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RESEARCH ARTICLE

BEHAVIOURAL AND COGNITIVE OUTCOMES IN A CHILD WITH TRAPS SYNDROME: A CASE STUDY

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ABSTRACT

Tumour Necrosis Factor Receptor-Associated Periodic Syndrome (TRAPS) is a rare autosomal dominant autoinflammatory disorder and the symptoms include recurrent fevers, significant multisystem involvement and systemic inflammation. (1)The physical symptoms of TRAPS, such as abdominal pain, arthralgia, and skin rashes, are well known but its impact on neurodevelopmental and behavioural outcomes is less understood. This case report presents a twelve-year-old male child diagnosed with TRAPS presenting with difficulties in attention, emotional dysregulation, and frustration, especially in academic settings. The patient is verbally expressive but struggles with writing and reading, requiring additional support during tests. Physical symptoms of TRAPS and well managed with canakinumab, though behavioural and cognitive challenges persist. Long-term neurodevelopmental impact of TRAPS is not well known and further research is necessary to understand how chronic inflammation and recurrent symptoms affect cognitive and emotional development.

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INTRODUCTION

TRAPS is caused due to a mutation in the TNFRSF1A gene, which encodes the TNF receptor-1 (TNFR1). The receptor's capacity to govern immunological responses is compromised by these mutations, which results in prolonged and uncontrollable inflammation.(2)Recurrent episodes of fever, stomach pain, arthralgia, myalgia, and a migrating erythematous rash are some common symptoms of TRAPS. Physical stress, infections, and hormonal changes, can cause inflammatory episodes in TRAPS patients, however the primary cause is poorly understood. (1)TRAPS patients also may show elevated levels acute-phase reactants, specifically serum amyloid A (SAA) which predisposes patients to amyloidosis causing a serious consequence linked to renal failure and higher mortality (3). TRAPS and Familial Mediterranean Fever (FMF) have similar presentations hence were initially confused.

But TRAPS was differentiated from FMF because of its autosomal dominant inheritance pattern and lack of sensitivity to FMF treatments like colchicine. Since then, developments in molecular genetics have established the involvement of TNFRSF1A mutations, which often impacts the extracellular domain of the receptor and causes misfolding and improper shedding of TNFR1. The overproduction of proinflammatory cytokines, especially IL-1β, results in an excessive inflammatory response (4). The impact of TRAPS on neurodevelopment and behaviour is still poorly understood, despite substantial advancements in our understanding of the molecular and immunological mechanisms underlying the condition.Cognitive delays, emotional dysregulation, and behavioural difficulties can be an outcome of chronic inflammation and recurrent pain which can disrupt normal neurodevelopment in children. The variable nature of TRAPS episodes, which can interfere with schooling, social interactions, and emotional well-being can make these challenges exacerbated.

Case Report: This case report presents atwelve-year-oldmale child referred by a speech and language therapist, who has been working with him for over a year. The patient was diagnosed with Tumor Necrosis Factor Receptor-Associated Periodic Syndrome (TRAPS), a rare genetic disorder. His initial symptoms included recurrent fevers lasting three weeks, migrating rashes, abdominal pain, headaches, conjunctivitis, and swelling around the eyes. According to thefamily members the patients is verbally expressive, but struggles with writing, reading, and spacing words, making his written work difficult to comprehend. They also report that he experiences frustration and attention difficulties, especially during tests, where he requires support from a teacher or a peer to help write for him. He often takes a leadership role, but tends to get into conflicts when things do not go his way. They describe him as impatient and fidgety but deny any depression, anxiety, or social anxiety. His parents note that he is good at initiating tasks, maintaining focus when interested, and does not struggle with procrastination or time management.

According to his speech and language therapist, the patient has made modest progress despite receiving extensive therapy and special education services. He continues to struggle significantly with attention, even in a quiet one-on-one setting, and often fidgets or gets up to move around the room. He has notable delays in both receptive and expressive language skills, as well as literacy skills, which hinder his academic progress. He also faces challenges with executive functioning especially in attention, working memory, problem-solving, and emotional regulation create further creating difficulties in both academic and social environments. The patient was born full term at 5.10 lbs after a normal delivery. No NICU care was required. At ten months of age, he started experiencing recurrent high fevers and abnormal white blood cell counts, leading to his eventual diagnosis of TRAPS. Despite his medical conditions, he met normal developmental milestones. He is currently being treated with canakinumab, which is an FDA-approved medication for managing TRAPS symptoms.

DISCUSSION

The patient's developmental curve may have been affected by his diagnosis of TRAPS syndrome, especially with regard to his academic and behavioural progress.His repeated absences from school due by the persistent fevers and long-term illness as a young childmight have exacerbated the delays in language and executive functioning. Despite these difficulties, he exhibits good leadership and social skills, which are positive signs of his ability to adjust in non-academic contexts. This case indicates that despite cognitive and intellectual strengths, the patient struggles with significant untreated executive dysfunction, leading to challenges in attention and impulse control. This is common in neurodevelopmental disorders, such as ADHD. The patient's challenges in maintaining concentration, especially in educational settings, emphasise how critical it is to take into consideration both his neurobehavioral requirements and his physical health associated to TRAPS. For children like this stimulant medications such as methylphenidate and amphetamine salts are often effective in managing ADHD symptoms. However, these medications may not always be well-tolerated due to side effects, including decreased appetite and insomnia.

In cases where stimulants are not suitable, non-stimulant alternatives like alpha-agonists can be considered. These medications target different neurotransmitter systems, offering a more tailored approach to managing attention and impulse control without the typical side effects associated with stimulants. This case demonstrates how difficult it is to treat ADHD when coexisting with a rare illness such as TRAPS, where controlling systemic inflammation is crucial yet neurodevelopmental issues necessitate cautious and frequently distinct attention. Given the chronic nature of both TRAPS and ADHD, early and consistent intervention, including both medication and behavioral therapies, is critical in supporting long-term development.

CONCLUSION

This case demonstrates the challenges of addressing major neurodevelopmental and behavioural disorders like as impulsivity and inattention linked to ADHD in addition to TRAPS. The patient's physical symptoms have improved with canakinumab treatment; nevertheless, because his executive dysfunction is unaddressed, his emotional and academic problems still persist. Treating the patients ADHD symptoms requires a customised strategy that includes behavioural therapies in addition to stimulant or non-stimulant drugs. To maximise his developmental trajectory, behavioural interventions, cognitive therapy, and continuous educational assistance are crucial in addition to pharmacological interventions. Continued research into the neurodevelopmental implications of TRAPS is necessary to enhance our understanding and improve treatment strategies.

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