

Diagnosis of Persistent Right Aortic Arch with an Aberrant Left Subclavian Artery using CT in a Poodle Dog

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Abstract : A 6-month-old, female poodle presented with a three-month history of persistent regurgitation immediately after eating. On physical examination, the patient was emaciated and dehydrated. Thoracic radiography showed ventral displacement of the trachea and increased radiopacity in the mediastinum, cranial to the heart base. A severely dilated esophagus was identified cranial to the heart on esophagram. Computed tomography (CT) revealed the esophagus was filled with gas, fluid and a little of contrast and dilated from caudo-cervical to cranio-thoracic part. The esophageal diameter was markedly decreased at the heart base. In addition, the trachea was displaced to the left-ventral side of the right aortic trunk and an aberrant left subclavian artery originating from the aorta was identified. There was no evidence of abdominal vascular anomaly. Based on diagnostic imaging, persistent right aortic arch (PRAA) with an aberrant left subclavian artery was diagnosed. The patient did not undergo surgery and died at 15 days after diagnosis. This report describes imaging diagnosis, including CT and radiography in a weaned dog with regurgitation due to esophageal obstruction by PRAA. When PRAA is suspected and conventional radiography or contrast study is insufficient for diagnosis, CT may be helpful for diagnosing PRAA.

Key words : persistent right aortic arch, CT, vascular ring anomalies, aberrant left subclavian artery, dog.

Introduction

Vascular ring anomalies (VRAs) are congenital malformations of the great vessels and structures of their branches. VRAs are relatively uncommon, considering that although persistent right aortic arch (PRAA) is the most common type of VRAs (95%), it merely accounts for approximately 3.1% of dogs with congenital heart defects (5,7). Also, PRAA has a tendency to be with a left retro-esophageal subclavian artery or double aortic arch and other VRAs have only been reported as complex combinations of abnormal vessels in a few dogs (5,14). Clinical signs of PRAA include postprandial regurgitation resulting from esophageal obstruction, weight loss with normal appetite and cough or dyspnea secondary to aspiration pneumonia (2). Many cases of PRAA are frequently involved in large-purebred dogs such as the German Shepherd (5,7,10,18,21). In a previous retrospective study for 25 dogs with PRAA, 23 (92%) dogs did not have clinical signs at the point of long-term (more than six months after surgical correction) follow-up, suggesting that accurate diagnosis and surgical correction may lead to a favorable outcome (20).

When compared to conventional radiography, computed tomography (CT) can provide additional diagnostic informations and aid in presurgical plans or treatment for PRAA (14,22). Although radiography is readily accessible imaging

tool, it does not provide a definitive information of VRAs or the three-dimensional structure of the causal vessels even with additional esophagram (22). CT requires general anesthesia, is expensive, and involves radiation overexposure. However, with a single intravenous injection and a short scan time CT provides direct identification of contrast-enhanced causative vessels and a three-dimensional anatomy of VRAs (22,23,25).

This case report described CT and radiographic findings of PRAA with an aberrant left subclavian artery in a poodle dog.

Case

A 6-month-old, female poodle, weighing 1.05 kg presented with a three-month history of persistent regurgitation immediately after eating, especially dry and solid foods. On physical examination, the patient indicated poor body condition score of 2/9 and had rough skin condition. The patient was on state of anemia (red blood cell, $3.2 \times 10^{12}/L$; reference range, 4.7 to $8.5 \times 10^{12}/L$, hematocrit, 22.7%; reference range, 32 to 55%, hemoglobin, 9.8 g/dL, reference range, 10.3 to 18.0 g/dL in complete blood cell counts).

Conventional thoracic radiographs revealed focal ventral displacement of the trachea and a gas- or fluid-filled mass in mediastinum. It obscured the normal left ventricular and auricular margin of the heart in a right lateral view (Fig 1A). The trachea was not focally narrowed or curved leftward on ventrodorsal radiographs. An esophagram with barium sulfate (Solotop Suspension 140; Taejoon Pharm) showed severe dilation of the esophagus in the thoracic cavity and focal

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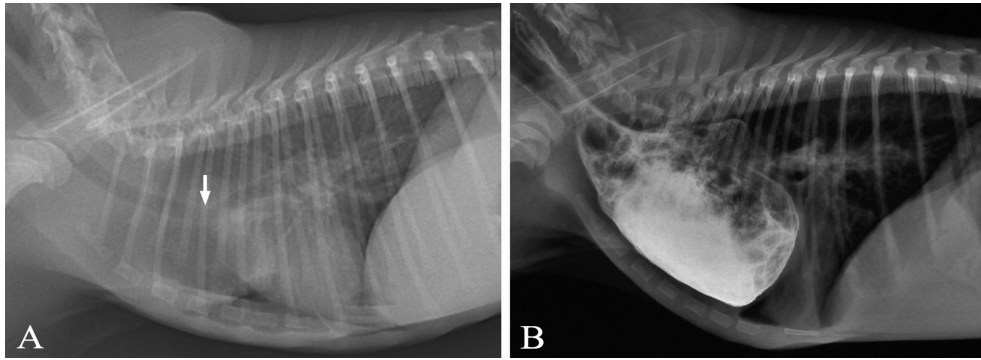


Fig 1. Right lateral thoracic radiograph (A) and esophagram (B). There are ventral curvature of the trachea (arrow) and mild loss of the auricular margin due to increased radiopacity in mediastinum cranial to the heart base. Esophagram shows the severely dilated esophagus.

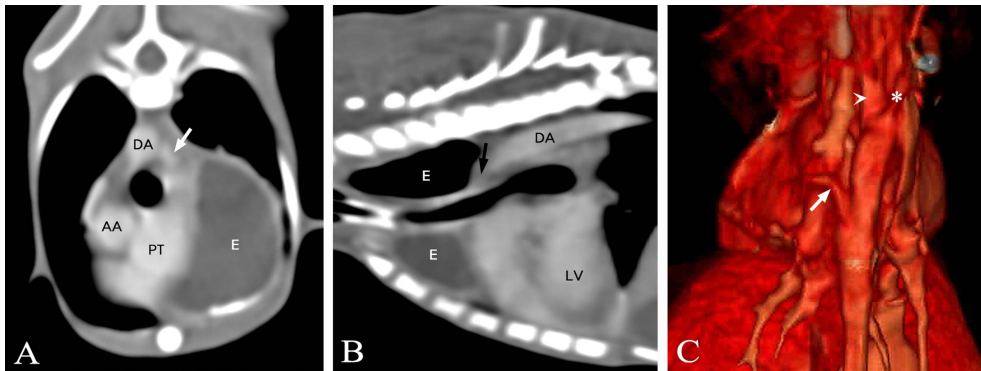


Fig 2. Post-contrast transverse (A), sagittal (B), and three-dimensional (3-D) reconstructed CT images (C). The dilated esophagus (E) becomes dramatically narrower at the heart base. The LSA (white arrows) originates from the left-lateral surface of the descending aorta and passes over the dorsal surface of the esophagus from the obstruction. The location of a non-enhanced LLA (black arrow) is surmised between the left-ventral portion of descending right aortic arch and the right-dorsal portion of main pulmonary artery near the compressed site of esophagus. The common carotid trunk (arrowhead) and right subclavian artery (*) are previously branched out from the early part of right aortic arch. AA, ascending aorta; DA, descending aorta; E, esophagus; LV, left ventricle; PT, pulmonary trunk.

esophageal compression at the level of the heart base (Fig 1B).

For CT scanning, anesthesia was induced by intravenous injection of 5 mg/kg propofol (Provive Inj. 1%; Claris Lifesciences) and maintained by inhalation of isoflurane (Ifran Solution; Hana Pharm). The patient was positioned in sternal recumbency and CT scanning was performed using a 32-multislice CT scanner (Alexion, Toshiba Medical System; Otawara, Japan). The scanning parameters were 120 mA, 200 kV, 1.0 mm slice thickness, and craniocaudal scan direction. A contrast study was performed after intravenous administration of 900 mg/kg Iohexol (Bonorex 300 Inj.; Daehan Pharm) at a speed of 0.5 ml/s for 20 sec, using a autoinjector. CT angiographic images were obtained 20 sec after injection.

CT revealed that the caudo-cervical to cranio-thoracic esophagus was severely dilated. The dilated esophagus was filled dorsally with gas and ventrally with fluid or a little of contrast media. The diameter of the dilated esophagus significantly narrowed at the heart base. The trachea was located left-ventral to the right aortic trunk. An aberrant branch vessel from aorta, 2.3 mm in diameter, was identified at the level of 4th intercostal space on post-contrast CT images. The

branch was identified as an aberrant left subclavian artery (LSA). The aberrant LSA originated from the left-lateral surface of descending aorta and went forward left-cranial direction over the dorsal surface of esophagus near to the obstructive portion. It was speculated that the left ligamentum arteriosum (LLA) was located near the compressed esophagus between the left-cranial portion of descending right aortic arch and the right-dorsal portion of main pulmonary artery (Fig 2). There was no evidence of abdominal vascular anomalies. Based on radiography and CT, PRAA with an aberrant LSA was diagnosed.

The owner declined surgical correction of the vascular anomaly. Although the owner chose conservative management, including feeding a liquid diet and intravascular nourishment, the patient died 15 days after diagnosis of PRAA.

Discussion

VRAs are congenital deformities of the aortic arches and their branches that entrap the esophagus. In the developing canine fetus, six pairs (first to sixth) of aortic arches are involved in the process of cardiovascular morphogenesis,

maturation, and differentiation. The aortic arches compose the mature aorta and its branches. The right fourth aortic arch normally becomes the proximal portion of right subclavian artery, while the left fourth aortic arch forms the normal adult aortic arch (3). In fetal development, if the right fourth aortic arch forms the main aortic arch instead of the left fourth aortic arch, non-regression of the right fourth aortic arch results in PRAA. Accordingly PRAA induces an abnormal LLA, a remnant of the ductus arteriosus, and esophageal obstruction resulted from the aorta and pulmonary trunk (or additional aberrant vessels) by surrounding the esophagus at the cranial heart base (2). A typical clinical sign is repetitive regurgitations after eating in weaned dogs, however the late-onsets of clinical signs have been reported (17).

Obstruction of esophagus may result from variant VRAs and a classification for these variants has been proposed (14, 23). Type 1, the most common, results in compression of esophagus at the level of heart base, entrapped by a LLA, right aortic arch and main pulmonary artery. Type 2 is less common and results in obstruction due to an aberrant LSA that pass across the esophagus in a cranio-dorsal direction. Cases of esophageal compression by LSA have been reported in Labrador Retriever and English Bulldog (2,14). Type 3 is double compression similar to type 2, with an additional LLA. Type 4 is the compression of esophagus by a double aortic arch. In addition, other various combinations of concurrent vascular anomalies at multiple locations can cause compression of the esophagus. Left aortic arch with a right ligamentum arteriosum and, left aortic arch with a patent ductus arteriosus have been reported (12,13). Three cases of unusual esophageal compression due to a connection between the LLA and an abnormal LSA have been reported in German Pinschers (18). In the present case, the patient was diagnosed PRAA, type 1 vascular ring anomaly. Since the ligament (remnant of the ductus arteriosus) was not filled with blood, the LLA causing the obstruction of esophagus was not contrast-enhanced and it was not indicated directly on post-contrast CT images (2,22). The patient also had an aberrant LSA, that did not compress the esophagus and no other compressions by defective vessels were detected. This form of PRAA coexisting with an aberrant LSA had occurred in 33% of dogs with VRAs in a previous study (5).

In dogs, PRAA is the most common type and accounts for 90% of VRAs (2,23). In a retrospective study of 55 dogs with one or more VRAs, 52 (95%) dogs had a PRAA complicated by other compressive vascular anomalies (5). Most cases of PRAA are frequently involved in young and large-breed dogs, such as German Shepherds, Greyhounds, and Irish Setters was included in the past, but recently excluded (5,6,10, 21). The breed-specific prevalence rates of cardiovascular malformations are higher in purebred dogs. Especially German Shepherd dogs have a 4.5 times of genetic predisposition for PRAA (7,21). In contrast to the medium or large-sized breed dogs described in most previous studies, our patient was a 1.05 kg small toy poodle. Cases of Yorkshire Terrier and domestic short haired cat also have been reported (5,10,15,17,18,24).

The obstruction of esophagus due to PRAA can result in life-threatening. The major clinical sign is regurgitation, the

degree of which is variable depending on the extent of esophageal compression caused by collaboration of the right aortic arch, LLA and main pulmonary artery. In addition, weight loss secondary to inadequate intake despite of normal appetite, cough and dyspnea may occur (2). The previous study of PRAA patients has reported 92% (23 of 25) dogs no indicated clinical signs at the point of more than six months after surgical correction in follow-up, suggesting that accurate surgical correction may lead to a good long-term outcome (20). In another previous retrospective analysis of the long-term outcomes for survival dogs following PRAA surgery, 87% (20 of 23) dogs indicated good or excellent outcomes (16). Surgery is an effective treatment for PRAA, and surgical planning based on accurate diagnosis is also important. CT can play a role in the pre-surgical procedure, including diagnosis and surgical approach (14,22). Unfortunately in the patient of this case, surgical treatment was not performed in spite of CT diagnosis.

In the previous cases, VRAs including PRAA were indirectly diagnosed by typical clinical signs, conventional thoracic radiography, contrast esophagram, endoscopic investigation, surgical exploration, or even post-mortem examination (1,2, 5,8,9,11,18,19,24). Especially tracheal deviation or focal narrowing on conventional thoracic radiography in suspected dogs was regarded as a reliable indicator of PRAA, even in the absence of an additional esophagram to identify the location of esophageal dilation (5). In a retrospective review of radiographs from 27 dogs with PRAA, 100% of the dogs had presented moderate or marked focal leftward curvature of the trachea near the cranial border of the heart in the dorsoventral or ventrodorsal view and, 74% indicated focal narrowing of the trachea (5). However, these methods are not always adequate for diagnosing PRAA and differentiating from megaesophagus or other respiratory diseases. In addition, these methods cannot directly demonstrate other vascular anomalies. In the present case, ventrodorsal thoracic radiographs did not show curvature of the trachea although there was ventral deviation of the trachea in the right lateral thoracic view. Focal leftward deviation of the trachea in ventrodorsal thoracic radiographs can be helpful in cases of suspected PRAA, however, this sign might be not always reliable in small breed dogs with a narrow thoracic cavity. CT directly provides features of aortic arch abnormally progressed on the right side of the trachea, other vessels constituting VRAs, and accurate location of luminal narrowing to enable differentiate from megaesophagus (4,11,14,23,25).

When a weaned dog repeatedly regurgitates after eating and is suspected to have PRAA, but radiographic evidences are insufficient for diagnosis, CT is recommended for diagnosis and preoperative plans. CT is a reliable diagnostic tool for PRAA that directly identifies the esophageal constriction near to the heart base and vascular malformations (22,25).

Conclusion

This case report described PRAA with an aberrant LSA diagnosed by CT in a toy poodle dog. Post-contrast and reconstructed CT may be useful to diagnose PRAA when conventional radiography or contrast study is not sufficient to

establish a diagnosis.

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