CASE PRESENTATION

Prolapsing Introital Masses in Two Young Sisters: Diagnosis and Management

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ABSTRACT

The diagnosis and management of two young Saudi sisters who presented with protruding masses in the introitus are described. The younger sister, aged three years, had Rhabdomyosarcoma of the urinary bladder, while the older sister, aged nine years, had urethral prolapse. Both conditions are rare. It was challenging and interesting to manage two sisters with similar symptoms, but totally different underlying diseases. Both were managed successfully.

A prolapsing mass from the introitus in a young girl is either a prolapsed ureterocele, urethral prolapse, condyloma or a Rhabdomyosarcoma of the bladder, vaginal or cervical origin. All these conditions are uncommon.

However, the associated urinary symptoms and spotting of blood should not deceive managing surgeons and paediatricians, since these symptoms could easily be attributed to the more commonly seen inflammatory conditions.

The diagnosis of a prolapsing mass at the introitus of a young girl requires careful history taking, physical examination and thorough judicious investigations. The management of such conditions requires close collaboration between the members of the managing team.

THE CASES

The younger sister (NR) aged three years, presented first with pain and difficulty during micturition. Her mother noticed a mass protruding from the child's introitus. The child had a past history of recurrent attacks of urinary tract infections over a few weeks prior to presentation. History of fever, nausea, vomiting and loss of weight was also obtained. Physical examination was unremarkable, except for a redness of the introitus. The urine culture revealed a significant number of E. Coli. Haemoglobin, renal and liver function tests were normal.

Intravenous pyelography revealed two normally functioning kidneys, but the cystogram phase and

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the voiding cystourethrogram showed a large irregular filling defect, involving the base of the urinary bladder (Fig 1).

Figure 1

Cystogram with basal and right-sided filling defects.

Sonographic and CT scanning (Fig 2) of the urinary bladder confirmed the presence of a solid intravesicular tumor. During cystoscopy, the classical "bunch of grape" – like tumor was seen along the base and right side of the bladder; a biopsy was taken which revealed a histopathological picture of embryonal Rhabdomyosarcoma (sarcoma botryoides). Vaginoscopy, using a paediatric cystoscope through the hymenal ring without disrupting it, revealed a polypoid mass pushing against the anterior vaginal wall, without invasion of the mucosa, and a normal uterine cervix. The metastatic survey was negative. The tumor was staged clinically as T_{21}, No, Mo.

The child was managed initially with antibiotics and pre-operatively with two cycles of combination triple chemotherapy of pulsed Vincristine, Actinomycin D and Cyclophosphamide (VAC) for a period of eight weeks. Re-evaluation revealed significant reduction in the tumor size. Re-evaluation at 16 weeks of chemotherapy revealed a minimal residual tumor on one side of the urinary bladder and partial cystectomy was then performed. The child made an uneventful recovery. Although there was no evidence of viable tumor cells in the histopathological examination of the surgical specimen, it was felt that it was best to give post-operative radiotherapy due to possible vaginal wall involvement on subsequent CT scans. This was completed and the child was continued on triple agent chemotherapy (VAC). At 12 months post-operatively, the child remained well; she was gaining weight and growing, and there was no evidence of recurrent tumor.

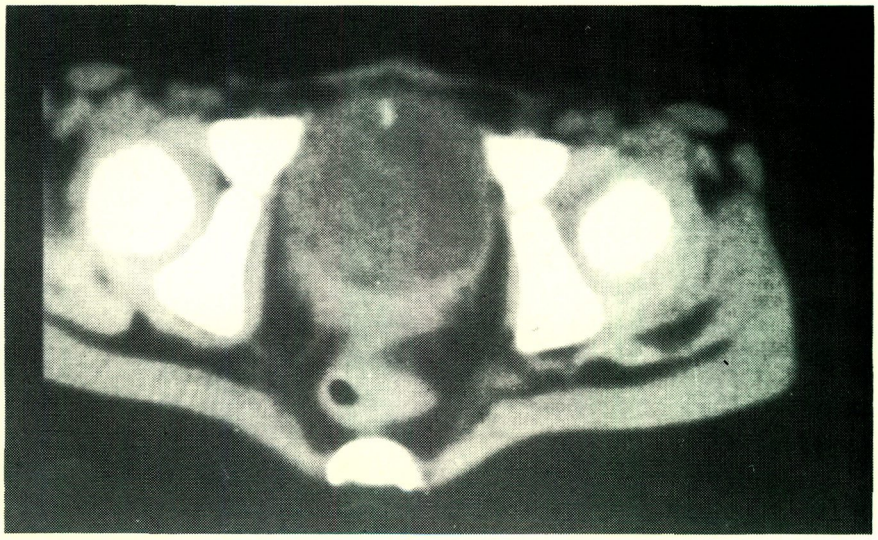

Figure 2

CT scan of the pelvis showing the solid intravesical tumor.

During the course of hospitalization of the younger sister, the older one (AR) aged nine years, presented with spotting of blood, dysuria and a protruding mass from the introitus. She was previously healthy. These symptoms alarmed everyone, knowing the younger sister's condition. However,

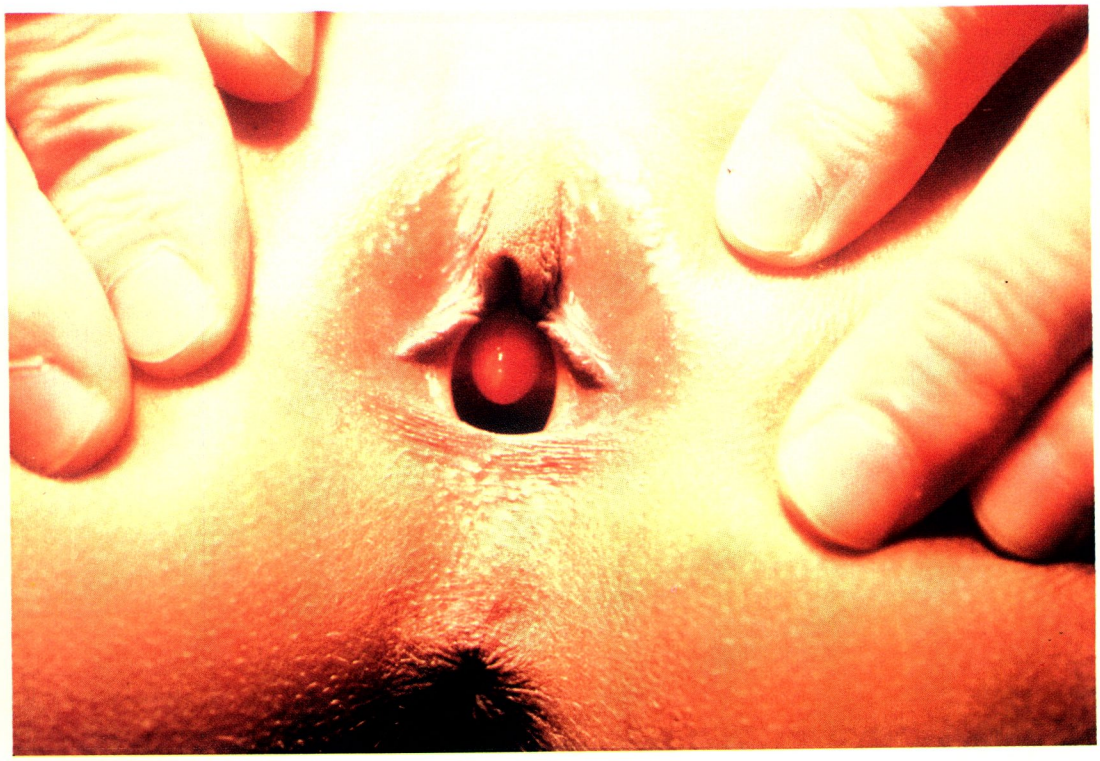

Figure 3. Urethral prolapse, a distinctive red mass projecting from the introitus.

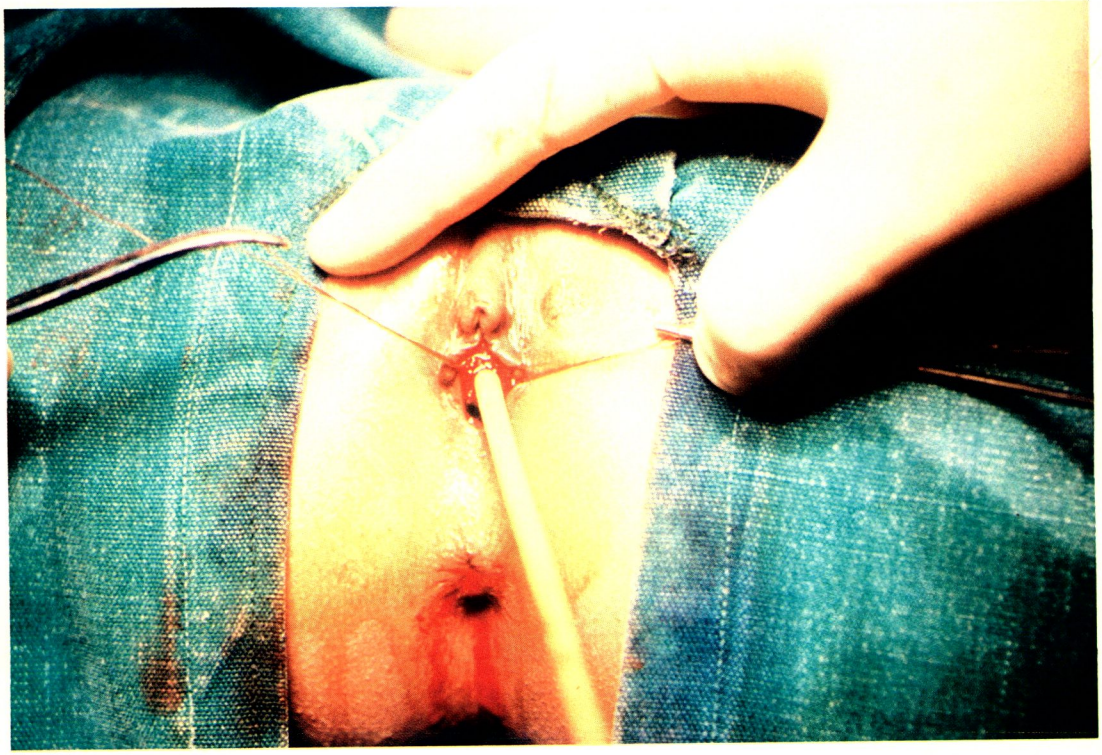

Figure 4. Urethra with Foley catheter in place after excision of the prolapse.

physical examination revealed a circumferentially distinct red mass protruding from the urethra. The remaining examination was normal. Her urine was clear of red blood cells and the culture was sterile. Bladder sonography and voiding cystourethrograms were also normal (Fig 3). Cystoscopy was negative. Vaginoscopy, using a paediatric cystoscopy through the hymenal ring without disrupting it, revealed a normal vagina and uterine cervix.

The child was managed by sharply excising the prolapsed portion of the urethra (Fig 4). Forty eight hours post-operatively, Foley catheter was removed and the child made an uneventful recovery. During follow-up in the outpatient clinic, the child remained asymptomatic and was voiding without any difficulty.

DISCUSSION

The symptom of a protruding introital mass in a young girl deserves careful evaluation, since it may represent a serious problem, such as malignancy of the genito-urinary tract[1]. Rhabdomyosarcoma of the genito-urinary tract constitutes 15-30% of all Rhabdomyosarcomas, which represents the most common solid tumor in childhood[1-4]. Rhab-domyosarcoma in childhood is an aggressive malignant tumor arising from undifferentiated mesenchyme[2]. Although the term "Rhabdomyosarcoma" implies an origin from the skeletal muscle, the tumor in fact arises from immature mesenchymal cells which exhibit varying degrees of differentiation towards skeletal muscles, smooth muscles and connective tissues[2].

Conventional radiographic investigations are valued in the diagnosis of genito-urinary Rhabdomyosarcoma, but ultrasonography and computerized tomograph are useful in defining the characteristics of a pelvic mass, determining its extent, resectability, as well as in the follow-up of the case[2].

The annual estimated incidence of genito-urinary Rhabdomyosarcoma is between 0.5 to 0.7 case per million children younger than 15 years[3]. Because of its rarity and the difficulties in evaluating the results of management of the small numbers seen in single centers, an intergroup Rhabdomyosarcoma study (IRS) was activated in the United States of America in 1972. Various treatment protocols were designed (IRS I - IRS III), incorporating several modalities and considering the cytohistoligic classification which divides the tumor into favourable and unfavourable histology[3].

Traditionally, Rhabdomyosarcoma has been divided into palermorphic, alveolar, embryonal and botryoides types[3]. The philosophy regarding treatment of pelvic Rhabdomyosarcoma has evolved slowly over the past few years, changing from radical ablative surgery towards more conservative surgery, preceded and followed by multi-agent cyclic chemotherapy and radiotherapy, when necessary[2,4,5].

We elected to follow the regime described by Kaplan et al[4] and reported by Kramer[3] in the management of this patient (IRS II).

Urethral prolapse was first reported by Keefe[5]. Urethral prolapse is most common in black females and presents with what is often misinterpreted as vaginal bleeding[6]. To our knowledge, this is the first report of a white Saudi female with urethral prolapse. The true incidence of urethral prolapse in females in general is unknown, but is has been estimated that approximately 1 in 2,500 to 3,000 paediatric admissions are for urethral prolapse with a marked predilection for black girls. Since this seems to be the first Saudi case to be reported in the medical literature, it is either an extremely rare condition among Saudi girls, or it is an overlooked diagnosis, for which this paper serves as a reminder.

The cause for urethral prolapse is unknown and the prolapsing red mass of urethral mucosa should be distinguished from more serious lesions, including trauma, prolapsing ureterocele, condyloma or Rhabdomyosarcoma[7].

Urethral prolapse has been reported in identical twins, and observed since birth, whereby both congenital and hereditary factors were inplicated[8].

Conservative management includes application of topical antimicrobial ointment[9], or application of topical estrogen cream, based on the theory that estrogen deficiency plays a role in the aetiology of this condition. However, we find that frequent topical applications of creams and ointments to the genitalia of young girls is unacceptable in our society.

Although conservative management using the application of topical antimicrobial ointment has

been advocated by some authors[9,10], the recommended treatment is to sharply excise the redundant mucosa around an indwelling catheter and to approximate the edges of the urethra with absorbable suture[6,11]. This latter method has been reported to have the lowest complication rate and shortest hospital stay[7,11].

CONCLUSION

The problem of a prolapsing mass in the intriotus of young females has been addressed and two representative cases, which happened to be sisters, were reported in this paper. Diagnosis was made and both sisters were managed successfully.

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