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Bladder Exstrophy: A rare case report

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Abstract: *The bladder exstrophy (BE) represents an anterior midline defect with variable expression comprising a spectrum of anomalies involving the abdominal wall, pelvis, urinary tract, genitalia, and occasionally the spine and anus. The vast majority of BE cases are classified as non-syndromic and the etiology of this malformation is still unknown. We are presenting a case report on BE. A 11 year old patient presented with continuous dribbling of the urine from the lateral side of a soft tissue mass present in the lower abdominal wall with inability to hold and pass urine. The lesion was present since birth. On examination a soft tissue mass was present in lower abdominal wall with two opening on the either sides. Penis was short with unremarkable scrotum. Testis was not palpated in the scrotum. Combined investigation - IVP, CECT and USG revealed pubic diastasis with externally placed bladder with deficient anterior bladder wall, bilateral ectopic ureteral opening and chronic cystitis with undescendant testis.*

Keywords: *Bladder exstrophy, Cystitis, Pubic diastases, Epispadias.*

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I. Introduction

Bladder exstrophy is a rare congenital birth defect occurring about 3 in 100, 000 births with males to female ratio of 3:1 & includes malformation of the bladder and urethra in which the bladder is turned inside out, flattened and exposed to outside the body & bladder neck fails to form and the anus and vagina appear anteriorly displaced [1]. Also, there is diastasis of the pubic bones.

II. Case report

A 12 year old male came to the radiodiagnosis department with swelling in lower abdomen & difficulty in passing urine since very long time. On elaborating the history the parents of the child told that the condition was from birth. His child never micturitated, rather the urine came out via the lateral margin of the protruded mass. There was no history of past surgical intervention.

On examination a defect noted in the lower anterior abdominal wall with protrusion of a globular soft tissue swelling from the defect. Confluent with this swelling, in the inferior aspect, were two openings, from where urine dribbled continuously, suggesting ureteral openings, with exposed posterior wall of the bladder. Umbilicus was not seen separately.

Due to continuous dribbling of urine, mild mucosal abrasion and scarring of the anterior surface of the protruded mass was seen. Penis was short, stubby, curved upwards and is drawn into the exstrophy area, suggestive of dorsal chordee. Bilateral testis was not palpated in the scrotum.

Figure – 1

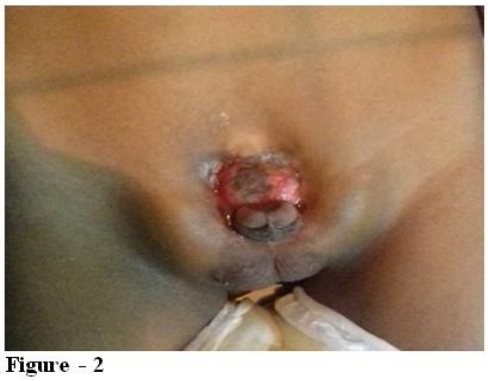


Figure - 2

Plain Picture (pp) KUB and IVP

KUB PP: Radiographs were taken in antero-posterior view [Figure – 3] which revealed a globular soft tissue shadow arising from the anterior aspect of lower abdomen and pelvis. There was evidence of diastasis of pubic bones. Coccyx was absent.

IVP: After contrast administration, there is symmetrical appearance of nephrogram and pyelogram on both sides [Figure – 4]. Position of both kidneys appears normal. Both kidneys shows normal size and smooth contour.There is prompt excretion of contrast into collecting system on both sides. Fornices appear sharp and papillary impressions are well preserved. Pelvis on both sides are normal. No evidence of malrotation seen. Upper 2/3 of both ureters appear normal in position and calibre. However distal end of ureter proximal to vesicoureteral juunction could not be well appreciated. Urinary Bladder: Inadequately distended with contrast mixed urine and shows irregular contour.

Diagnosis was put as exstrophy of bladder with bilateral ectopic ureteral openings and chronic cystitis.

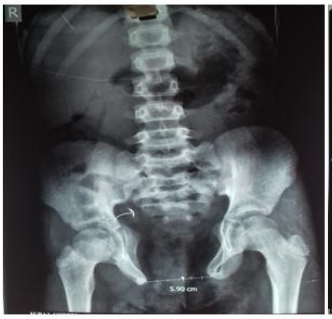


Figure – 3: Plain picture KUB

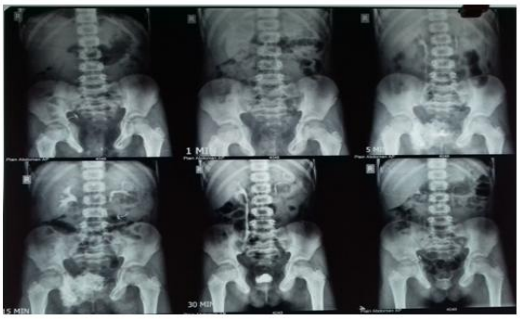
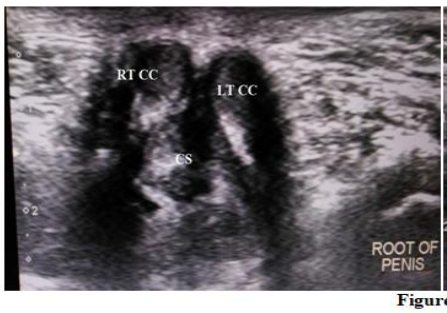


Figure – 4: IVP

USG: On USG both corpora cavernosa (CC) and corpora spongiosa (CS) was normally visualized at the root of penis (Figure - 5). However there was hypospadias and significant dorsal chordee. Penis was underdeveloped with short penile shaft. Bilateral testis (TT) was not visualized in scrotum; it was present in the inguinal canal (Figure - 6), suggestive of undescendent testis. Bilateral lower abdominal wall was underdeveloped as evidenced by diverication of lower rectus muscle (Figure - 7). Anterior wall of the bladder was not developed (Figure - 8) with direct visualization of its posterior wall with reduced bladder capacity.



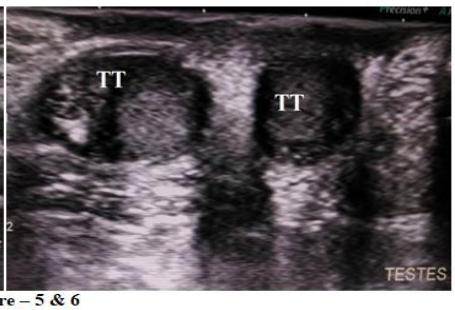
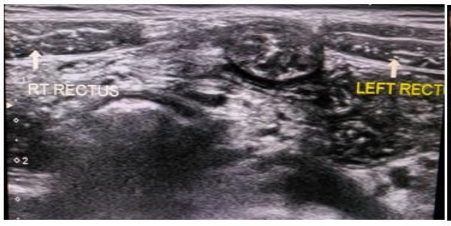


Figure – 5 & 6



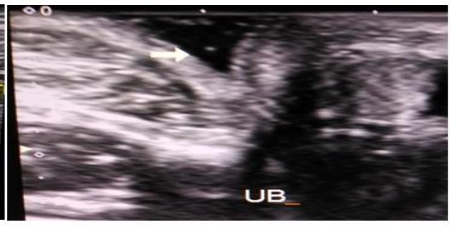


Figure –7 & 8.

III. Cect Kub Findings

There was marked widening of the symphysis pubis with absence of normal urinary bladder (figure 9 & 10). Focal defect seen in anterior abdominal wall near midline in suprapubic region with protrusion of the posterior wall of bladder with overlying thickening of the soft tissue planes.

Right and left kidneys measure 7.6 (CC) x 3.6 (AP) x 3.1 (TR) cm and 7.8 (CC) x 3.5 (AP) x 3.2 (TR) cm respectively. Both kidneys reveal normal size, shape, position and attenuation. Normal functioning and excreting bilateral kidneys. Bilateral upper 2/3 of ureters were normal in location, contour, size & enhancement without any hydronephrosis. Delayed phase shows normal contrast excretion in both kidneys & ureters. Distal

1/3 of both ureters shows mild dilatation with acute anteromedial course in lateral pelvis reaching up to the abdominal wall defect in suprapubic region with further short segment of narrowing of caliber of ureters coursing anteriorly near midline upto the superior margin of base of penis simulating epispadias. Bilateral mucosal thickening noted in the region of vesicoureteric junction. Penile shaft is blunted & smaller in size. Scrotum is unremarkable with presence of testes in the inguinal canal.

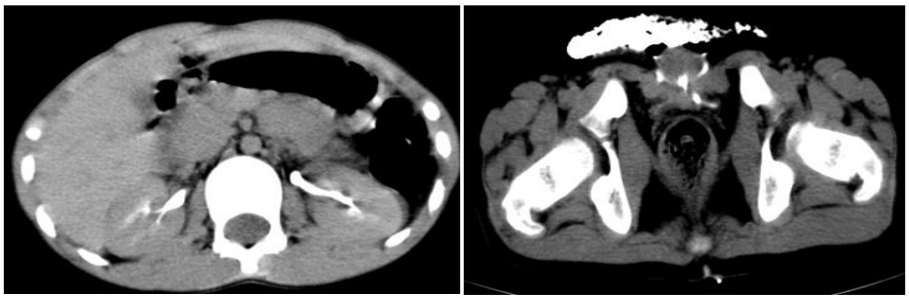


Figure –9 & 10.

IV. Discussion

Bladder exstrophy (also known as **ectopia vesicae**) refers to a herniation of the urinary bladder through an anterior abdominal wall defect. The severity of these defects is widely variable. The condition is thought to be caused by incomplete development of the infra- umbilical part of the anterior abdominal wall, associated with incomplete development of the anterior wall of the bladder owing to delayed rupture of the cloacal membrane. Persistence of the cloacal membrane prevents medial mesenchymal ingrowth, causing the abdominal wall to remain lateral and the posterior bladder wall to be exposed to the external surface [2, 7]. The anterior abdominal wall defect involves the entire urethra and bladder neck [4].The pubic symphysis is always widened [3] with diastasis of rectus abdominis. Umbilicus is low set [4, 7]. Frequently there is omphalocoele [4, 7] which is confluent with exstrophic bladder. [4] In males the penis is short, stubby, curved upwards and is drawn into the exstrophic area [2, 4]. Unilateral or bilateral cryptorchidism may be present [2, 4]. Inguinal hernia may be associated [2, 4]. In females, the urethra is short, often buried in the exstrophied bladder. The clitoris tends to be bifid. The labia are also widely separated. The vagina is short and orifice may be stenotic. Uterine prolapse or unicornuate uterus may be present [2, 4]. Distal ends of ureters are slightly dilated, and curve laterally, then medially and slightly upwards in the shape of a hook before entering the bladder [4].

V. Conclusion

Bladder exstrophy a rare congenital birth defect can be diagnosed by prenatal USG and necessary steps can be taken out in further management. In postnatal case renal USG, Spinal USG, CT KUB and MRI must be done to exclude renal agenesis, hydronephrosis, myelodysplasias and vertebral anomalies. An early assessment by examination under anaesthesia should be carried out in a center experienced with the condition.

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