Multiple Congenital PSS in a Dog: Case Report and Literature Review

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ABSTRACT

A 4 yr old spayed female mixed-breed dog presented with a 2 yr history of recurring increases in liver enzymes. Two congenital portosystemic shunts (PSSs) were identified using computed tomography (CT) angiography, which included a portoazygous and portorenal extrahepatic shunt. Double right renal veins were also identified. The shunts were successfully identified and attenuated with cellophane banding. Multiple congenital PSS is a rare phenomenon, but should be considered during exploratory laparotomy for PSS and in dogs with poor response to surgical attenuation of a single PSS. CT proved to be a crucial part of accurate diagnosis and surgical planning for this dog with multiple congenital PSS. (J Am Anim Hosp Assoc 2013; 49:281–285. DOI 10.5326/JAAHA-MS-5877)

Introduction

Portosystemic shunts (PSSs) are abnormal communications between the portal vasculature and the systemic venous circulation.1 The communication allows blood from the gastrointestinal tract to bypass the liver, which often results in clinical signs related to hepatic insufficiency. A PSS can be characterized either as acquired or congenital. Developmentally, congenital shunts arise from aberrant connections between the vitelline and cardinal venous systems.1 They can also be anatomically classified as being either intrahepatic or extrahepatic. Congenital shunts are usually single vessels, whereas acquired shunts are typically multiple vessels that develop in response to chronic portal hypertension. The presence of acquired portosystemic collateral circulation secondary to a single congenital PSS has also been reported, but appears to be a rare and controversial phenomenon.2

The purpose of this case report was to describe the diagnostic imaging, anatomic characterization, and successful surgical attenuation of two separate congenital extrahepatic PSS in a dog.

Case Report

A 4 yr old spayed female mixed-breed dog weighing 17.5 kg presented for evaluation of recurring increased liver enzymes. Initial laboratory abnormalities included an increase in alanine aminotransferase (ALT; 279 μkat/L; reference range, 9–86 μkat/L), which was first reported approximately 2.5 yr prior to presentation. The elevation was detected after a serum biochemical analysis, complete blood count (CBC), and urinalysis were performed to rule out causes of inappropriate urination and pollakuria. Urinary cultures were not submitted. All other laboratory values were within normal limits. The dog was prescribed a prescription diet formulated for hepatic insufficiencya. Serum biochemical analysis was repeated intermittently over a 1 yr period, and ALT returned to within the normal reference range. The prescription diet was then discontinued for a period of time due to normal liver enzyme values; however, additional analyses were performed due to recurrence of clinical signs indicative of an increase in ALT (168 μkat/L), and the prescription diet was reintroduced. One mo prior to referral, ALT remained increased (659 μkat/L).

Physical examination at the time of referral was unremarkable, with the exception of a juvenile vulva. Serum biochemical analysis revealed a high ALT (764 μkat/L; reference range, 17–86 μkat/L) and elevated aspartate aminotransferase (93 μkat/L; reference range, 9–42 μkat/L). Resting and postprandial serum bile

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Acids were 133 μmol/L (reference range, 0.0–5.0 μmol/L) and 278 μmol/L (reference range, 0.0–25.0 μmol/L), respectively. No other laboratory diagnostic tests were performed at that time.

Abdominal ultrasonography revealed microhepatica with normal echogenicity. There were multiple, linear, hyperechoic regions in the right and left kidneys and multiple small (< 5 mm in length) hyperechoic structures seen along the ventral border of the urinary bladder. Those hyperechoic structures were gravity dependent and most consistent with small ammonium urate cystoliths. The porta hepatitis was difficult to visualize due to the patient’s size, and no abnormal vessels were found. A hypoechoic nodule, measuring 5 mm × 4 mm, was seen within the tail of the spleen. Abdominal radiographs revealed microhepatica and splenomegaly. Nonselective contrast was administered IV during abdominal CT, and two large extrhepatic PSSs were identified. The first was a splenoazygous PSS originating from the splenic vein adjacent to its communication with the portal vein (Figure 1). The vessel then coursed cranially and to the left. At the level of the stomach, the PSS coursed dorsally through the diaphragm and continued into the azygous vein. Cranial to the communication of the portal and splenic veins, the portal vein was 2 mm smaller in diameter than caudal to the communication. The second smaller shunt was seen originating from the portal vein, caudal to the insertion of the splenic vein, and inserting into a focal dilation on the craniodorsal aspect of the left renal vein (Figure 2).

An exploratory laparotomy was performed through a ventral midline approach. Upon entering the omental bursa and reflecting the stomach ventrally, an anomalous vessel from the splenic vein coursing craniodorsally and through the diaphragm was identified. The right pararenal gut was exposed by retracting the mesoduodenum toward midline, and a second extrhepatic shunt was found originating from the portal vein, traveling a short distance caudally and entering the left renal vein. Two right renal veins were also noted. The distal aspect of each shunt was carefully isolated with gentle dissection. Cellophane was then placed around each of the anomalous vessels and secured with two hemostatic clips. The liver appeared small with some multifocal discoloration. A wedge biopsy was taken from the right medial liver lobe. Recovery was uneventful, and the dog was discharged 2 days following surgery.

The liver biopsy results revealed moderate lobular hypoplasia and diffuse mild duplication of small diameter arteries. The midzonal to centrlobular hepatocytes were moderately swollen with fragmented cytoplasm. Some portal triads had either very few veins or lacked them altogether. Those findings were consistent with the presence of a PSS and microhepatica.

CT angiography was performed 1 mo postsurgically to re-evaluate the attenuation of the shunts. The splenoazygous shunt was approximately 50% smaller in diameter at the level of the cellophane band. The lumen of the shunt at the level of the cellophane was not visualized due to artifact associated with the hemoclips placed during surgery. The diameter of the portal vein cranial to the splenoazygous shunt increased in diameter by approximately 33%. The portorenal shunt was also attenuated at the level of the cellophane band, but the degree of shunting could not be accurately assessed. There was no change in diameter of the shunt proximal to the cellophane.

Three mo postsurgically, fasting and postprandial bile acid results were 19.8 μmol/L (reference range, < 10.0 μmol/L) and 36.4 μmol/L (reference range, < 20.0 μmol/L), respectively. A CBC and serum biochemical analysis were within normal limits at that time. Six mo postsurgically, fasting and postprandial bile acids were 14 μmol/L (reference range, 0–13 μmol/L) and 12 μmol/L (reference range, 0–30 μmol/L), respectively. Approximately 1 yr after surgery, a CBC, serum biochemical analysis, and urinalysis results were all within normal limits. The dog was maintained on the prescription diet.

FIGURE 1 Parasagittal multiplanar reformat computed tomography (CT) angiography image of the portoazygous shunt arising from the splenic vein. The portoazygous shunt (hollow arrows) passes through the diaphragm when followed cranially. The portal tributaries (long white arrow) and communication between the splenic vein and anomalous vessel (short white arrow) are also seen. Ao, aorta.
and no clinical signs of PSS were reported at the time of that examination.

Discussion

Multiple congenital extrahepatic PSSs are rare, as they are usually singular vascular anomalies.1–3 To the authors’ knowledge, multiple congenital PSS has only been previously reported in eight cases.2–5 One dog was identified as having two distinct anomalous vessels on transsplenic portal scintigraphy, which was confirmed during exploratory laparotomy.3 In that dog, one vessel originated from the splenic vein, bifurcated, and terminated with one branch to the caudal vena cava and one branch through the diaphragmatic crus, presumably to the azygous vein. The second shunt was a portocaval shunt.3 A similar pattern of multiple congenital PSS was reported in a miniature schnauzer with both portocaval and portoazygous shunts. The shunts arose from a common trunk of the portal vein and could be traced to the vena cava and azygous vein, respectively.4 An unusual description of multiple congenital extrahepatic PSS was seen in a young golden retriever with one aberrant vessel arising from the portal vein and a second arising from the splenic vein. Those two anomalous vessels anastomosed along the midline at the level of the twelfth thoracic vertebra, coursing cranially before terminating in the posthepatic caudal vena cava.2 Another study reported a relatively high incidence of multiple congenital shunting, where 5 of 46 dogs with congenital shunts had double extrahepatic PSS.5 Detailed descriptions of the shunts were not included in that study. For all dogs previously identified with multiple congenital PSS, specific clinical signs, surgical intervention, and outcomes were not reported.

Congenital PSS may be amenable to direct occlusion, whereas no successful surgical treatment options exist for acquired shunting. Therefore, multiple congenital extrahepatic PSS should not be confused with multiple acquired shunts. Several patterns of collateral shunting, which develop secondary either to portal hypertension or cranial vena cava syndrome, have been described. They can be either multiple, small, tortuous vessels in the renal region, which are found most commonly, or a longer, tortuous vessel between the splenic vein and left renal vein.6 This may also be termed a splenogonadal shunt. Those types of acquired shunts were not suspected in the dog described in this case due to the pattern of shunting, lack of clinical portal hypertension, absence of cranial vena cava syndrome, and absence of multiple small tortuous vessels.

It could be suggested one of the dog’s shunts was congenital, while the other was acquired; however, it is unlikely that a dog with a congenital PSS would develop spontaneous portal hypertension and acquired portosystemic collaterals.6,7 While resistance of blood flow through the hepatic sinusoids via the portal vein in dogs with congenital PSS may be high, studies have shown that 95–100% of the portal blood flows by way of the congenital PSS instead of the hypoperfused vessel because the resistance is lower through the shunt.6–10 Portal pressures then equilibrate with systemic venous pressures and are either within the reference range or lower than normal.7 Acquired shunting in the presence of a congenital PSS would require occlusion of the congenital vessel, which would cause increases in portal pressures (such as by a thrombus, tumor, or surgery).

Persistently high serum bile acids, requirement for ongoing medical management, and recurrence of clinical signs are not uncommon after surgical attenuation of congenital PSS.1,11 Possible causes for those complications include residual flow through the shunt, persistent hepatic insufficiency due to inadequate adaptation by the liver, incorrect identification of the anomalous vessel during surgery, or the presence of multiple PSS. This case report highlights the importance of performing a thorough evaluation of the portal vasculature during surgery, even after a single congenital PSS has already been identified.
Currently, abdominal ultrasound and nuclear scintigraphy are the most commonly used imaging modalities for dogs suspected to have PSS. There are considerable differences in the reported accuracy for use of ultrasound to detect PSS, with sensitivity and specificity values reported as low as 47% and 67%, respectively.\textsuperscript{12,13} Transspenic scintigraphy has shown more promise in identifying either multiple congenital or acquired PSS, but the anatomic detail provided by those studies is limited.\textsuperscript{1} CT angiography, which is considered the “gold standard” for assessing intra-abdominal vasculature in human medicine, is becoming a more popular modality for evaluation of the hepatic and portal vasculature in animals.\textsuperscript{14} It has the main advantage of providing superior characterization of normal and abnormal vascular anatomy, which can be viewed in either multiple two-dimensional planes or as a three-dimensional reconstruction.\textsuperscript{6} In this case, abdominal ultrasonography failed to confirm the presence of the PSS, whereas CT angiography precisely characterized the course of both anomalous vessels. Moreover, the authors of this report believe the identification and appropriate attenuation of the portorenal PSS in particular would not have been accomplished without the use of preoperative CT angiography in this case. With more widespread use of this advanced imaging modality, it is likely the detection of multiple congenital PSS and other unusual vascular anomalies will be identified more frequently than previously reported.

Assessing vascular occlusion following surgery has been attempted using nuclear scintigraphy with limited success.\textsuperscript{15} Based on this case, it appears that CT may be one noninvasive imaging modality option that allows accurate assessment of postoperative shunting attenuation, as the authors were able to quantify the degree of shunt narrowing in the portoazygous shunt postoperatively. Although cellophane was used instead of an ameroid ring on the portorenal shunt to decrease metal artifact, some artifact remained due to hemoclips used to secure the cellophane. If an alternate method of securing the cellophane was used, there may have been a better assessment of shunt attenuation postoperatively. Gradual vascular occlusion could have also been achieved without resulting in metal artifact on CT angiography by using partial attenuation with either silk suture or hydraulic occluders.\textsuperscript{16} However, complete occlusion using silk is unpredictable and hydraulic occluders were not available at the time of surgery.\textsuperscript{16} Despite the lack further follow-up imaging, complete attenuation of the shunts was likely in this case as bile acids returned to within normal limits and there was no relapse of clinical signs associated with hepatic insufficiency identified during any of the follow-up examinations.

**Conclusion**

This case report characterized multiple congenital extrahepatic PSS in a dog, which is regarded as a rare phenomenon. The shunts were not visualized via ultrasonography and may have been overlooked during exploratory laparotomy without the information obtained from nonselective abdominal CT angiography. It is imperative to be thorough during the pre- and intraoperative evaluation of the portal vasculature in dogs suspected to have PSS because there may either be more than one shunt or other vascular anomalies present. Poor response to surgical intervention or persistence of clinical signs or abnormal laboratory values may be the result of an unidentified second congenital PSS. More sophisticated imaging techniques, such as CT angiography, may better help characterize unusual shunt morphologies and identify multiple vascular anomalies.

**FOOTNOTES**

\textsuperscript{a} Hill’s I/d; Hill’s Pet Nutrition Inc., Topeka, KS

\textsuperscript{b} Omnipaque; GE Healthcare Co. Ltd., Shanghai, China

**REFERENCES**


