or childhood acute-onset neuropsychiatric syndrome (CANS), suggesting that several infectious agents, rather than Group A streptococcal infection only, may be involved.[4]

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CASE REPORT

A 15-year-old female presented to the pediatric emergency department (ED) for acute onset of verbal and motor tics. She was known to have a major depressive episode (MDE) in 2018 as well as generalized anxiety disorder (GAD), but had been in remission for the past six months. She also had a brief self-resolving episode of facial and motor tics that occurred after a streptococcal infection, for which the presumed diagnosis was PANDAS, also in 2018 prior to her diagnosis with GAD and MDE. The patient was on fluoxetine 30mg per os daily.

The patient described upper and lower motor tics worsening over the past three days. The most common tic was a contraction of her left arm that caused her to hit herself in the chest. She would also regularly contract and arch her back and swing her head back and forth or side to side. She had concurrently developed speech tics, most commonly forming a click sound with her tongue. During the interview, there were several episodes of coprolalia when answering questions about her symptoms or about her family. The father described that the patient had suddenly begun to swear inappropriately when asked any question, often having no relation to the subject being discussed. She had also been described to laugh inappropriately when asked questions. The patient described the spastic movements as sharply painful and worsening in intensity over the past 24 hours. The symptoms were starting to interfere with the quality of her activities of daily living and were affecting her ability to fall asleep while her ability to maintain sleep was unaffected. She denied any substance use, no recent viral infection symptoms, suicidal or depressive ideation, trauma or social stressors.

Her vital signs on presentation were within normal limits (heart rate of 100 beat per minute, blood pressure 118/71, temperature 36.6°C, respiratory rate of 18 breath per minute, SpO2 98% on room air). Her cardiac, upper and lower respiratory as well as abdominal exams were all normal. She had bruising on her right arm (stated to have occurred from hitting herself), as well as healed linear horizontal scarring to her left forearm. Otherwise, she had no bruises or signs of trauma. Her neurological exam revealed symmetrical reflexes in upper and lower extremities, a normal cranial nerve exam, normal gait, normal proprioception and cerebellar function. Her motor strength was appropriate in all four limbs and symmetrical; however, assessment was limited due to the frequency of motor tic movements. Blood work including Antistreptolysin O were normal.

The patient was referred to her treating psychiatrist after neurology team clearance with suggestions to begin a course of clonidine and for further monitoring.

DISCUSSION

PANDAS, as a clinical entity, has become very popular in the past few years yet has caused intense debates among clinicians and researchers; the relationship between the streptococcus infectious exposure and the explosive onset of symptoms, the progression of this disease, and the treatment including the different symptomatic and disease -modifying entities are all issues that remain unanswered and have yet to be addressed.[5]

Usually, PANDAS patients initially present with complex motor and vocal tics in an abrupt fashion, rather than with a progressively more complex tics (that are usually more typical of Tourette syndrome), similar to our presented case.[1] Interestingly, although coprolalia is one of the most notorious symptoms of tic disorder, studies have shown that its incidence is rather low, occurring in less than 25% of patients.[1]

Although the patient was previously diagnosed with PANDAS, her initial presenting symptoms, reported motor and vocal tics at the time, were described as self-limited and completely resolved. She now presented to the ED, 3 years later, with the recurrence of both types of tics. The swift and intense onset of symptoms is very characteristic of PANDAS, as well as the course described by remissions and exacerbations following possible new infections. This pattern is usually helpful in differentiating PANDAS from other forms of tics, Tourette's Syndrome, and OCD, all of which usually have a slow onset.[6] Perhaps the most controversial aspect is the association concerning group A streptococcus and the development of PANDAS.[2] Some population-based studies have been able to prove an association between GABHS infection and the development and worsening of neuropsychiatric symptoms; in fact, in a report by Mell et al., patients with tics and OCD were 2.2 times more likely to have had a streptococcal infection in the 3 months prior to symptoms.[7] Other articles however did not find a connection between this specific infection and exacerbations of proposed PANDAS-related symptoms, further contributing to the confusion regarding this entity.[8] Different infectious agents may cause symptoms through nonspecific immune activation means and therefore all infections should be considered and treated on a case-by-case basis, according to existing recommendations.[5] In some cases, onset and exacerbations may happen in the

absence of any identifiable contagion, suggesting that further disease mechanisms may be implicated; however, this needs to be further studied.[8]

A strong association between patients with PANDAS and first-degree relatives with autoimmune diseases also exists, further supporting the hypothesis that PANDAS might be immune related. This has led to the proposal of the use of immune-modulating agents and the development of guides for the use of these agents, especially in kids with more severe presentations, a chronic-static or a chronic-progressive illness progression.[1]

Interestingly, our patient was diagnosed with GAD and MDE after her diagnosis with PANDAS. Both anxiety and depression have been shown to be very common in patients diagnosed with PANDAS.[9] Both disorders require treatment that could range from psychotherapy to pharmacotherapy, depending on the severity of symptoms[9]; our patient necessitated treatment with the selective serotonin reuptake inhibitor, fluoxetine, suggesting that her symptoms were impacting functionality. In fact, in a study by Leon et al., 33% of patients diagnosed with PANDAS received a psychiatric diagnosis during follow-up.[10] This highlights the importance of regular follow-up and neuropsychiatric assessments in these specific patients as timely and correct diagnosis of these syndromes will allow the use of targeted and evidence-based interventions.

CONCLUSION

PANDAS and other neuropsychiatric syndromes can have different acute presentations. The ED physicians should be familiar with such disorders and presentations as they are associated with subsequent development of other long-term psychiatric conditions.

Informed consent: Patient's informed consent was obtained for the publication of this case report.

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REFERENCES

1. Blackburn JS. Tic disorders and PANDAS. Semin Pediatr Neurol. 2018;25:25–33. https://doi.org/10.1016/j.spen.2017.12.003

2. Shulman ST. Pediatric autoimmune neuropsychiatric disorders associated with streptococci (PANDAS): update. Curr Opin Pediatr. 2009;21(1):127-30. https://doi.org/10.1097/MOP.0b013e32831db2c4

3. Swedo SE, Grant PJ. Annotation: PANDAS: a model for human autoimmune disease. J Child Psychol Psychiatry. 2005;46(3):227-34. https://doi.org/10.1111/j.1469-7610.2004.00386.x

4. Baytunca M, Donuk T, Erermis HA. Evaluation of a neuropsychiatric disorder: from PANDAS to PANS and CANS. Turk Psikiyatri Dergisi. 2016;27(2). https://pubmed.ncbi.nlm.nih.gov/27370066/

5. Chiarello F, Spitoni S, Hollander E, Matucci Cerinic M, Pallanti S. An expert opinion on PANDAS/PANS: highlights and controversies. Int J Psychiatry Clin Pract. 2017;21(2):91-8. https://doi.org/10.1080/13651501.2017.1285941

6. Gilbert DL. Inflammation in tic disorders and obsessive-compulsive disorder: are pans and PANDAS a path forward?. J Child Neurol. 2019;34(10):598-611. https://doi.org/10.1177/0883073819848635

7. Schrag A, Gilbert R, Giovannoni G, Robertson MM, Metcalfe C, Ben-Shlomo Y. Streptococcal infection, Tourette syndrome, and OCD: is there a connection? Neurology. 2009;73(16):1256-63. https://doi.org/10.1212/WNL.0b013e3181bd10fd

8.Leckman JF, King RA, Gilbert DL, et al. Streptococcal upper respiratory tract infections and exacerbations of tic and obsessive-compulsive symptoms: a prospective longitudinal study. J Am Acad Child Adolesc Psychiatry. 2011;50(2):108-18.e3. https://doi.org/10.1016/j.jaac.2010.10.011

9. Thienemann M, Murphy T, Leckman J, et al. Clinical management of pediatric acute-onset neuropsychiatric syndrome: part I—psychiatric and behavioral interventions. J Child Adolesc Psychopharmacol. 2017;27(7):566-73. https://doi.org/10.1089/cap.2016.0145

10. Leon J, Hommer R, Grant P, et al. Longitudinal outcomes of children with pediatric autoimmune neuropsychiatric disorder associated with streptococcal infections (PANDAS). Eur Child Adolesc Psychiatry. 2018;27(5):637-43. https://doi.org/10.1007/s00787-017-1077-9