

PP-90**Not Quite Hickam's Dictum**

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INTRODUCTION

Myasthenia gravis is a rare autoimmune neuromuscular disease. Rarer still is the combination of myasthenia gravis and Graves' disease, especially so in the paediatrics population, bringing to mind Hickam's Dictum. However, considering that both diseases are antibody-mediated, it should not be surprising to find them occurring simultaneously. Of more concern, though, is the effect these diseases have on each other's natural progression, and the challenges in managing them. Indeed, it has been observed that concurrent myasthenia gravis worsens hyperthyroidism, and improved thyroid function worsens muscle weakness.

CASE

We present a 3-year-old girl with concurrent exophthalmos and ptosis, diagnosed to have ocular myasthenia gravis and Graves' disease. We discuss the course of her disease and challenges in managing it. The course of her disease was as reported in adults, where her hyperthyroidism was difficult to control, and her ophthalmoplegia worsened when her hyperthyroidism improved.

CONCLUSION

We are reminded that the presence of exophthalmos and ptosis simultaneously should ring warning bells of concurrent myasthenia gravis and Graves' disease.

PP-91**Dilemma in Gender Assignment in Vanishing Testis Syndrome: Report of Two Cases**

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INTRODUCTION

Vanishing testis syndrome, or Testicular regression syndrome (TRS) is a developmental anomaly characterized by the absence of one or both testicles with partial or complete absence of testicular tissues. Vanishing testis syndrome may vary from normal male with unilateral non-palpable testis through phenotypic male with micropenis, to phenotypic female. The phenotype depends on the extent and timing of the intrauterine accident in relation to sexual development.

Here we present 2 cases of vanishing testis syndrome with different gender assignment.

CASE 1

10-month-old baby with chromosome 46XY presented with genital ambiguity with phallus-like structure, labioscrotal fold and non-palpable gonads. Follicular stimulating hormone (FSH) and luteinizing hormone (LH) level suggest of gonadal failure. Beta human chorionic gonadotropin stimulation test shows poor testosterone response. Laparoscopic exploration revealed bilateral spermatic cord and vessels with atrophic testes. In view of severe undervirilised phenotype, parents registered child as a female.

CASE 2

12-year-old child already registered as a boy, was referred for bilateral undescended testes and micropenis. Chromosome is 46XY. Laparoscopic exploration revealed atrophic testicular tissues at the end of spermatic cord. Histopathological examination shows structure resemble vas deferens and benign adrenal rest tissues. The child was given testosterone replacement during pubertal age.

CONCLUSION

Diagnosis of vanishing testis is based on clinically non-palpable testis with a blind-ending spermatic cord. It has a typical clinical and histopathological characteristic, but with a wide spectrum of clinical presentation and phenotype. The degree of masculinization depends on the duration of testicular function prior to its loss. Thus, gender assignment needs careful consideration and involves extensive discussion among the parents and multidisciplinary teams. Long-term outcome of clinical status, gender identity and sexual orientation need to be monitored.