■ CASE STUDIES

# PANDAS Syndrome: A New Tonsillectomy Indication?

*Ángel Batuecas Caletrío, Fernando Sánchez González, Santiago Santa Cruz Ruiz, Pablo Santos Gorjón, and Pedro Blanco Pérez*
Servicio de Otorrinolaringología y Patología Cérvico-Facial, Hospital Universitario de Salamanca, Salamanca, Spain

PANDAS Syndrome (Paediatric Autoimmune Neuropsychiatric Disorders Associated with *Streptococcus*) is a rare disease described in 1998. In this disease, there is a relationship between group A beta haemolytic streptococcal tonsil infections and the exacerbation of neuropsychiatric disorders.
A case report of a 9-year-old child with PANDAS syndrome is presented. This child has had no further symptoms after tonsillectomy. The understanding about PANDAS syndrome and tonsillectomy is reviewed.

**Key words:** PANDAS syndrome. Group A beta haemolytic *Streptococcus*. Tonsillectomy.

**Síndrome PANDAS: ¿una nueva indicación de amigdalectomía?**
El síndrome PANDAS *(pediatric autoinmune neuropsychiatric disorders associated with* Streptococcus) es una entidad rara que se describió en 1998. Los pacientes con esta afección presentan una exacerbación de los síntomas neuropsiquiátricos coincidiendo con las infecciones amigdalares causadas por *Streptococcus* betahemolítico del grupo A.
Presentamos el caso clínico de un paciente de 9 años con síndrome PANDAS sometido a amigdalectomía que mejoró de su sintomatología tras la intervención. Del mismo modo revisamos la literatura sobre esta entidad.

**Palabras clave:** Síndrome PANDAS. *Streptococcus* betahemolítico del grupo A. Amigdalectomía.

## INTRODUCTION

PANDAS syndrome (paediatric autoimmune neuropsychiatric disorders associated with Streptococcus) has recently been described and included among childhood neuropsychiatric disorders, in which symptoms such as tics or manifestations typical of obsessive-compulsive disorder are exacerbated following streptococcal infections.

Because it is a recently described disease and therefore not fully known, there is no full consensus with regard to diagnostic criteria and the most appropriate treatment for these patients. There is also no agreement as to whether or not these patients should undergo tonsillectomy.

Few cases of this disease in connection with tonsillectomy have so far been reported; hence, this treatment is not standardized.

We present here a case study of a boy with PANDAS syndrome who underwent tonsillectomy and improved after surgery.

Correspondence: Dr. A. Batuecas Caletrío.
Gustavo Doré, 2-12, p 6, 3.º A. 37007 Salamanca. España.
E-mail: abatuc@yahoo.es

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## CASE STUDY

A 9-year-old boy was referred to our surgery due to recurrent bacterial tonsillitis and worsening eye tics and facial grimaces. This exacerbation coincided with acute infection and improved after the infectious process resolved, although the tics did not disappear entirely. He was diagnosed as having PANDAS syndrome.

Furthermore, the patient had night time snoring with respiratory pauses, observed by his parents, without any other associated symptoms.

On otorhinolaryngological examination, bilateral hypertrophy of the tonsils was seen, larger on the left, as well as a slight adenoid hypertrophy; the rest of the examination was normal.

The complementary tests were performed with the following noteworthy results: an analysis in which the antistreptolysin antibody (ASLO) titres were high (550 µm/mL), the culture of the tonsillar exudate isolated *Streptococcus pyogenes* and a polysomnography recorded 125 respiratory events, with an apnoea-hypopnoea rate of 16.9 per hour of sleep with a mean duration of 7 s.

Once the patient had been evaluated, it was decided to perform a tonsillectomy to treat obstructive sleep apnoea syndrome (OSAS), since PANDAS syndrome has not been established as an indication for tonsillectomy.

After the tonsillectomy had been performed and the patient had been seen at the clinic on a regular basis, the eye and facial tics were seen to have disappeared, as had the clinical symptoms of OSAS.

## DISCUSSION

Tics and obsessive-compulsive disorders (OCD) are fairly common during childhood. They affect 1% to 4% and 2% to 3% of the paediatric population, respectively.[1-3]

Swedo[4] identified a sub-group of patients whose tics and/or obsessive-compulsive disorder temporarily got worse during infections provoked by group A beta-haemolytic *Streptococcus*. This condition has been encompassed under the acronym PANDAS.[4]

The diagnosis of PANDAS syndrome is based on the following criteria[5]:

– Tics or OCD
– Onset during childhood (between 3 years of age and puberty)
– Episodic presentation (sudden onset of symptoms, with subsequent remission and sudden reappearance of symptoms)
– Infection due to group A beta-haemolytic *Streptococcus* (determined by pharyngeal culture or elevated ASLO)
– Abnormal neurological signs (abnormal movements, hyperactivity...)

The patient presented here meets all the criteria for PANDAS syndrome, with childhood onset of symptoms and exacerbation of the clinical signs coinciding with recurrent bacterial tonsillitis.

The physiopathology of PANDAS syndrome is believed to be similar to Sydenham's chorea. The antibodies generated by a cross immune reaction would act on the basal ganglia, causing the reported movement and behavioural disturbances.

Children with PANDAS syndrome display no symptoms or symptoms are minimal outside infectious processes. There should be at least 2 episodes during which there is a clear relation between *Streptococcus* infection and clinical exacerbation. During the acute episode, the ASLO titre can be ordered. Elevated titres do not confirm the diagnosis; there must later be a subsequent decrease.[6,7]

These patients usually also exhibit symptoms of anxiety, emotional lability, attention difficulties, hyperactivity, and others.[6]

No treatment has been defined for PANDAS syndrome patients. The use of penicillin has not been proven useful in preventing clinical recurrences.[8]

Several articles show positive results from antibiotic therapy during acute episodes, with disappearance of the psychiatric clinical symptoms.[9,10] In some cases, cognitive behavioural therapy[11] or intravenous immunoglobulin and plasmapheresis have been applied with favourable outcomes.

Tonsillectomy is not included as an indication in the criteria established by the Academy of Otorhinolaryngology and Head and Neck Surgery. Its indication in the treatment of PANDAS syndrome is controversial and debated. They tend to be patients who present recurrent tonsillitis, and in whom the clinical symptoms of tics or OCD improve after tonsillectomy.

Orvidas et al[6] report 2 clinical cases. They are 2 siblings with recurrent tonsillitis and PANDAS syndrome. The neuropsychiatric symptoms disappeared after they underwent tonsillectomy.

Heubi et al[7] report clinical improvement in 2 patients with OCD and tics, with recurrent tonsillitis, who improved after removal of their tonsils.

However, Arostegui et al[10] report 1 patient who displayed OCD 1 week after tonsillectomy, with elevated ASLO titre after surgery.

In our case, the patient underwent tonsillectomy for OSAS. The eye tics and facial grimaces disappeared after the surgery was performed.

Despite the fact that in our case the tonsillectomy resulted in resolution of the patient's neuropsychiatric disorder, as well as his OSAS, we believe that broader studies are needed to conclude that tonsillectomy can be included as standard treatment for patients with PANDAS syndrome.

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