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To cite this article: Karin Frånlund & Charbél Talani (2023) PANDAS – a rare but severe disorder associated with streptococcal infections; Awareness is needed, Acta Oto-Laryngologica Case Reports, 8:1, 104-107, DOI: 10.1080/23772484.2023.2231146

To link to this article: https://doi.org/10.1080/23772484.2023.2231146

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Published online: 06 Jul 2023.

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PANDAS – a rare but severe disorder associated with streptococcal infections; Awareness is needed

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**ABSTRACT**

PANDAS is an acronym for ‘paediatric autoimmune neuropsychiatric disorders associated with streptococcal infections’ and is defined as a subgroup of PANS, ‘Paediatric acute-onset neuropsychiatric syndrome’. PANDAS is considered an autoimmune disease based on a streptococcal infection with a verified immune response. The onset is often dramatic, with symptoms of OCD developing within 24-48h. Symptoms include both psychological symptoms and physical symptoms. Treatments of PANDAS must address both physical and psychiatric symptoms. Supplemental treatments for PANDAS include tonsillectomy to prevent further streptococcal infections. We present a case of PANDAS with the aim of raising awareness of its diagnosis. PANDAS is a rare condition with an impact on the patient’s quality of life; thus, awareness is needed. A multidisciplinary team is needed for diagnosis and treatment. Otorhinolaryngologists may play an important role in the well-being of these patients when performing tonsillectomy.

**Background**

PANDAS is an acronym for ‘paediatric autoimmune neuropsychiatric disorders associated with streptococcal infections’ and is defined as a subgroup of PANS, ‘Paediatric acute-onset neuropsychiatric syndrome’. Although the validity of the diagnosis has been debated, criteria for PANS have recently been established (Table 1) \cite{1}. PANDAS is a rare condition and might be hard to diagnose.

PANDAS was first described by Swedo et al. in 1998 in 50 cases with sudden onset of obsessive-compulsive disorder (OCD) and behavioural disorders in a group of children with B-haemolytic streptococcal infection. It affects males more frequently than females 2:1, and occurs most frequently among children aged between 3 and 12 \cite{1, 2}. PANDAS is considered an autoimmune disease based on a streptococcal infection with a verified immune response. Molecular mimicry is the most frequent physio-pathological mechanism, where a foreign antigen shares structural or sequence similarities with self-antigens \cite{3}. Streptococcal infections hide from the host immune system by mimicking host cells. This causes the production of cross-reactive antibodies that can cross the blood–brain barrier. Anti-neuronal autoantibodies react with autoantigens in the basal ganglia and cortical structures, causing the onset of neuropsychiatric symptoms \cite{4}.

The onset is often dramatic, with symptoms of OCD developing within 24-48h. The psychological symptoms may include repetitive, obsessive-compulsive behaviours, fear, separation anxiety, panic attacks, irritability, frequent mood changes, emotional and developmental regression, visual or auditory hallucinations, depression and suicidal thoughts. The physical symptoms may consist of sensitivity to light, sound and touch, tics, deterioration of motor abilities (tested by

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handwriting), hyperactivity, memory problems, sleep disorders, refusal to eat and weight loss, joint pain, frequent urination and enuresis. Unlike other childhood psychiatric diseases that develop gradually, symptoms of PANDAS develop quickly and can reach maximum severity as early as within 2-3 days [3].

PANDAS is a rare but severe disorder, and there is no biomarker for the syndrome [3]. It is a clinical diagnosis and requires a thorough medical history and physical examination. Parents or caregivers play an important role in describing the symptoms. Laboratory tests included complete blood count, erythrocyte sedimentation rate, C-reactive protein, urine analysis, pharyngeal swab and anti-streptococcal antibodies. Children with psychiatric and neurological signs require analysis of the cerebrospinal fluid (CSF) and neuroimaging exams [5]. Differential diagnosis includes other disorders with psychiatric manifestations, such as Sydenham’s chorea, Tourette’s syndrome, OCD, central nervous system vasculitis, autoimmune encephalitis and neuropsychiatric lupus [3,4]. As a diagnosis of exclusion, PANDAS is diagnosed after all other possibilities have been eliminated [3].

Treatment of PANDAS must address both physical and psychiatric symptoms. Cognitive behavioural therapy is the primary evidence-based therapy for OCD, supplemented with a small dose of selective serotonin reuptake inhibitors (SSRIs) when indicated [6]. Treatment of the streptococcal infection is often post-fiftum but may include amoxicillin, penicillin, azithromycin and cephalosporins. The evidence for using antibiotics for PANS and PANDAS is inconclusive, but it is assumed that certain anti-streptococcal antibiotics may have a neuroprotective effect [3,7]. Supplemental treatments for PANDAS include tonsillectomy to prevent further streptococcal infections. To suppress the immune system and reduce OCD symptoms, corticosteroids, therapeutic plasma exchange (TPE) and intravenous immunoglobulin (IVIG) or anti-CD20 monoclonal antibodies (rituximab) have been given [8].

PANS and PANDAS are distressing for both the affected child and the parents and cause parental stress due to fear of the symptoms, frustration from the lack of scientific knowledge of the disorder and the perception of not being heard by members of the health care system [9].

The aim of this case report is to spread knowledge of this disorder after our search in today’s literature and present a case of PANDAS to raise awareness among fellow ear, nose and throat (ENT) practitioners.

Case report

A 9-year-old girl was referred from the primary care unit for an ENT assessment of the tonsils. She had a previous history of mononucleosis and tonsillitis 3 years prior but was not found to be a candidate for tonsillectomy due to transient symptoms. Her tonsils were then assessed as Brodsky grade 2 [10].

She had a Streptococcus group A verified pharyngeal infection in March 2020 treated with penicillin V. A few months later, she slowly developed progressing anxiety and obsessions. Her parents wrote a self-referral to the child and adolescent psychiatrist clinic and were placed in queue. While the symptoms worsened during the months in queue, the parents googled her symptoms and were the ones who first came to suspect PANDAS. Isolation due to the SARS-CoV-2 pandemic worsened her symptoms. Her symptoms were daily and excessive and prevented her from participating in social activities and attending school. Investigation at a private child and adolescent psychiatrist office was commended. In October 2021, she was referred to our ENT clinic for preoperative assessment of tonsillectomy. Examination showed hypertrophic tonsils Brodsky grade 2-3 and sore jugulodigastric lymph nodes [10]. Self-administered NSAID treatment had decreased the OCD symptoms, and she agreed to tonsillectomy, which had previously been out of the questions due to fear and anxiety.

Table 1. Diagnostic criteria for PANS [1].

| Abrupt, dramatic onset of OCD or severely restricted food intake |
| Additional neuropsychiatric symptoms, ≥2 of the following: |
| • Anxiety |
| • Emotional lability and/or severely oppositional behaviour |
| • Behavioural regression |
| • Deterioration in school performance |
| • Sensory or motor abnormalities |
| • Somatic symptoms, including sleep disturbances, enuresis or urinary frequency |
| Symptoms not better explained by a neurological or medical disorder |
Tonsillectomy was thought to lessen the inflammatory burden.

Her parents had made a thorough review of available literature on the subject and insisted upon a preventive antibiotic treatment to minimize the risk of neural damage. Preoperative prophylactic penicillin V was prescribed after consultation with her child and adolescent psychiatrist. Bacterial cultures taken from the pharynx were negative. There were no neurological symptoms, i.e. motor impairments. Twenty days after the first visit at our ENT clinic, she underwent tonsillectomy and abrasion of a small residual adenoid, still under prophylactic antibiotics. Due to complications with minor intraoperative bleeding in the nasopharynx, she stayed overnight at the children’s ward. To ensure a neuroprotective effect perioperatively, the antibiotic regimen was changed to amoxicillin/clavulanic acid for 2 weeks postoperatively. NSAID treatment was eliminated 5 days after surgery to minimize the risk of postoperative haemorrhage.

At follow-up three weeks later, the patient self-estimated that her symptoms had gradually decreased, but her parents had not noted any dramatic changes, possibly small ones. The symptoms had, most importantly, not increased. The pharynx had healed as expected, and no sore lymph nodes were noted at palpation. The patient was undergoing cognitive therapy at the child and adolescent psychiatrist clinic for her psychiatric symptoms.

At follow-up at the neuroimmunology team at the child and adolescent psychiatric clinic 6 months later, she had made an almost complete recovery and had had a good effect from NSAIDs. The caregivers contributed this to the tonsillectomy, after which they started to notice the recovery. When infected by the common cold or a smaller infection, OCD symptoms recurred, and she was then administered NSAIDs for relief. Cognitive therapy given for 6 months had been completed. The final diagnosis was suspected PANDAS.

Discussion

Otorhinolaryngology symptoms (ENT) in a paediatric population affected by PANDAS were reported in approximately 67.7% of patients, suggesting that PANDAS patients may undergo a specific otolaryngologic consultation [11].

Our case had symptoms in line with previous studies, including a history of streptococcal infection and abrupt onset of OCD, where neither parents nor family practitioners initially recognized the condition. Due to deeply committed parents, the diagnosis was thought of, and the child was referred to the child and Adolescent Psychiatrist Clinic for investigation.

Knowledge of this disorder is important due to the abrupt onset of symptoms that may lead to patients and caregivers searching medical care not primarily at child and adolescent psychiatrists but more likely at their family practitioner or their closest ENT clinic. A multidisciplinary team needs to be involved in the investigation and treatment. Supportive therapies may be indicated, not only for the affected child but also for his or her parents or caregivers, as the symptoms of PANDAS can be distressing and most often affect the whole family [1,9].

Conclusion

PANDAS is a rare condition with an impact on the patient’s quality of life; thus, awareness is needed. A multidisciplinary team is needed for diagnosis and treatment. The finding from this case report mirrors the findings from other studies noting that parents are usually the first ones to suspect PANS [9].

Otorhinolaryngologists may play an important role in the well-being of these patients when performing tonsillectomy.

Informed consent

Informed consent was given by the patient and caregivers.

Disclosure statements

No potential conflict of interest was reported by the author(s).

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