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Case Report / Vaka Sunumu

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A possible variant of PANDAS: Dystonic subtype

Olası bir PANDAS varyantı: Distonik alt tip

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ABSTRACT

Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections (PANDAS) is characterized as exacerbation of obsessive-compulsive disorder and/or tic disorder secondary to streptococcal infections. Recently accumulated data suggest that PANDAS have some variants such as myoclonic type, adult onset type, infantile-onset type and dystonic type. In this report, we present a 17 year-old patient who was treated with antidepressant and antipsychotic medicine for a long time until the diagnosis of late-onset PANDAS dystonic variant was made.

Keywords: *PANDAS, late onset, antibiotic treatment, dystonic variant*

ÖZ

Streptokok enfeksiyonu ile ilişkili pediatrik otoimmun nöropsikiyatrik bozukluk (PANDAS) streptekok enfeksiyonu sonrası alevlenen obsesif kompulsif bozukluk ve/veya tik bozukluğudur. Güncel biriken bilgiler PANDAS'ın bazı varyasyonları olduğunu söylemektedir; infantil başlangıçlı tip, yetişkin başlangıçlı tip, myoklonik tip ve distonik tip. Bu yazıda, 17 yaşında olup, antidepresan ve antipsikotik ile uzun süre tedavi edildikten sonra geç başlangıçlı PANDAS distonik tip tanısı konulan bir vakayı sunacağız.

Anahtar kelimeler: *PANDAS, geç başlangıçlı, antibiyotik tedavisi, distonik alt tip*

INTRODUCTION

Streptococcal infections are very common in childhood. Following group A streptococcal (GAS) infections, the sudden onset of tics/obsessive-compulsive behaviours has been diagnosed as a pediatric autoimmune neuropsychiatric disorder associated with streptococcal infections (PANDAS). PANDAS was firstly described by Swedo et al.[1] as having five components, including early onset, tics/obsessive-compulsive disorder (OCD), relationship with GAS infection, an intermittent course of symptom severity and neurological abnormalities. Although PANDAS usually occurs secondary to oro-pharyngeal infections, cases secondary to skin infections have been reported[2]. Some post-streptococcal autoimmune disorders have been diagnosed, including paroxysmal dystonic choreoathetosis[3], acute disseminated encephalomyelitis[4], dystonia[5], myoclonus[2] and anorexia nervosa[6]. In addition to clinical variants of PANDAS, other classifications have been reported based on age of onset (infantile or adult)[1,7], neurological features (myoclonic)[2] and psychiatric features (body dysmorphic disorder, attention deficit and hyperactivity disorder)[8,9]. The underlying pathophysiology may be autoimmune, with an autoantibody that attacks basal ganglia, and activated by streptococcal infection[10]. Kumar et al.[11] demonstrated increased neuroinflammation in the lentiform nucleus and bilaterally in the caudate nucleus in PANDAS patients compared with those in healthy controls. We present here a case of PANDAS with a dystonic variant, which was treated with antibiotics.

CASE

A 17-year-old male patient, the younger of two siblings, who lives in the city centre with his family, was admitted to our clinic because of involuntary movements. There was no medical history until four

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months earlier, when a contamination obsession first occurred, followed by phonic tics in the form of howling. To accommodate the howling, the patient went to the bathroom more often. He stated that he was in distress and would be bored stiff if he did not yell. Subsequently he developed a religious obsession (thinking he was a sinner). His symptoms were exacerbated by stress. He was anxious, with emotional lability and shaking hands. At the time of admission, the patient's Yale-Brown obsession score (Y-Brown) was 32. He was repeating some words for a couple of days. When interviewed, phonic tics were observed. The patient stated that he felt a voice of "shout" inside of himself. After shouting, he felt sad and guilty. Before being admitting to our clinic, the patient was treated for a month in another psychiatric inpatient clinic with risperidone, fluvoxamine, ziprasidone, clomipramine and olanzapine, consecutively. Because the decrease of his symptoms with treatment was not sufficient, the patient was referred to our clinic.

Physical examinations and investigations (including a complete blood count (CBC), biochemical analyses, a brain MRI, C-reactive protein (CRP) levels and erythrocyte sedimentation rate (ESR)) were normal, but his serum antistreptolysin O (ASO) titer was twofold higher (520 IU) than the normal reference range (0-200 IU). A throat culture was negative. He was on 150 mg clomipramine, and we added 2 mg clonazepam, 2 mg haloperidol (for phonic tics) and propranolol (for shaking hands). When asked again, his mother told us that the patient had had tonsillitis just before his symptoms started. In addition to psychiatric treatment, we started the patient on penicillin G benzathine (1.2000.000 IU IM) and amoxicillin clavulanate 2000 mg orally. After antibiotic treatment, the patient's symptoms (obsessions and phonic tics) and his Y-Brown score significantly decreased.

A couple of days before his planned discharge, the patient had a sore throat and his symptoms increased. A physical examination revealed tonsillitis. At this time, dystonia had developed, so the patient was started on 2 mg biperiden. Antibiotic treatment was restarted, and the clomipramine dose was increased to 225 mg. The patient's symptoms were reduced dramatically after antibiotic treatment, and he was discharged. His symptoms were considered to be related possibly to PANDAS, and the patient was continued on clonazepam 2 mg/day, clomipramine 225 mg/day and penicillin G benzathine 1.2000.000 IU for three weeks.

DISCUSSION

The patient's abrupt onset of OCD and/or tic symptoms, episodes of severe symptoms association with GAS infection and neurological abnormalities met the criteria of PANDAS. His symptoms started after tonsillitis, decreased with antibiotic treatment and increased again after recurring tonsillitis during treatment. Although the patient was 17 years old which was a late age for onset of PANDAS, other late-onset cases have been also reported[2]. Various OCD symptoms may arise after GAS infection[12], and PANDAS may account for up to 25% of all pediatric OCD cases[13]. A 10-year-old boy presented with decreasing school performance[14]. The present case had manifestations of loud shouting and obsessions with contamination and religion. Although there was no growth on a throat culture, a high ASO level and exacerbation of the patient's symptoms after tonsillitis suggested PANDAS. Dale et al.[5] reported post-streptococcal autoimmune dystonia with basal ganglia necrosis. The dystonia that occurred in the course of illness in the present case could be due to the antipsychotic medication, although the dose that the patient had been taking (2 mg haloperidol for a month) does not typically result in dystonia. Additionally, the dystonia developed just after an infection and resolved after antibiotic treatment, indicates that the dystonia was associated with PANDAS and not with the antipsychotic medicine.

In this case, there was no significant improvement until antibiotic treatment was started, providing additional support for a diagnosis of PANDAS. Penicillin is an easily accessible and effective treatment for patients with PANDAS. Anti-inflammatory medicine can also be used. If antibiotic treatment is insufficient,

other treatment options that have proven successful in reducing OCD symptoms in children with PANDAS are therapeutic plasma exchange and intravenous immunoglobulin (IVIG) administration[15].

Since PANDAS is associated not only with streptococcal infections, in Diagnostic and Statistical Manual of Mental Disorder 5 (DSM 5), the name was changed from PANDAS to PANS (Pediatric Acute-Onset Neuropsychiatric Syndrome), and it was moved to the category of Other Obsessive Compulsive and Related Disorders[16]. This categorisation emphasises the symptoms instead of the etiology.

The present case will help professionals to look at PANS from a different angle. Though PANS is known as a rare disease, some professionals claim that it is much more common than generally thought[17]. Clinicians who have pediatric patients with unusual or treatment-resistant symptoms should review the patient's diagnosis with an emphasis on the patient's medical history, keeping PANS in mind. Patients with PANS may have uncharacteristic symptoms, which can be effectively resolved with antibiotics.

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