Case Report

Juvenile Allergic Urethritis with Urethro-vasal Reflex Masquerading Cauda Equina Syndrome

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We report a case of severe Juvenile Allergic Urethritis secondary to double concentrate orange squash of a famous brand in a 3-year-old boy who presented with urethral and perineal pain resulting in an abnormal gait and urinary symptoms suggestive of Cauda Equina Syndrome. Ultrasound of the Urinary Tract was normal as was the Magnetic Resonance Imaging (MRI) of the Spine. Withdrawal of the allergen produced complete recovery. Symptoms recurred on food challenge. There are several learning points and take-home messages in this case such as (1) Allergic Urethritis can have a dramatic presentation, mimicking serious conditions such as Cauda Equina Syndrome. (2) Food challenge provided the definitive diagnosis: this is the first report of double concentrate orange squash induced urethritis. (3) Complete avoidance has resulted in an enduring cure. (4) Appropriate timely referral by general Practitioner and cohesive and well-coordinated multidisciplinary team management at the University Teaching Hospital is required to successfully manage such rare and challenging case.

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Key words **: Juvenile Allergic Urethritis, Urethro-vasal Reflex, Cauda Equina Syndrome, Congenital Secreto-motility Disorders**

In the last four decades, there has been a dramatic increase in the number of children suffering from allergic diseases eg, Asthma, Hay fever, Eczema and Food allergies. In the UK, 18 million people have an allergy, 12 million people will be receiving allergy treatment in any one year, 6 million people will have sufficiently serious allergy to require specialist help and hospital admissions for allergy increased three-fold during the 1990s[1]. Allergies have a tremendous impact on the quality of life of children, affecting a child's growth, development, Educational, Social and Psychological well being[2]. Organ systems most commonly affected are those with direct exposure to an allergen, ie, Skin, Respiratory and Gastro-intestinal Systems. Primary involvement of Genitourinary Tract in allergic reactions is rare. We report a case of Allergic Urethritis secondary to double concentrate orange squash in a 3-year-old boy.

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Editor's Comment :

- Juvenile Allergic Urethritis is an emerging new condition secondary to food processing and preservative sensitivity which can have dramatic presentation mimicking serious conditions secondary to the histamine release and tissue edema.
- Food avoidance leads to complete recovery of symptoms and food challenge causes discovery of symptoms thus help to establish clinical diagnosis. High index of suspicion and early referral to specialist help reduce morbidity.

Case Report

A 3-year-old boy first presented to their General Practitioner with a six-week history of intermittent abnormal wide-based tip-toe Gait and Urethral Dysuria. He was otherwise fit and well, with normal developmental milestones and stable family dynamics. He had commenced walking at 10 months and achieved dryness by day one month before the onset of symptoms.

Having previously been fully-ambulatory, the child would take only a few steps on tip-toe, legs wide-apart and had stopped running. There were no other neurological or musculoskeletal symptoms. Episodes of urinary urgency and marked Urethral Dysuria were followed by a failure to void and screaming for 15 minutes. Once initiated, the Urinary Stream was normal. There was no urinary frequency or macroscopic hematuria. The parents thought that the entire penis had appeared purple at times and he had received treatment for Balanitis on one occasion. Sporadically, he complained of pain in the left scrotum particularly at night. Bowel frequency was alternate daily. There was no history of recent trauma or fevers. However, he had suffered a fall from a slide one

year earlier without apparent sequelae. There was no history of Allergy or Atopy.

On examination, he was apyrexial. Height was on the 75th centile, weight above the 9th and head circumference on the 25th centile. Musculoskeletal, neurological and abdominal examinations were normal. In particular, a full range of movement was obtained for all joints and no overlying erythema or tenderness present. The Spine also was normal to inspection and palpation: there were no stigmata of spina bifida occulta. Examination of the perineum revealed a normal penis with a physiological phimosis and descended testes in a normal non-tender scrotum.

Urine dipstick detected Protein ++, Red Blood Cells + and White Blood Cells + but no nitrites. Urine Culture repeatedly showed no organisms.

Ultrasound scan of the Urinary Tract was normal. The bladder, although sub optimally distended, showed no focal bladder wall abnormality. The Right Kidney measured 6.9 cm and Left 6.5 cm in bipolar axis (50th centile for age = 7 cm), both demonstrating normal internal echoes without any focal parenchymal lesions or Hydronephrosis. Despite waiting for 1.5 hours, the patient did not void. The Epididymis was reported to have been tender during the Ultrasound examination.

Magnetic Resonance Imaging (MRI) scan under General Anesthetic of the Cervical, Thoracic and Lumbo-sacral Spine was normal, including the Cauda Equina with cord termination at the upper third of L2.

Having excluded neurological, orthopaedic and congenital uropathy causes, idiopathic urethral inflammation was considered as possible differential diagnosis.

General advice was given on regular toileting, an adequate fluid intake and avoidance of "bladder irritants", in particular concentrated urine, caffeinated or fizzy drinks, citrus/orange squash and blackcurrant drinks. Hydrocortisone 1% cream was prescribed topically for the physiological phimosis in view of the episodes of penile discoloration.

When the family ran out of double concentrate orange squash all symptoms resolved. Subsequent food challenge of double concentrate orange squash resulted in prompt recurrence of the symptoms. Fresh orange juice did not produce any of the symptoms.

Having consistently removed double concentrate orange squash from the diet for the last 10 months, he has remained entirely symptom-free. The parents have declined referral to a specialist allergy clinic and further allergy testing.

Discussion

To the best of our knowledge, this is the first case of Acute Severe Juvenile Allergic Urethritis caused by double concentrate orange squash. Food challenge enabled definitive determination of the cause and hence treatment. Consistent avoidance has resulted in cure. double concentrate orange squash consists of water, orange fruit from concentrate (20%), sugar, citric acid, sweeteners (aspartame, saccharin), natural flavoring, an acidity regulator (sodium citrate), preservatives (potassium sorbate, sodium metabisulphite), stabilizer (cellulose gum), natural color (carotenes) and phenylalanine. Further testing of the child would be needed to ascertain the exact antigen.

Non-specific Urethritis in children typically occurs in adolescence, as a mild, self-limiting disease[3]. Putative underlying mechanisms include infection[4], inflammation[5], chemical irritation or allergy[6] and dysfunctional voiding[7]. An allergic basis to Urethritis was first proposed by Weston[8] in 1965 but was deemed an unlikely cause of Urethritis in a series of 17 boys aged 5 to 15 years published by DI Williams[4]. Food challenge conclusively proved the allergic aetiology in our case. Additionally, urethral inflammation may have been exacerbated by disturbed voiding. The patient had become dry by day only a month earlier. Bladder control is usually associated with resolution of detrusor-sphincter-dyssynergia of infancy[9]. However, the child suffered urgency, hesitancy and would delay voiding for as long as possible during these episodes, at times voiding only twice a day.

The severity of the accompanying symptoms was dramatic. We assume that the abnormal wide-based Gait related to an attempt to minimize movement-exacerbated urethral and perineal pain. Joint examinations were normal and Serum Inflammatory Markers were within the normal range. The intermittent left scrotal pain and epididymal tenderness observed during the Ultrasound examination in our case may have related to the allergic/inflammatory process extending beyond the confines of the Urethra to the adjoining tissues, or to urethro-vasal reflux secondary to detrusor-sphincter-dyssynergia precipitated by the urethral inflammation and avoidance of voiding. Ninan, *et al* demonstrated Urethro-Vasal Reflux (UVR) on Micturating Cystourethrography (MCUG) in a case of idiopathic adolescent posterior urethritis leading to recurrent left Epididymitis[5]. In view of the normal Ultrasound of the urinary tract and resolution of symptoms on avoidance of the allergen, we did not perform a MCUG or Cystourethroscopy.

Children with allergic diseases require holistic management rather than a fragmented systems-based approach. Rapid diagnosis was facilitated for our patient by appropriate referral by General Practitioner, joint assessment and close collaboration between paediatricians (General, Neurology and Immunology) and Paediatric Surgeons and Urologists. Food challenge enabled a definitive diagnosis and prompt cure of severe Allergic Urethritis in this 3-year-old boy by avoidance of double concentrate orange squash.

Since our index case of Juvenile Allergic Urethritis was seen and treated, few more publications have appeared with the use of topical steroids, antihistamines, instillation of water-soluble contrast, topical steroids and local anesthetic medications cocktails into the Urethra

and Bladder and even systemic steroids have been found useful[10-13]. Recently, Allergic Disorders in the Genitourinary Tract and Secreto-motility Disorders as well as histamine intolerance in children has been considered additional factors to be considered in such cases[14-15].

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Contributions

All authors contributed to the study conception and design. Material preparation, data collection and analysis were performed by all authors. The first draft of the manuscript was written by RP and all authors commented on or edited previous versions of the manuscript. All authors read and approved the final manuscript.

Ethics Declarations

Conflict of Interest : Authors Anthony, Govani Patel and Patel declare that they have no conflict of interest.

Ethical Approval : All procedures performed in studies involving human participants were in accordance with the ethical standards of the Institutional and/or National Research Committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed Consent : Informed consent was obtained from all individual participants included in the study.

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