Persistent mullerian duct syndrome in a dog

Z. Szabo[1], J. Moser[2], S. Vincenti[1]

[1]Abteilung Chirurgie, Departement für Kleintiere, Vetsuisse Fakultät Universität Bern, Schweiz;
[2]Freelance Veterinarian, Schweiz

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Persistierendes Müller-Gang-Syndrom bei einem Hund

Dieser Fallbericht beschreibt einen 14-jährigen, männlichen kastrierten York Shire Terrier, der wegen Strangurie und Tenesmus vorgestellt wurde.

Durch eine Computertomographie (CT) des Abdomens wurde die Verdachtsdiagnose eines persistierendes Müller-Gang-Syndrom gestellt. Während der Laparotomie wurden Strukturen vorgefunden, welche als persistierende Eierstöcke und Gebärmutter vermutet wurden. Der abnormale Uterus war mit Flüssigkeit gefüllt und hatte auf beiden Seiten ein blindes Ende. Die abnormen Strukturen wurden chirurgisch entfernt und pathologisch untersucht. Die Pathologie bestätigte ein persistierendes Müller-Gang-Syndrom. Der York Shire Terrier erholte sich gut von der Operation, konnte am selben Tag Urin absetzen und am nächsten Tag entlassen werden.

Schlüsselwörter: Fallbericht, Computertomographie, Hund, persistierendes Müller-Gang-Syndrom, Strangurie, Tenesmus

Abstract

This case report describes a 14-year-old, male castrated York Shire Terrier, which was presented due to stranguria and tenesmus.

An abdominal computed tomography (CT) scan raised a high suspicion of a persistent mullerian duct. During laparotomy structures were found that were suspected to be a persistent ovary and uterus. The abnormal uterus was filled with fluid and had a blind end on both sides. The abnormal structures were surgically removed and pathologically examined. Pathology confirmed a persistent mullerian duct. The patient recovered well from the surgery and was able to urinate spontaneously the same day and was discharged the next day.

Keywords: Case report, computed tomography, dog, persistent mullerian duct syndrome, stranguria, tenesmus

Introduction

Intersexuality is a condition in which sex determination and sex differentiation does not occur normally. In male pseudohermaphroditism, the animal has XY chromosomes, testes and feminization of internal or external genitals. In the persistent mullerian duct syndrome, fallopian tubes and uterus develop at the same time.[7, 8, 15]

In humans, the disease results from a mutation of the anti-mullerian-hormone produced by Sertoli-cells. The hormone causes regression of the mullerian duct in male fetuses. Alternatively, there may be a mutation in the anti-mullerian-hormone-receptor.[3] It has been shown, that canine persistent mullerian duct syndrome is inherited in an autosomal recessive manner in the miniature schnauzer breed.[11]

The persistence and formation of mullerian duct in male dogs allows the development of uterine horns, cervix and cranial vagina. Testicles-produced testosterones stimulate the development of Wollfian ducts into the epididymis, vasa deferentia, seminal vesicles, and prostate, in close anatomical association with the female tubular reproductive organs. Dogs with a persistent mullerian duct syndrome can develop pyometra, prostate infection and urinary tract infections. Neoplastic degeneration of the interested, abnormal organs can also occur.[7]

Gonadohysterectomy is recommended to reduce the risk of pyometra, urinary tract infection and prostate infection. In addition, affected animals should be removed from the genetic pool, as inheritance cannot be ruled out.[3, 12]

Diseases of sexual development are rare. The unique feature of this case is the appearance of symptoms very late in the patient's life. The aim of this case report was to show how persistent mullerian duct syndrome can present clinical signs in adult-senior animals, how it can be diagnosed and treated.

Case report

A 14-year-old male castrated Yorkshire terrier, with known mitral endocardiosis, tracheal collapse and occasional recurrent cystitis for 2 years, was referred to the emergency department of the Vetsuisse Faculty of the University of Bern in June 2021, due to stranguria and tenesmus.

On clinical examination, the dog was alert, in good general condition and vital signs were within normal limits. Clinical examination revealed bilateral cloudiness of the lenses, a 3/6 heart murmur and tracheal collapse. On abdominal palpation, the urinary bladder appeared to be large, but otherwise the abdomen was not painful.

A blood test showed mild metabolic acidosis and mild hypokalemia. The remaining values were within normal limits.

An abdominal ultrasound was performed, which revealed a fluid-filled tubular structure between the urinary bladder and the descending colon. In addition, bilateral chronic nephropathy with mild pyelectasia, diffuse hepatopathy with nodular changes and jejunal diffuse enteropathy were seen.

Subsequently, a computed tomography (CT) scan was performed. A fluid-filled tubular structure with mass effect on the colon and urinary bladder was seen in the caudal abdomen (figure 1). Other finding included multiple choleliths in the gallbladder, bilateral cystic nephropathy, ventral spondylosis and a protrusion between L7 and S1 with moderate cauda equina compression.

Based on the clinical symptoms, a laparotomy and a thorough abdominal inspection, with particular focus on the caudal abdomen, were performed by an experienced surgeon (figure 2). A large (6 cm × 5 cm) cystic-like structure looking like a malformed uterus with two uterine horns and on the left side with malformed ovarian tissue, was identified. This structure was attached to the serosa of the dorsal aspect of the urinary bladder and the ventral aspect of the rectum. The structure had a blind end on both sides and was filled with fluid, making the structure morphologically resemble a cyst. The remainder of the abdomen was within normal limits for a dog of this age. To avoid any iatrogenic trauma, the urethra was catheterized, and the urinary bladder was gently retracted with the help of a holding suture in its apex (Prolene™ 4–0, Ethicon, Johnson & Johnson, Switzerland). The uterine structures were carefully isolated from the urinary bladder, ureters and the rectum. Double ligatures of the uterine and ovarian arteries were placed (PDS II 4–0, Ethicon, Johnson & Johnson, Switzerland). During the procedure a fusion between the cystic mass and the serosa of the urethra was observed, without a clear communication of the lumen of the urethra with the cystic structure. To avoid any trauma to the urethra, the removal of the cystic structure was performed by sharp transection as close as pos-

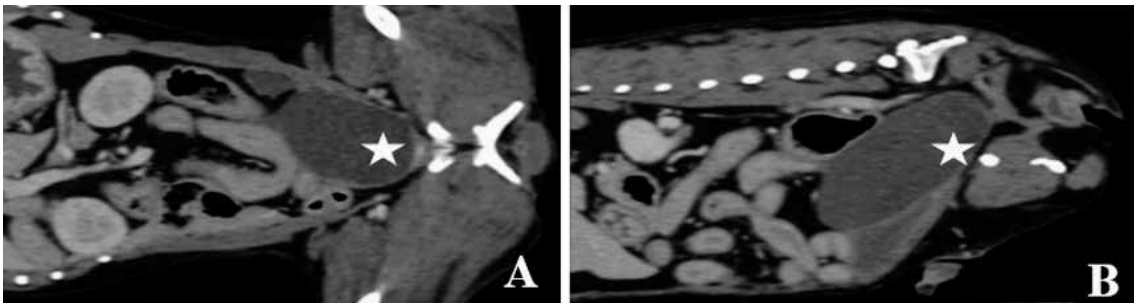

Figure 1: Coronal (A) and sagittal (B) computed tomography (CT) pictures of the abdomen. Cranial is left in the images. There is a large fluid-filled tubular structure visible in the caudal abdomen (white star), located between the descending colon and urinary bladder with a moderate mass effect.

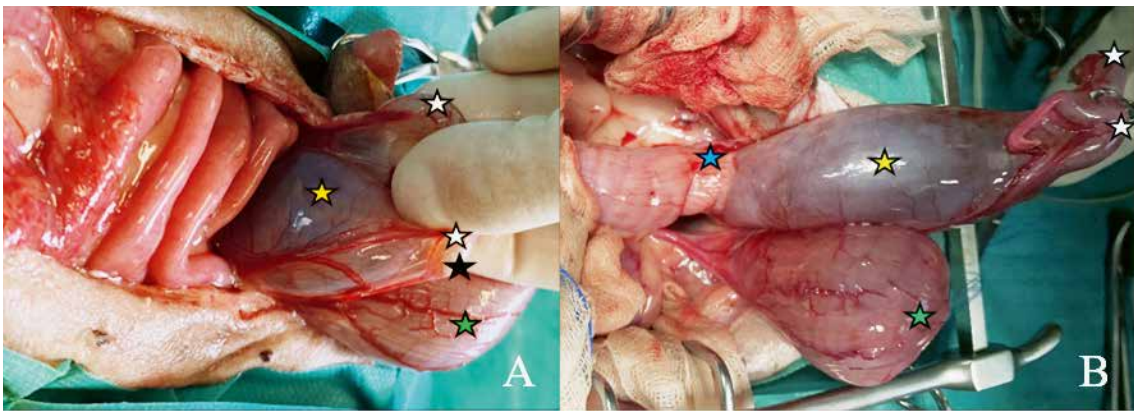

Figure 2: Intraoperative images of the ovariohysterectomy. Cranial is to the left in the images. (A) At the dorsal aspect of the bladder (green stars) a big cystic like structure (yellow stars) was revealed, which looked like a malformed uterus, with two uterine horns (white stars). On the left side of the uterine horns some ovarian tissue was present (black star). (B) There was a fusion between the cystic mass and the serosa of the rectum (blue star).

sible to the urethra. Before closing the abdomen, a peritoneal lavage and omentalization of the minimal residual portion of the cystic lesion attached to the urethra were performed.

The excised cystic uterine structure was sent for histopathological and bacteriological examination. The histologic image showed a cavity, lined by multilayered non-keratinizing epithelium. Subepithelially, there was a thick wall consisting of smooth, interrupted muscle fiber bundles and fibroblast-like cells. Histology confirmed the suspected diagnosis of mullerian duct. No bacterial growth was detected.

The dog recovered well from the anesthesia and was kept hospitalized overnight. He received paracetamol (Perfalgan Inf. Lös. 1 g/100 ml, Bristol-Myers Squibb SA, Schweiz, 10 mg/kg i.v. TID), ampicilline-sulbactam (Ampicillin/Sulbactam Kabi 1000 mg/500 mg, Fresenius Kabi Deutschland GmbH, Deutschland, 30 mg/kg TID) and fluid therapy (Plasma Lyte A solution, Baxter AG, Schweiz, 2 ml/kg/h). Opioids were not given due to a reaction with cyanosis and dyspnea after administration of methadone (Methadon Streuli Inj. Lös., 10 ml/ml, Streuli Pharma AG, Schweiz, 0, 2 mg/kg i.v.). These symptoms stopped witrh the administration of oxygen and naloxone (Naloxon OrPha Inj. Lös. 0, 4 mg/ml, OrPha Swiss GmbH, Schweiz, 0, 02 mg/kg i.v.). After surgery, the patient could urinate without difficulties. The following day, the patient was discharged to the owners' care. The owners reported that the dog urinated normally after surgery and was energetic.

Discussion

Male pseudohermaphroditism, that is caused by a persistens mullerian duct syndrome, often remains undetected due to the normal appearance of the external genitalia in affected dogs.[2, 15] This is probably due to the fact that after puberty, the genitalia exert their function and thus cystic endometrial hyperplasia, pyometra, cystitis, hydrometra, Sertoli-cell tumors, and epididymal malformations may occur.[15] In the present case, the dog showed no symptoms of persistent mullerian duct syndrome until 12 years of age, which has not been described until now. He became symptomatic when intermittent cystitis occurred.

With the very slow and gradual filling of the hollow organ with fluid, and a consequent mass effect at the caudal abdominal and cranial intrapelvic area, stranguria and tenesmus occurred at the age of 14 years. The urinary difficulties probably led to inflammation, which then provoked the symptomatology of cystitis. The symptoms can also be caused by ascending bacterial infections, but the bacteriological examination of the urine in this dog was negative.

It is not apparent why fluid had formed in the hollow organ. Bacteriological examination of the hollow organ showed no growth and no connection to the urinary organs could be found during surgery. Thus, a pyometra or a urine filled structure was not considered. Since no glandular tissue were identified in histology, the origin of the fluid formation is unknown.

As mentioned above, there is a high suspicion that the recurrent cystitis in this dog was also due to persistent mullerian duct syndrome. Since the treatment of choice is gonadectomy with hysterectomy to resolve and prevent additional clinical signs, earlier surgical exploration and therapy could have been considered in this dog.[3, 12] Most case reports describe early surgical therapy to prevent further symptoms.[3, 8, 13–15] As this dog initially showed mild symptoms that always resolved, already had other pre-existing diseases (mitral endocardiosis, tracheal colapse) and due to his age, earlier therapy was not undertaken.

In dogs and humans, persistent mullerian duct syndrome is commonly associated with cryptorchidism.[1, 9, 10, 15] Approximately 50 % of dogs with persistent mullerian duct syndrome are thought to be unilateral or bilateral cryptorchid,[2, 12] accordingly, cases described in several case reports were cryptorchids.[7, 8, 13, 15] This may lead to the development of Sertoli cell tumors in undetected cases.[15] In humans, cryptorchidism is thought to be caused by mechanical obstruction by the retained organs such as the ovaries, fallopian tubes, and uterus. If the persistent mullerian duct can be mobilized, the testicular descent is possible.[6] In dogs, the cause is not yet clear. The dog in this case report was not a cryptorchid according to the medical records. He was castrated at the time of presentation and no testicular-like structures were found at laparotomy described in this case report.

To the author's knowledge, there are no studies on the inheritance of persistent mullerian duct syndrome in Yorkshire Terriers, but there are some case reports about sexual development disorders of the breed. In these case reports, the dogs were found to have karyotype XY, with presence of both testes and internal female reproductive organs. Laparotomy was also performed in these animals to confirm the findings and to remove the internal reproductive organs.[4, 5, 14] No karyotyping was performed in the patient from this case report. The diagnosis was based on clinical history, diagnostic imaging, laparotomy, and histopathology. Further studies would be necessary to investigate possible inheritance and gene mutations in Yorkshire Terriers.

The diagnosis and treatment of sexual development disorders are challenging in small animal medicine, because they are not common. This case report can be used as an example for the diagnosis and treatment of persistent mullerian duct syndrome. In addition, it shows that even in a 14-year-old patient with recurrent cystitis, persistent mullerian duct syndrome should not be excluded as a differential diagnosis.

Persistent mullerian duct syndrome in a dog

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Syndrome du canal de Müller persistant chez un chien

Ce rapport de cas décrit un York Shire Terrier mâle castré de 14 ans, qui a été présenté en raison d'une strangurie et d'un ténesme.

Une tomodensitométrie (TDM) abdominale a soulevé une forte suspicion d'un canal de Müller persistant. Une laparotomie a été pratiquée, au cours de laquelle on a découvert des structures suspectées d'être un ovaire et un utérus persistants. L'utérus anormal était rempli de liquide et avait une extrémité aveugle des deux côtés. Les structures anormales ont été retirées chirurgicalement et ont fait l'objet d'un examen pathologique. La pathologie a confirmé la présence d'un canal de Müller persistant. Le patient s'est bien remis de l'opération et a pu uriner spontanément le jour même. Il a été autorisée à sortir le lendemain.

Mots clés: Rapport de cas, tomographie assistée par ordinateur, chien, syndrome du canal de Müller persistant, strangurie, ténesme.

Resoconto della sindrome da persistenza dei dotti mulleriani in un cane

Questo resoconto descrive il caso di un York Shire Terrier maschio castrato di 14 anni, che manifestava stranguria e tenesmo.

Una tomografia computerizzata (TC) addominale ha sollevato il sospetto di una sindrome da persistenza di un dotto mulleriano. È stata eseguita una laparotomia, durante la quale sono state trovate strutture che si sospettavano essere un'ovaia e un utero persistenti. L'utero anomalo era pieno di liquido e presentava un'estremità cieca su entrambi i lati. Le strutture anomale sono state rimosse chirurgicamente e sottoposte a esame patologico. La patologia ha confermato la presenza di un dotto mulleriano persistente. Il paziente si è ripreso bene dall'intervento ed è stato in grado di urinare spontaneamente il giorno stesso ed è stato dimessa il giorno successivo.

Parole chiave: Resoconto, tomografia computerizzata, cane, sindrome da persistenza dei dotti mulleriani, stranguria, tenesmo

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Korrespondenzadresse

Zita Szabo
Kleintierklinik Bern
Länggassstrasse 128
CH-3012 Bern
Telefon: +41 76 561 03 13
E-Mail: zita.szabo@vetsuisse.unibe.ch