Segmental Caudal Vena Cava Aplasia and Porto-Azygos Shunt in a Female Shih-Tzu Dog

Yaffe, M.* and Aroch, I.

*The Hebrew University Veterinary Teaching Hospital, Koret School of Veterinary Medicine, The Robert H. Smith Faculty of Agriculture, Food and Environment, The Hebrew University of Jerusalem, P.O. Box 12, Rehovot, 761001, Israel.

* Corresponding author: Dr. Marganit Yaffe, Koret School of Veterinary Medicine, The Hebrew University of Jerusalem, P.O.Box 12, Rehovot, 761001, Israel. Email address: marganit.yaffe@gmail.com

ABSTRACT

Segmental aplasia of the caudal vena cava (CVC), also termed azygos continuation of the CVC, CVC uniting with the azygos vein, and CVC absence, interruption or discontinuation, is a very rare congenital anomaly in dogs, reported only several times in dogs, and is similar to segmental aplasia of the inferior vena cava (IVC) in humans (also termed IVC atresia). An intact female Shih-Tzu dog, aged 18 months was presented with a history of aimless walking, hematochezia and hypersalivation. Laboratory tests showed hyperammonemia, increased liver enzyme activity and decreased liver functions, suggestive of hepatic encephalopathy. Abdominal ultrasonography and computed tomography (CT) with angiography showed several marked vascular anomalies, including segmental CVC aplasia and porto-azygos shunt. This combined vascular anomaly is an extremely rare congenital condition in dogs, since azygos continuation of the CVC occurs most often in the absence of portosystemic shunting. Based on the imaging findings, surgical correction of this shunt was not an option. The dog was therefore treated conservatively with oral lactulose and a commercial prescription hepatic diet. The dog improved clinically, and was stable over a nine month follow-up period. CT scanning with contrast media angiography was a sensitive tool to detect the portosystemic shunt and to demonstrate this unique and rare combined vascular anomaly.

Key words: Canine; CVC Aplasia; Porto-Systemic Shunt; Hyperammonemia; Hepatic Encephalopathy; Computed Tomography.

INTRODUCTION

Segmental aplasia of the caudal vena cava (CVC), also termed azygos continuation of the CVC, CVC uniting with the azygos vein, and CVC absence, interruption or discontinuation, is a congenital anomaly in dogs (1,2). This condition in dogs, as well as the similar segmental aplasia of the inferior vena cava (IVC) in humans (also termed IVC atresia), are rare congenital vascular anomalies (1, 2). It is usually identified as an incidental finding in imaging studies performed for diagnosing portosystemic shunts (PSSs), or upon laparotomy or anatomic dissection (3, 4). In this condition, the segment of the CVC between the kidneys and the liver is not formed, while the renal and post-renal caval blood is shunted instead towards an enlarged azygos vein, allowing functional venous blood return to the heart (4,5). In dogs, the anomaly may or may not be associated with portocaval shunting (6). Understanding the anatomy and function of the anomalous but vital cavo-azygos shunt vessel is essential when considering surgical intervention for correcting an associated PSS (1).

In humans, congenital caval vein anomalies, first described in 1793, are often associated with additional abnormalities, such as heart defects, situs inversus or a polysplenia-asplenia-syndrome (7, 8). Isolated, congenital malformations, such as IVC aplasia are rare, with an estimated prevalence of ap-
proximately 1% of all congenital vascular anomalies (9). It is found in approximately 5% of cases of unprovoked lower extremity deep venous thrombosis in young adults, which occurs significantly more commonly before the fourth decade of life (10).

In congenital PSS, there are abnormal connections between the portal system blood vessels (i.e., splenic, phrenic, cranial mesenteric, caudal mesenteric, gastric, or gastroduodenal veins) to the CVC or to the azygos vein (11). Porto-azygos shunt is a congenital extrahepatic PSS, mainly caused by a splenic and azygos vein connection, and is the most common extrahepatic congenital PSS in dogs in North America (12).

Clinical signs of congenital PSS (cPSS) result from nervous, urinary tract and digestive system abnormalities (13). General signs in dogs include poor weight gain, stunted growth and poor recovery from anesthesia. Neurological manifestations are the most common clinical abnormalities, resulting from hepatic encephalopathy (HE), and include behavioral changes, depression, disorientation, dementia, tremors, seizures, blindness, ataxia, stupor and coma (13). Older dogs may only present signs of cystitis or urinary tract obstruction from urate urolythiasis and ammonium-biurate crystaluria. Gastrointestinal signs of PSS include vomiting and diarrhea (13).

The laboratory abnormalities of cPSS in dogs may include microcytic anemia, leukocytosis, decreases urea concentration, hypoalbuminemia, hypocholesterolemia and hypoglycemia, and increased liver enzyme activities (13, 14). Serum bile acid concentration is increased. Hyperammonemia occurs in 90% to 100% of dogs with cPSS. Urine is often isosthenuric or hypostenuric. Urine sediment examination might show ammonium-biurate crystals (13, 14).

Imaging diagnosis of cPSS mainly includes abdominal ultrasonography and computed tomography (CT). Most cPSSs are detected by ultrasonography, with 80-95% sensitivity and 67-100% specificity, when performed by well experienced sonographers (15, 16). This wide variation in diagnostic accuracy between different studies likely reflects high inter-operator variability, inconsistent aberrant vessel visualization and advances in equipment imaging quality and technology over time (14-16). Even if the PSS is not visualized upon ultrasonography, other findings, such as renal and urinary bladder sediment, renomegaly, microhepatica, reduced portal marking and ascites, may support the diagnosis (15).

Portograms provide the diagnosis and location of the PSS (17). Radio-opaque, sterile, water soluble contrast is injected intraoperatively into a catheterized jejunal vein, or ultrasound-guided to the splenic vein preoperatively. The sensitivity of intraoperative portography is between 85% and 100%, and depends on the patient’s positioning (13). Nowadays, with the increasing availability of less invasive imaging modalities (i.e., advance ultrasound, scintigraphy and CT angiography), portography is less commonly performed in many large facilities (13).

Nuclear scintigraphy can also be used for diagnosing cPSS, either trans-splenically or rectally. Both methods enable calculation of the shunt fraction, which represents the blood volume flowing through the shunting vessel, bypassing the liver. The reported sensitivity and specificity for per-rectum and trans-splenic scintigraphy are 88% and 67%, and 100% and 100%, respectively (18).

The diagnostic gold standard for cPSS is dual phase CT angiography, allowing reconstructing the abnormal blood vessels, and identifying their number, location, and termination, with 96% sensitivity and 89% specificity (16, 19). Magnetic resonance imaging (MRI) with angiography (MRA) provides 3-dimensional imaging of the shunt with good to excellent detail (20). Although MRA is a promising diagnostic modality for cPSS, CT angiography provides similar detail, is performed more quickly, and is more cost-efficient than MRA (14).

In this report, we describe a case of concurrent segmental CVC aplasia and porto-azygos shunt in an 18 months old female Shih-Tzu dog, which initially presented with signs of HE.

**CASE REPORT**

An 18-month old, intact female Shih-Tzu dog was referred to the Koret School of Veterinary Medicine Teaching Hospital (KSVMT) with a history of aimless walking, hematochezia and hypersalivation of 24-hour duration. The dog was adopted as a puppy, and was described by its owner as a “sleepy” dog. Two months previously, the dog was presented to the KSVMT with an episode of aimless walking and polakiuria. It tested positive for tetrahydrocannabinol (THC), and was also diagnosed with cystitis. It received supportive treatment, including amoxicillin-clavulanic acid (Augmentin, SmithKline Beechman Ltd., Worthing, UK; 70 mg q12h PO for 14d), and recovered.
At presentation to the KSVMTH, the dog had a low body condition score (3/9), mildly decreased rectal temperature (37.8°C), tachycardia (160 bpm), hypersalivation, tightly closed jaws, bloody stool upon rectal examination and showed aimless walking and did not respond to her owner.

Complete blood count with microscopic examination of the blood smear showed microcytic hypochromic regenerative anemia (based on presence of polychromasia and mild metarubricytosis), neutrophilic leukocytosis (Table 1), while upon examination of the stained blood smear, the neutrophils showed mild left shift and cytoplasmic toxicity. Serum chemistry abnormalities included mildly increased activities of some hepatobiliary enzymes, hypoglycemia, mild hyperbilirubinemia, hypocalcemia, hyponatremia, hypochloridemia, mild hypocholesterolemia and fasting plasma hyperammonemia (Table 1).

Ultrasonographic evaluation of the abdominal structures was performed with a micro-convex array transducer in B-mode at a frequency range of 3-11 MHz (Mindray DC-8 and Mindray M-9, Mindray Bio-Medical Electronics, Shenzhen, China). Harmonic imaging was used in all scans.

Abdominal ultrasonography demonstrated marked distension of what was then suspected to be a segment of the CVC, between the renal veins and the liver, with abnormal vascularization within the hepatic hilus, mildly decreased liver size, enlarged kidneys, sediment within the urinary bladder and small volume of free, anechoic abdominal fluid. Extrahepatic PSS was suspected. Ultrasonographic evaluation of the abdominal structures was performed with a micro-convex array transducer in B-mode at a frequency range of 3-11 MHz (Mindray DC-8, Mindray Bio-Medical Electronics, Shenzhen, China).

A CT scan with angiography was performed using non-ionic contrast medium (Omnipaque, GE Healthcare, Cork, Ireland; 300 mg/kg IV). Post contrast scanning was performed, 5 and 28 sec post manual injection, to mimic arterial and portal phases. A 16 single slice helical scanner (CT imaging: Philips MX8000 IDT; 16 slice MDCT; Philips, Cleveland, OH, USA) was used, with acquisition parameters of 1 mm thick contiguous slices, at 120 kVp and 320 mA. Images were viewed with dedicated viewing software, with 3D capabilities (Fujifilm Synapse, FujiFilm Medical System USA, Stamford, CT), and a soft tissue algorithm. In the pre-contrast images, the liver was irregular, and its left

![Figure 1: Volume-rendered 3-dimensional projection of the computed tomography angiography study. The shunted portal vein (P) merges the azygos vein (Az). There is severe azygos vein and caudal vena cava (CVC) distention (Ao, aorta; Ce, celiac artery; CMa, cranial mesenteric artery; *, location of the shunt).](image-url)
lobes were markedly small, while bilateral renomegaly was noted. Post-contrast scans demonstrated the portal vein, originating from the confluence of the cranial and caudal mesenteric veins, coursing dorsally to insert the pre-hepatic CVC, cranially to the renal veins. The portal trunk was tortuous and dilated (Figures 1 and 2). The splenic, cranial pancreaticoduodenal and gastro-duodenal veins traveled caudally, to join the portal vein as it coursed dorsally (Figure 3). The pre-hepatic CVC was markedly dilated at the shunt insertion, and several centimeters cranially, it merged dorsally to join the azygos vein at its origin, near the second lumbar vertebra (Figure 4). This severely dilated right sided azygos vein paralleled the aorta, crossed the diaphragm dorsally, and entered the right atrium directly at the coronary sinus (Figure 3). Cranial to the confluence of the CVC and azygos vein, there was no apparent continuation of the CVC through the liver. The portal vein was not visible between the portal trunk and the liver, and the portion of the pre-hepatic and the entire hepatic CVC were absent (Figure 2). The small hepatic veins converged to form a post-hepatic vena cava that was positioned in the right diaphragmatic crus, and traveled slightly to the right, towards the right atrium. In addition, post contrast scanning demonstrated moderate thickening of the small intestine wall (Figure 3A).

With the high risk and low survival chances of surgical correction of such vascular anomalies, the owners elected conservative treatment. This treatment included commercial prescription hepatic low protein diet, (Monge VetSolution Hepatic, Monge & C. S.p.A, Monasterolo di Savigliano, Italy) and lactulose (Avilac, Perrigo Israel Pharmaceuticals Ltd., Shoham, Israel). Nine months later, the dog was clinically normal and has gained weight.
DISCUSSION
This report describes a rare congenital anomaly of azygos continuation of the CVC, combined with PSS. Similar cases have been infrequently reported in the veterinary literature, all in dogs (1, 4, 6, 21), and none were reported in cats. In dogs, azygos continuation of the CVC occurs most often in the absence of portosystemic shunting (22), unlike the present case. In humans, the prevalence of azygos continuation of the IVC is approximately 0.6%-1% in patients with congenital heart disease (9, 23).

The embryonic CVC is formed through a complicated process of development, anastomosis and replacement of three vascular systems, namely the posterior cardinal, subcardinal and supracardinal venous channels, leading to hepatic, prerenal, renal and post-renal segments of a right sided CVC (2). In the case of fusion failure between the right subcardinal and the hepatic veins, the post renal CVC segment and the cranial portion of the supracardinal vein (the later azygos or hemiazygos veins) will persist, forming this anomaly (2). Thrombus formation in an aneurysmal cavo-azygos shunt vessel is a possible rare outcome in dogs (1), while in humans, deep venous thrombosis is the main cause that eventually triggers the diagnosis of segmental IVC aplasia (9).

CT angiography with multi-phase technique is the gold-standard diagnostic imaging technique, which allows evaluation the entire vascular system (i.e., arterial, portal and caval) using a single iodinated contrast medium bolus, injected into a peripheral vein. In this case, an automatic injection system was not used, and instead, the dog was rescanned immediately after manually injecting the contrast media, as well as at 28 seconds later, approximately providing resolution first the arterial phase, and later the portal phase, respectively. The high sensitivity and specificity of this method is beneficial for surgical or minimally invasive interventional radiology shunt correction (19), or, as in this case, to avoid unnecessary surgical interventions. Furthermore, detailed axial images and additional techniques, such as 3-dimensional volume rendering and thick-slab maximum intensity projections (MIP), were used to demonstrate the abnormal vessels, along their entire course, and for surgical planning (Figures 1-4).

This dog presented a microcytic anemia, which occurs commonly in dogs with cPSS (24-26), although the precise pathogenesis of cPSS-associated anemia remains unclear. In such dogs, hypoferremia is common, occurring in 56 to 70% of cases, but based on evaluation of other iron status markers (e.g., normal to decreased total iron binding capacity, variable transferrin saturation and increased serum ferritin concentration and hepatic stainable iron) it is likely unassociated with absolute iron deficiency (24, 25, 27). Iron parameters have normalized post partial shunt attenuation, which is suggestive of a causal relationship between these abnormalities and cPSS (24). This anemia in dogs with a cPSS is unassociated with dysregulated hepcidin production (28). As to the increased activities of ALP, GGT and AST observed in the present dog, hepatobiliary enzyme activities, especially increased ALT and ALP activities, these are mild to moderately increased in75% of dogs with PSS (29, 30). Hyperbilirubinemia, hypoglycemia and hypocholesterolemia, observed herein, were likely due to reduced liver function (31).

In conclusion, this is a report of a rare combination of congenital segmental CVC aplasia and porto-azygos shunt in a dog, which is a rare phenomenon. The segmental CVC aplasia was diagnosed incidentally, when the dog presented signs of HE due to the PSS. In this case, CT angiography was a crucial tool for demonstrating the complexity of this unusual vascular anomaly. The poor prognosis of surgical intervention to correct the present vascular anomalies, combined with the excellent short-term response to conservative treatment, warrant continuing the latter form of treatment.

REFERENCES
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