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CASE REPORT

Urorectal Septum Malformation Sequence in A Calf

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Summary

Urorectal Septum Malformation Sequence was described in a 3-day-old, Simmental-Brown Swiss crossbred calf characterized by absence of anal and vaginal openings, ambiguous genitalia, sacral hypoplasia, anuria, colonic atresia and lumbal vertebral scoliosis.

Keywords: *Urorectal septum malformation sequence, Sacral hypoplasia, Anuria, Colonic atresia, Lumbal scoliosis, Calf*

Bir Buzağıda Ürorektal Septum Malformasyon Serisi

Özet

Üç günlük, Simental-İsviçre Esmeri melezi bir buzağıda anal ve vaginal deliklerin bulunmaması, sakral hipoplazi, kuyruk bulunmaması, kolon atrezisi, lumbal omur skoliozisi ve anormal genital organ anomalisiyle belirgin Ürorektal Septum Malformasyon Serisi tanımlandı.

Anahtar sözcükler: *Ürorektal septum malformasyon serisi, Sakrum hipoplazisi, Kuyruksuzluk, Kolon atrezisi, Lumbal skoliozis, Buzağı*

INTRODUCTION

The developmental anomalies may be caused by genetic or environmental factors or combination of both and, in most cases the etiology remains obscured [1]. The occurrence of congenital malformations in food animals has economic importance and might provide material for developing appropriate understanding of the embryologic pathogenesis [2].

URSM (Urorectal Septum Malformation Sequence) in humans is a well-known specific developmental abnormalities originally described in about 25 years ago including the absence of anal, urethral and vaginal openings, ambiguous genitalia and genito-urinary anomalies [1]. Before the introduced the term in human medicine, authors used the terms including 'female pseudohermaphroditism with anorectal malformation, hypoplastic pelvic outlet and persistent cloaca' [2]. The extent of variation in human URSM cases is from complete to partial or milder variant forms characterized by a single openining draining the cloaca might be compatible with life [3].

In veterinary medical literature; the presence and frequency of the URSM in domestic animals was much less documented. To the authors' knowledge, URSM was described in one of the dizygous twin lambs [2], in 6 calves [5-8], in a foal [9] and 40 fetus or aborted pig embryos [10] up to date. Generally, most of the authors in veterinary medicine have used to term of 'persistent cloaca or complex intersex condition' to describe these series of anomalies [5-7], whereas URSM was introduced in veterinary medicine for describing in a lamb with urogenital malformations including abnormal external genitalia imperforate anus, fistulous connection between rectum, bladder and vagina [2].

This study describes the morphologic features of URSM together with colonic atresia, rectum aplasia, anuria, sacral hypoplasia, and lumbar scoliosis in a cross-bred calf.

CASE HISTORY

Three-day-old, Simmental-Brown Swiss crossbred calf was presented to the Animal Hospital of Veterinary Faculty

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of Firat University, with the signs of the absence of perianal opening and tail, and postural abnormality *(Fig.1a)*. There was no record of previous defects and the cow had delivered one normal calf before. External appearance of the calf resembled the male due to presence of phallic structure in normal location for penis *(Fig. 1b)*, however there was neither scrotum nor testes. Urethra was not patent. In radiographical and macroscopical examinations; coccygeal vertebrae were absent, there was scoliosis in 4-5 lumbar vertebrae and the sacrum was moderately hypoplastic.

Upon the owner's request and the situation is not compatible with the life, the calf was sacrificed humanly.

In macroscopic examination; genital and urinary canal was not separated and combined as a cloacal sac measured as12x5x5 cm. The following organs were directly associated with the cloaca: urinary bladder (diameter of 5 cm); urethra (length of 8 cm); and uterus *(Fig. 1c)*. Cloaca, urinary bladder and uterus including the uterine horns and Fallopian tubes were all distended massively by straw-colored fluid accumulation and they considerably filled the pelvic and peritoneal cavity. The total fluid having the characteristics of urine was measured as 2800 ml. There was neither rectum nor anus. The vagina and cervix were also absent.

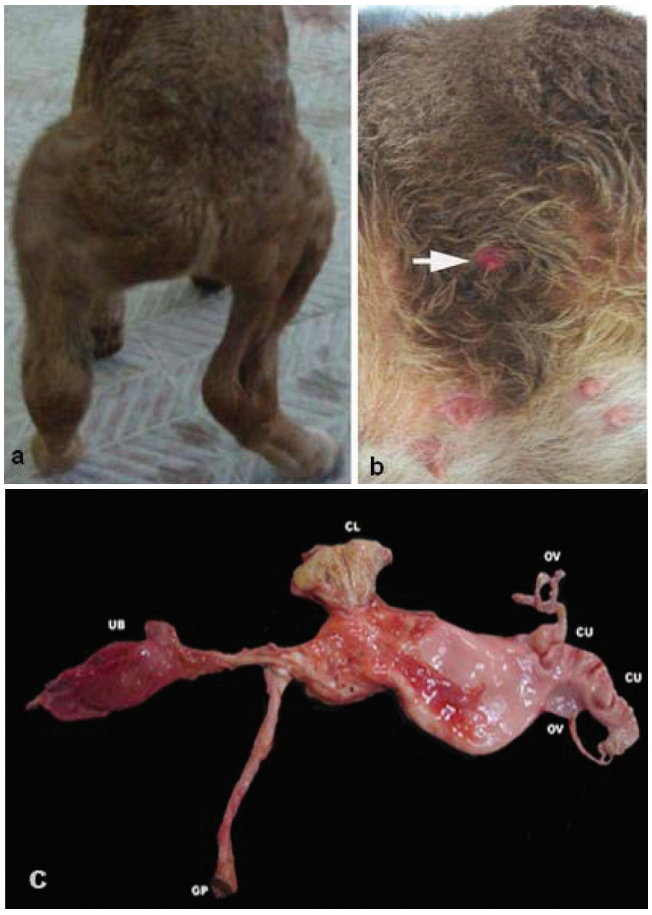

Fig 1. a- Anuria, pelvic hypoplasia and postural abnormality in the calf, **b-** Phallus like structure *(arrow)* and absence of perianal openings, **c-** Dissected organs around the cloaca (CL) including; colon, urinary bladder (UB), cornu uterie (CU), ovaries (O), urether and prepitium (P)

Şekil 1. a- Buzağıda kuyruksuzluk, pelvis hipoplazisi ve duruş bozukluğu, **b-** Penis benzeri yapı *(ok)* ve perianal deliklerin bulunmaması, **c-** Kloaka'yı (CL) içevreleyen dokular; kolon, idrar kesesi (UB), kornu uteri (CU), ovaryumlar (O), üreter ve prepisyum (P)

Colon had no distal portion and ended blindly forming a pouch measuring 2x3x1.5 cm due to meconium accumulation. The urethra was completely atrethic at the point where enter the urinary bladder. Although there was no cervix uteri and vagina; corpus and cornu uteries, oviducts and ovaries were identified. Left cornu uteri was more prominently dilated than the left one *(Fig. 1d)*. The ovaries were in normal size and appearance. No testes, prostate or seminal vesicle was identified.

For histological examination, tissue samples from cloaca, penis, urinary bladder, colon, small intestines, heart, liver, ovary, kidney, and brain were collected and embedded in paraffin wax and then sectioned at 5-6 µm thickness. The prepared sections were stained with haematoxylineosin.

Microscopically; the cloacal wall was lined by a strongly folded, non-keratinized cubiodal epithelium and making villous projections to the lumen *(Fig. 2a)*. It's lamina propria consisted of primitive mesenchymal tissue.

The wall of the urinary bladder consisted of a two to three cells thick transitional epithelium with typical large covering cells having rounded free surface.

The colonic mucosa composed of goblet cells, resorbing epithelial cells and crypts *(Fig. 2b)*. The stroma had highly cellular connective tissue and smooth muscle cells.

The inner surface of the uterus contained thickened regions of the endometrium and the caruncles were present. There was no uterine glands in these areas *(Fig. 2c)*. The mucosa contained single columnar epithelium, lamina propria, and muscularis mucosa. The lamina propria was highly cellular. Myometrium was made up of several layers of smooth muscles, which were demarcated by connective tissue.

The oviducts were lined with simple ciliated cells, cuboidal to columnar in shape *(Fig. 2d)*. The muscularis externa consisted of an inner circular layer of smooth muscles and a less developed outer longitudinal layer. The serosa has a layer of simple squamous epithelium in some connective tissue.

The ovaries covered by single squamous epithelium and tunica albugenea composed of cortex and medulla. Cortex contained numerous primordial, single and multilayered primary follicles *(Fig. 3a)*.

The phallus consisted of corpora cavernous and the corpus spongiosum surrounding the urethra *(Fig. 3b)* and the glans penis covered by prepuce. The corpora cavernous had a thick and tough connective tissue fibers. The sponge-like cavernous tissue consisted of cavernous sinuses and

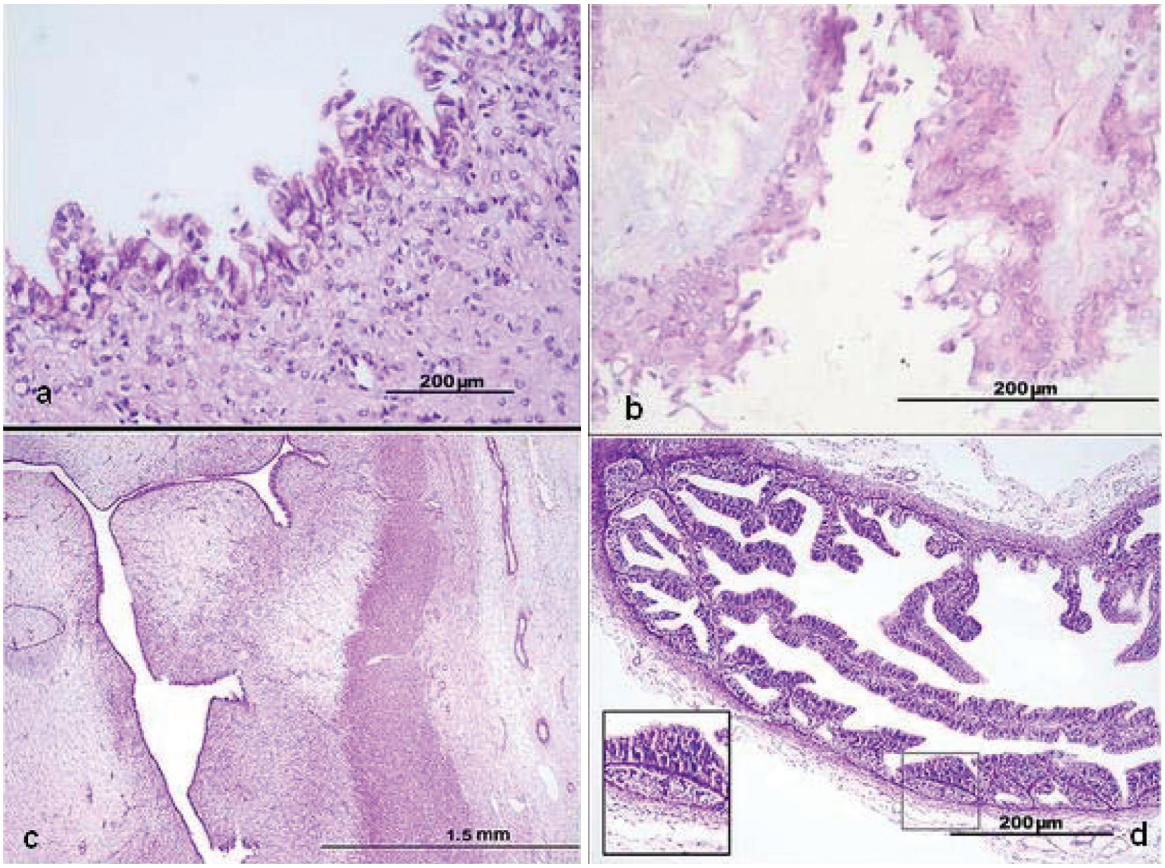

Fig 2. a- Colonic villus composed of goblet and resorptive cells, **b-** Cloacal mucosa containing pseudostatified epithelium and primitive stroma, **c-** The uterine carunculas composed of single columnar epithelial tissue, lamina propria and muscularis mucosa, **d-** The uterine tube lined with simple ciliated cells (inset), cuboidal to columnar in shape, Hematoxylin-eosin (HxE)

Şekil 2. a- Kadeh hücreleri ve rezorbtif hücreler içeren kolon villusu, **b-** Yalancı çokkkatlı epitelyum ve primitif mezenkimden ibaret kloaka mukozası, **c-** Tek katlı kolumnar epitelyum, propria ve musküler tabak içeren üterus karunkulaları, **d-** Silyumlu (inset) kübik-kolumnar epitelyum içeren yumurta yolu, Hematoksilen-eosin (HxE)

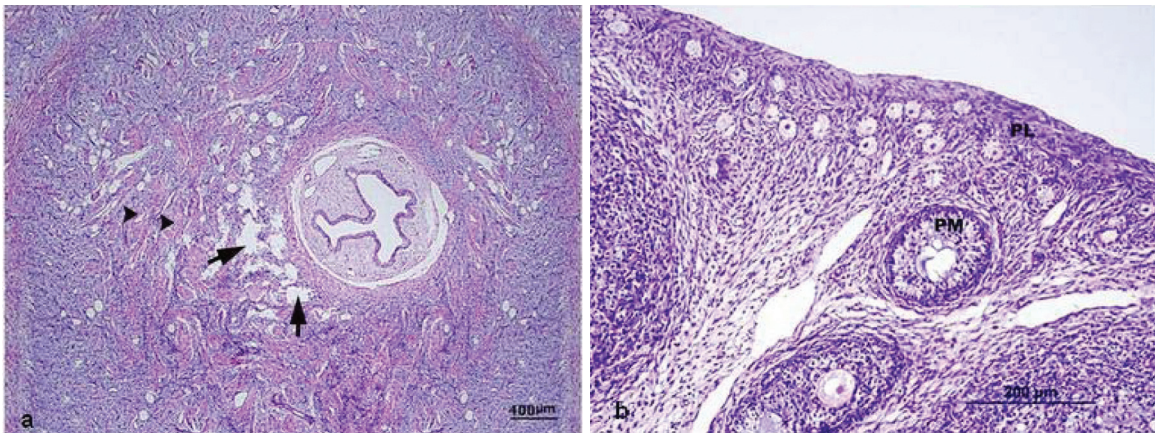

Fig 3. a- The corpora cavernosa *(arrow heads)* and the corpus spongiosum *(arrows)* surrounding the urethra, **b-** The ovarium containing primordial (PL) and primary (PM) follicules, Hematoxylin-eosin (HxE)

Şekil 3. a- Üretrayı çevreleyen korpora kavernoza *(okbaşı)* ve korpus spongiozum *(oklar)*, **b-** Ovaryum'da primordial (PL) ve primer (PM) foliküller, Hematoksilen-eosin (HxE)

septa with smooth muscle fibers. The urethra consisted of a stratified mainly cuboidal urothelium. The lamina propria composed of connective tissue appeared free of glands. The prepuce had stratified keratinized epithelium and showed intraepithelial and subepithelial neutrophilic infiltration.

DISCUSSION

The incidence of URSM is unknown in animals, however, in human neonates, it was reported as one in 250.000 [1]. In this report, we presented URSM sequence in a calf

characterized by absence of anal, urethral and vaginal openings, ambiguous genitalia and genito-urinary anomalies. Additionally, sacral hypoplasie [6,7] anuria and vertebral anomalies [5-7] were described in the present study as in an earlier bovine cases.

Renal changes or defects including horse-shoe kidney [6,8] and renal dysplasia [6], hydronephrosis, hypoplasia or aplasia [5] were commonly reported in bovine and human cases [1], but not such changes were detected in the present report. The absence of this defects in the present case might be explained by the presence of colonic atresia that might protect the obstructive uropathy. Similar to our report, renal defects have not been detected in the lamb [4], in a calf [7], and about 6 of 18 human URSM cases [2].

Originally, it was proposed and widely accepted that the specific sequence is caused by failure of urorectal septum to migrate to and/or fuse with cloacal membrane [1]. However, which factor or factors leading to this failure is unknown. Possible hypothesis including caudal mesodermal deficiency, mutations and teratogens, lateral compression, and vascular steal have been proposed [4].

Similar to the present report, tail abnormalities such as anuria (4 of 6 bovine cases) [5,7] or short and deviated tail (1/6) [6] have been reported co-existently in previous bovine URSM cases. Although anuria seems to be most concurrent lesion in bovine URSM cases, anuria and/or urogenital defects were also co-existently reported in atresia ani in calves [11,12].

Interestingly; amongst the 6 total bovine URSM cases, 3 were Holstein calf [5,7,13], one was Limosin [5], one animal was Simmental [6]. and one was Hereford X Longhorn crossbred calf [8]. The subject of this observation was also Simmental crossbred.

As a result, the case reported here consist of female counterpart of URSM which was reported in two bovine [6,7], and one lamb case and the additional cases are needed to elucidate the mechanism leading to formation of this defects.

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