A CASE OF BLADDER EXSTROPHY AND A NOVEL THEORY ON ITS NATURAL HISTORY

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Case Report:

This is a 34 years-old patient gravid 2 para 1, at 18 weeks of gestation, with a history of a prior uncomplicated vaginal delivery and healthy daughter. The patient was referred to our hospital because a fetal ultrasound demonstrated a cystic tumor in the anterior part of the fetal abdomen of such dimensions that limited hip flexion bilaterally. The referring diagnosis was omphalomesenteric cyst. We decided to perform a genetic amniocentesis that showed a normal male karyotype. During subsequent follow up, the cystic apperance of the tumor disappeared and turned into a solid mass that resembled a bladder exstrophy. Intra-abdominal cystic images later appeared which added a concomitant intestinal atresia to the differential diagnosis. An elective cesarean section was performed at 39 weeks of gestation and a live born infant with a birth weight of 3100 g and Apgar scores of 8 and 10 was delivered and transferred to the intermediate care unit. Physical examination confirmed the antenatal diagnosis and added a diagnosis of anorectal atresia. Surgical treatment included neonatal colostomy and closure of bladder exstrophy (three weeks later). Once the baby reaches six months of age the plan is to correct the epispadias and perform reversal of the colostomy.
Figures 1-4: 16 weeks of gestation. Top left: Sagittal view of the fetus with a cystic structure in the anterior part of the abdomen. Top right: represents an axial view of the abdomen with a cystic structure, please note the lateral abdominal folds. Images below: Path of the umbilical cord around the cystic bladder.
**Figure 5:** 20 weeks of gestation: the cystic appearance of the mass disappears and is replaced by solid triangular mass in the lower abdomen. Abdomen with umbilical arteries running alongside the mass.

**Figure 6:** 24 weeks of gestation. Bladder exstrophy. Umbilical vessels around the tumor. The penis and scrotum are displayed (Top right).
Figure 7: 37 weeks of gestation. Bladder exstrophy. Multiple intra-abdominal cystic images compatible with intestinal atresia.

Figure 8: Newborn with bladder exstrophy and imperforate anus.
**Figure 9:** Newborn with bladder exstrophy after colostomy. In the right thigh there is part of the scrotal skin. Inguinal hernias are present bilaterally.

**Figure 10:** Sequence of images of corrective surgery with satisfactory results
Comments:

The true cause of bladder extrophy has not been elucidated. In classic bladder extrophy, the lower urinary tract, genitalia, and musculoskeletal system are affected. The prevalence of classic bladder extrophy is 3.3 per 100,000 births (range 1:10,000 – 1: 200,000) (2). Most cases, of this condition with variable expression, occur sporadically, but there are some cases that are inherited in an autosomal dominant fashion (5).

Antenatal ultrasound findings suggestive of extrophy-epispadias complex include the following: failure to visualize the bladder on ultrasonography on repeated examinations, lower abdominal wall mass, low-set umbilical cord, images compatible with intestinal atresia and abnormal genitalia (11).

Neonatal clinical findings include the following: the bladder is open on the lower abdomen, with mucosa fully exposed through a triangular fascial defect. The abdominal wall appears long because of a low-set umbilicus on the upper edge of the bladder plate. The distance between the umbilicus and anus is foreshortened. Rectus muscles diverge distally, attaching to the widely separated pubic bones (8). Indirect inguinal hernias are frequent (>80% of males, >10% of females) due to wide inguinal rings and the lack of an oblique inguinal canal. Divergent rectus muscles remain attached to the pubis. The phallus is short and broad with upward curvature (dorsal chordee). There may be cryptorchidism or abnormalities in the scrotum and in some cases penile agenesis has been reported.

Conclusions

Bladder extrophy is a complex malformation involving the abdominal wall muscles, pelvic bone, uro-genital and intestinal tracts. The sequence of events leading to this malformation remain unknown and this would be the first case that documents the natural history of this anomaly. It’s management is difficult and it leads to serious sequelae (1-13). Umbilical cord cyst as a clue in the prenatal diagnosis of bladder extrophy was first described by Tong et al. (12)
Considering the evolution of this case, it is possible to formulate the hypothesis that the natural history of this congenital anomaly begins with a failure of the closure of the lower abdominal wall, through which emerges a megacystis (bladder ectopia) which subsequently explodes and everts exposing its internal lining in the lower part of the abdominal wall. As a consequence, the anomaly becomes a complex one, where the genitourinary tract, the muscular system and intestine are affected. Thus, a detailed ultrasound evaluation is necessary and requires an experienced operator. As this abnormality has important aesthetic and functional consequences, a multidisciplinary approach is required in order to provide appropriate management and parent counseling (4).

**Figure 11**: Schematic representation of the natural history of bladder exstrophy.
References


11. Nguyen Ha, Eva Racanska, Bladder extrophy. 2010-1-11-15


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