An Interesting Case Of Atrial Septal Defect, Patent Urachus And Valvular Anus Presenting In Adulthood

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Abstract

Patent urachus presenting in adulthood is extremely rare. Majority of these would have sought medical attention in their infancy or early childhood. This report describes a female patient with weeping umbilicus brought by a social worker. There were additional anomalies, namely: anorectal malformation (ARM) and atrial septal defect (ASD). This is a rarity of a trinity of genitourinary, anorectal and cardiac anomalies.

Introduction

The urachus is a fibrous cord located in the extraperitoneal tissues of the anterior abdominal wall. When it fails to get obliterated, four distinct types of anomalies arise. In the order of frequency, they are 1) patent urachus (50%), 2) urachal cyst (30%), 3) umbilical urachal sinus (15%), and 4) vesicourachal diverticulum (3%-5%) (1, 2). ARM occurs more frequently in boys than girls. The sex ratio varies from 55% to 70% in favor of boys (3, 4). The cardiac anomaly is associated most commonly with anorectal malformation but not with patent urachus. But, combination these three congenital anomalies are extremely rare.

Case Report(s)

The patient was a middle aged female brought to us by a social worker with history of long standing weeping umbilicus. She was well preserved with a normal mental status. There was no fecal incontinence. She complained of chest pain occasionally. The most conspicuous point in the history was absence of repeated episodes of urinary tract infection in the past. She had normal menstrual cycles and had not conceived after 15 yrs of marriage.

Examination showed a leaking opening in the lower lip of a low placed umbilicus. There was peri-umbilical ammonical dermatitis.

Genital examination showed the urethral, vaginal and anal openings were within the vestibule. 2D ECHO of heart showed atrial septal defect measuring 39 mm in diameter with right to left shunt. Ultrasound of the abdomen revealed a small uterus and a small sized right kidney. Urinalysis showed 5-7 WBC/HPF. Hemoglobin was 10 Gms/dl. Blood Urea was 40 mg/dl and Serum creatinine was 1.3 mg/dl. No organism was grown in urine cultured.

A short channel connecting the dome of the bladder with an external opening close to the umbilicus was noted at Cystoscopy. Ureteral orifices were orthotopic. This tubular connection was then excised into to with a cuff of bladder and the defect in bladder was repaired. The wound healed well and she could void well without incontinence or discomfort. Histopathology confirmed that the excised tract was urachus. No intervention was done for ARM and ASD as she was not willing for the same.

Discussion

A valvular anus as in this case results because the anus lies within the vestibule as a result of very little development of perineum. In this anomaly, the anal orifice is usually small but not stenotic and is surrounded by wet vestibular epithelium.

This patient had a low translevator ARM in which all the 3 openings were within the vestibule i.e. urethral, vaginal and the anus. Patent urachus is explained by non-descent of the bladder or, more commonly, failure of the epithelial-lined urachal canal to obliterate (5).

Bladder obstruction during fetal development has been blamed for the urachus remaining patent. It is seen that urachal patency is often absent in severely obstructed bladders in utero as in a case of posterior urethral valves. So the obliteration of the urachus may be independent from the level of bladder distention (6, 7). In our case the bladder was well within the pelvis with a normal outlet and orthotopic ureteric orifices.

Therefore re-tubularization, rather than primary patency, might be the cause for urinary drainage from the umbilicus (8, 9).

Delayed presentation and absence of repeated urinary tract infections despite the fact that the
urethra, vagina and anus being within the vestibule facilitating colonization by bacteria are unique features in this report. Combination of cardiac, urinary and anorectal anomalies makes it extremely rare.

References

Illustrations

Illustration 1

Clinical Photographs

Fig 1:- Low placed umbilicus
Fig 3:- All the 3 openings within the vestibule
Fig 5: Chest X ray showing increased pulmonary vascularity with prominent hilar markings and cardiomegaly.

Fig 6: Post op picture after repair of umbilicus.
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