A rare presentation of true hermaphroditism: an abnormal inguinoscrotal mass

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Abstract

This study examined the differential diagnosis of inguinoscrotal masses and true hermaphroditism as well as incarcerated inguinal hernia in the differential diagnosis of abnormal inguinoscrotal masses. Inguinoscrotal masses are a rare presentation of true hermaphroditism. A child with an inguinoscrotal mass of the right groin and penoscrotal hypospadias was diagnosed. Our case suggested that abnormal inguinoscrotal masses occur in true hermaphroditism as well as incarcerated inguinal hernia. These congenital anomalies should be considered in the differential diagnosis of inguinoscrotal masses of the groin.

CASE REPORT

INTRODUCTION

True hermaphroditism (TH), due to intersex anomalies (IAs), is a rare anomaly in children. The incidence of IAs is 1.7%, but there is nothing in the literature about the incidence of TH (Coran & Polley, 1991; Glick & Boulanger, 2006). TH is the least common form of IAs (Coran & Polley, 1991). Most children with TH have ovaries and/or testes that grew well (Coran & Polley, 1991). The diagnosis of this rare disorder is clinically and radiographically difficult because it can produce the same symptoms and present similar radiographic findings as incarcerated inguinal hernia. Identifying gender in a newborn with ambiguous genitalia (AG) is emergent and important (Coran & Polley, 1991; Griffin & Wilson, 1992). In the present study, to elucidate TH and to investigate the association between TH and IM, we evaluated a case of inguinoscrotal mass (IM).

Abbreviations

TH	- true hermaphroditism
IAs	- intersex anomalies
AG	- ambiguous genitalia
IM	- inguinoscrotal mass
PH	- penoscrotal hypospadias

CASE

A 4-year-old child was presented with an IM in the right groin and scrotum. On examination, a mass, which was hyperemic, tender, mobile, and measuring approximately 4–5 cm in diameter, was identified in the right groin and scrotum associated with penoscrotal hypospadias (PH) (Figure 1). The patient was operated on under emergent conditions for incarcerated inguinal hernia (IIH). During the operation, it was observed that there were an abscess, two testes, two ovaries, and a uterine tube in the right inguinal exploration. Giemsa Trypsin Banding (GTB) showed a 46 XY karyotype and fluorescent in situ hybridization (FISH) showed that the SRY (sex-determining region Y gene) was positive. The testosterone level, which was appropriate for his age, was determined in his blood after the human chorionic gonadotropin stimulation test. Ten months after the first operation, the rudimentary uterus was resected. Two years after the first operation, the PH was repaired by preputial transverse island flap after parenteral testosterone treatment once a month for 3 months (Figure 2). Two years after the surgical procedures, the patient had no urinary symptoms and no sign of an inguinoscrotal mass.

DISCUSSION

TH is rare in IAs. However, it may also appear as an underlying etiological factor that is responsible for a wide spectrum of IAs. While the forms of IAs can vary, TH is the least common (Dessouky, 2003). However, the anatomical interpretation of TH and IM is unclear (Dessouky, 2003) and it remains controversial whether the primary lesion is TH or IIH. IIH can cause intestinal obstruction (Glick & Boulanger, 2006).

The diagnosis of TH is generally made by physical examination, and biochemical, radiological, and genetic investigations (Sax, 2002). At first we suspected IIH. Later, the diagnosis was confirmed by histopathologic examination (Sax, 2002). A similar clinical presentation between IIH and TH has been identified.

While various forms of AG are present, such as female pseudohermaphroditism, male pseudohermaphroditism, and mixed gonadal dysgenesis, there are few reports of TH occurring with IIH presentation (Dessouky, 2003; Bidarkar & Hutson, 2005)

Two of 9 patients with true hermaphroditism were identified as female and clitoral resection and vaginoplasty were carried out (Coran & Polley, 1991). Six patients with true hermaphroditism associated with penoscrotal hypospadias and bilateral undescended testes were brought up as males and the reconstruction of hypospadias together with bilateral orchidopexy were performed (Coran & Polley, 1991).

In our case, the preoperative physical examination demonstrated hyperemic and tender IM in the right groin. In general, children with IM are evaluated with regard to incarcerated hernia, hydrocele, hydrocele of cord, and undescended testis (Glick & Boulanger, 2006).

In conclusion, our case indicated that TH should be considered in the differential diagnosis of IM. In order to accurately assign gender in a newborn with abnormal external genitalia, a team consisting of a pediatric endocrinologist, pediatric surgeon, psychologist, and radiologist is necessary. Later, reconstructive surgery should be performed (Glick & Boulanger, 2006).

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Figure 1. The appearance of the right inguinoscrotal mass.

Figure 2. The appearance of the external genitalia after the repair of penoscrotal hypospadias.