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Case Series

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Isolated unilateral absence of the right pulmonary artery in two cats visualized by computed tomography angiography

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Abstract

Case series summary Two cats were evaluated for progressive exercise intolerance, dyspnea and unilateral infiltrate of the left lung. Computed tomography angiography (CTA) revealed absence of the right pulmonary artery in both cats with systemic arterial collateral vessels perfusing the right segmental pulmonary arteries. In one case, the collateral vessels arose from the esophageal artery, while in the other case they derived off the right costocervical trunk. One cat was diagnosed with pulmonary hypertension and was euthanized owing to progressive respiratory distress despite medical management with sildenafil, pimobendan, clopidogrel and furosemide. The other cat, without echocardiographic evidence of pulmonary hypertension, was successfully managed with furosemide and enalapril for more than 4 years.

Relevance and novel information CTA allowed visualization of a rare congenital heart malformation, unilateral absence of the right pulmonary artery, in two cats and accurately characterized the source of collateral blood supply to the affected lung. Severe pulmonary hypertension may be a negative prognostic factor in cats with this condition as medical therapy in the cat without evidence of pulmonary hypertension resolved clinical signs, while the cat with severe pulmonary hypertension died from the disease.

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Case series description

Case 1

A 5-year-old, spayed, female, domestic shorthair cat (cat 1) presented to The Ohio State University's Veterinary Medical Center for evaluation of acute disorientation, tachypnea, lethargy and anorexia. A grade III/VI systolic right parasternal heart murmur and focal crackles in the ventral left thorax were noted on auscultation. Complete blood count, serum chemistry, urinalysis and blood pressure were within reference intervals. Heartworm antigen, antibody and D-dimers were all negative. Thoracic radiographs (Figure 1) revealed moderate cardiomegaly (vertebral heart size of 8.8), a severe unstructured interstitial pattern throughout the left lung lobes, leftward shift of the mediastinum with hyperinflation of the right lung lobes and a mild unstructured interstitial pattern within the right caudal lung lobe. The left caudal lobar pulmonary artery and vein were severely dilated.

On transthoracic echocardiography (TTE), trace pericardial effusion was noted with, subjectively, moderate dilation of the right atrium and right ventricle. The pulmonary trunk (PT) and left pulmonary artery (LPA) were severely dilated with a pulmonary valve annulus of 12.4 mm compared with an aortic valve annulus of 8.5 mm, and the right pulmonary artery (RPA) was not

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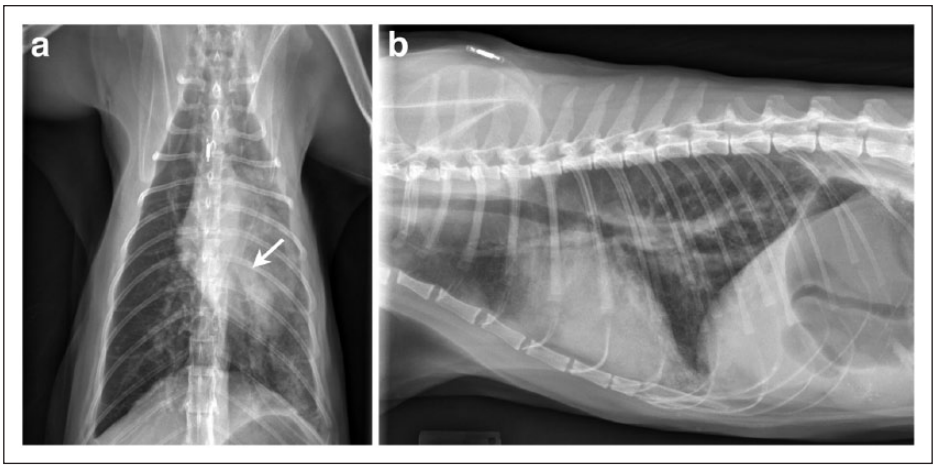

Figure 1 (a) Ventrodorsal and (b) lateral thoracic radiographs from cat 1, with unilateral absence of the right pulmonary artery. An unstructured interstitial pattern is apparent in the left lung lobes with marked dilation of the left caudal lobar pulmonary artery (arrow) and vein. There is a leftward mediastinal shift present. Moderate cardiomegaly is apparent with a gas-distended stomach.

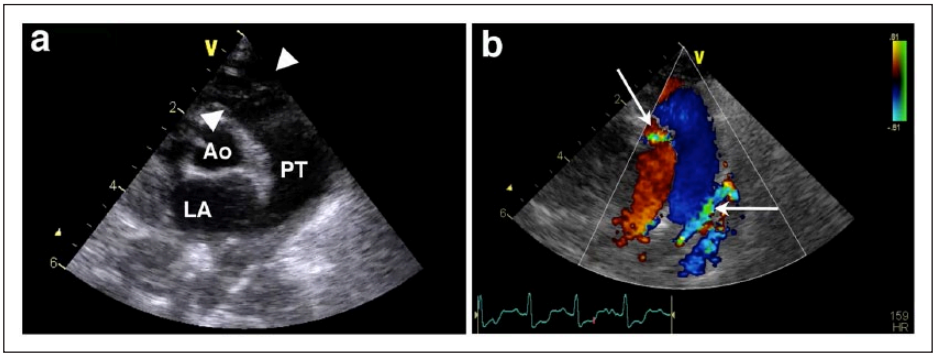

Figure 2 Echocardiographic images from cat 1 with unilateral absence of the right pulmonary artery. (a) A two-dimensional image at the heart base showing the aorta (Ao), left atrium (LA) and pulmonary trunk (PT). The pulmonary valve annulus is denoted by the arrowheads. (b) A similar image to that in (a) is shown with color Doppler imaging demonstrating the turbulent, continuous flow of aberrant vessels around the heart base (arrows).

visualized. Several tortuous vessels with continuous, turbulent flow on color and spectral Doppler imaging were noted at the heart base (Figure 2). Moderate tricuspid regurgitation with a peak velocity of 4.6 m/s and no evidence of pulmonary valve stenosis predicted a systolic right ventricular systolic pressure of at least 90 mmHg. Differential diagnoses included pulmonary hypertension (PH) secondary to extra-cardiac left-to-right shunt flow, peripheral pulmonary arterial stenosis, pulmonary artery dissection, congenital absence of the RPA, heartworm disease or thromboembolic disease.

Thoracic computed tomography angiography (CTA) images (Figure 3) were acquired with an eight-slice CT scanner (GE LightSpeed Ultra 8 slice) using the following parameters: 120 kVP, 49 mA, 1.25 mm slice thickness, spiral pitch of 1.35 and 0.6 s/rotation. Iodinated contrast medium (240 mg iohexol/ml; Omnipaque) was injected as a single bolus (12 ml; 3 ml/kg IV) into the right cephalic vein using a power injector and post-contrast images were acquired starting 3 s after injection from cranial to caudal, followed by an additional series from caudal to cranial. The CT revealed absence of the RPA and severe dilation of the LPA. The right segmental pulmonary arteries were supplied by tortuous blood vessels within the mediastinum, which were interpreted as bronchial arteries. There were no obvious filling defects within the pulmonary arteries to suggest pulmonary thrombi or heartworm emboli.

The cat was diagnosed with congenital absence of the RPA and prescribed clopidogrel (18.75 mg PO q24h), sildenafil (6.25 mg PO q8h) and pimobendan (1.25 mg PO q12h). On re-evaluation 2 days later after a marked decrease in energy level and mild dyspnea at home, repeat thoracic radiographs demonstrated increased distention

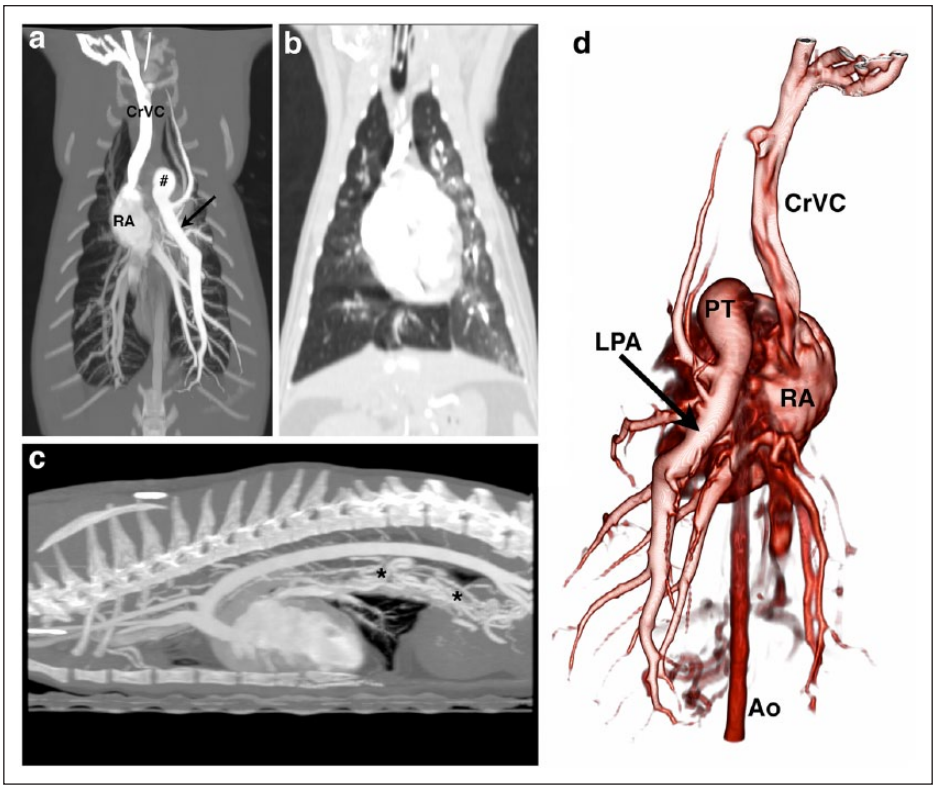

Figure 3 Thoracic CT angiography of cat 1 with unilateral absence of the right pulmonary artery. (a) Dorsal multi-planar reformat showing the single left pulmonary artery (arrow) with absence of a bifurcation at the pulmonary trunk (#). Contrast is seen in the cranial vena cava (CrVC) and right atrium (RA) as well. (b) Dorsal reformat in a lung window showing diffuse interstitial infiltrates in the mid-to-ventral left lung with reduced lung volume compared with the right. (c) Sagittal multi-planar reformat showing multiple tortuous systemic collateral arterial vessels (*) arising from the celiac and esophageal arteries, which course cranially to perfuse the right segmental pulmonary arteries. (d) A volume-rendered three-dimensional reformatted image from a dorsal perspective demonstrates the lack of a right pulmonary artery with the pulmonary trunk (PT) continuing solely as the left pulmonary artery (LPA). Note that the left–right orientation of (c) is opposite to that of (a). Ao = aorta.

of the left caudal lobar pulmonary artery, resolution of the mediastinal shift, an unstructured interstitial pattern throughout all lung lobes most pronounced in the left caudal lung lobe and mild pleural effusion. Right-sided congestive heart failure was presumed from the pleural effusion and furosemide prescribed (10 mg SC q12h). The cat was euthanized without clinical evaluation 8 days later owing to progressive dyspnea and lethargy at home, despite medical management. Autopsy revealed severe cardiomegaly with marked right atrial dilation, mild pericardial effusion and diffuse hyperinflation and locally extensive emphysema of the left lung. The PT was dilated and continued solely as the LPA with absence of the RPA (Figure 4a). Small-caliber arterial varices derived from the esophageal branch of the celiac artery were found as the primary supply blood to the right segmental pulmonary arteries (Figure 4b), rather than hypertrophy of the bronchoesophageal circulation, as initially suspected. Microscopically, there was severe unilateral subacute interstitial pneumonia in the right lung, consistent with decompensated PH. Sections of right lung contained multiple thick-walled and tortuous arterioles, reflecting aortic collateral origin and systemic arterial pressures. The left lung histopathology revealed severe necrotizing interstitial pneumonia, marked alveolar histiocytosis, type II pneumocyte hyperplasia, high protein edema and alveolar emphysema. Microscopically, the myocardium was unremarkable other than rare atrophic myofibers in the right ventricle and moderate, subacute multifocal myofiber degeneration in the right atrium. Liver histopathology revealed moderate, diffuse centrilobular congestion; Kupffer cells contained hemosiderin.

Case 2

A 3-year-old, castrated, male, domestic shorthair cat (cat 2) presented for cardiac evaluation following a 2 month history of tachypnea. On physical examination, a grade III/VI systolic murmur was ausculted over the right parasternal border. Renal values, packed cell volume, total protein and blood pressure were within reference limits. The cat was receiving furosemide (12.5 mg PO, AM; 18.75 mg PO, PM) at the time of initial evaluation.

Prior thoracic radiographs (Figure 5) from cat 2 were reviewed, which showed a normal heart size (vertebral

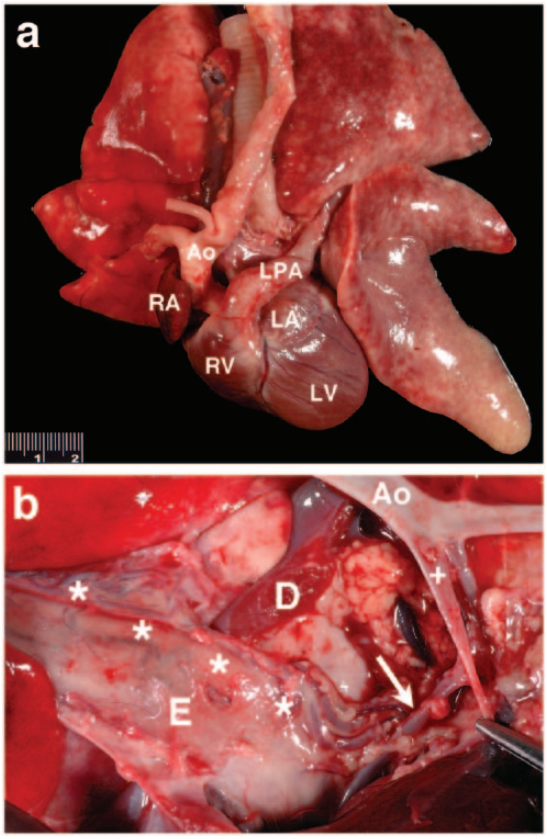

Figure 4 Autopsy images from cat 1 showing unilateral absence of the right pulmonary artery in a 5-year-old cat. (a) The left lung is hyperinflated, while the right lung appears partially collapsed. The right atrium (RA) and right ventricle (RV) are enlarged. The pulmonary trunk continues solely as the left pulmonary artery (LPA); no remnant of right pulmonary artery can be seen. LA = left atrium; LV = left ventricle. (b) The image shows the origin of the systemic arterial collateral vessels supplying the right lung. The aorta (Ao) gives rise to the celiac artery (+), from which derives the left gastric artery (arrow) and many tortuous varices (*) tracking along the esophagus (E). Cranial is to the left of the image. D = diaphragm.

heart size of 7.3), enlarged left caudal lobar pulmonary artery and vein, a mediastinal shift to the right, and slight interstitial opacity throughout the left lung lobes. On TTE, the PT and LPA were visualized, but two-dimensional and color Doppler evidence of a RPA could not be observed. Pulmonary hypertension could not be documented, as tricuspid regurgitation and pulmonary insufficiency were not apparent, and the right ventricle was read as equivocal concentric hypertrophy with a normal right atrium. Mild concentric left ventricular hypertrophy was present and interpreted as pseudohypertrophy from mild hypovolemia.

Thoracic CTA images (Figure 6) were acquired similarly to cat 1. CTA revealed absence of the RPA with a dilated LPA and associated vasculature. A tortuous systemic arterial collateral vessel arose from the right costocervical trunk and extended caudally to anastamose with the right segmental pulmonary arteries. The parenchymal changes noted in the left lung on thoracic radiographs had resolved following diuretic therapy administered for 2 weeks prior to CT evaluation. Based on echocardiographic and CT findings, the cat was diagnosed with congenital absence of the RPA. The cat was prescribed enalapril (1.25 mg PO q12h) in addition to the previously prescribed furosemide. At the time of writing, 4 years have passed and the cat has remained asymptomatic, without recurrence of pulmonary infiltrates. The cat has continued to receive furosemide and enalapril as previously prescribed. On follow-up TTE performed annually since CTA, unchanged mild thickening of the right ventricular free wall and interventricular septum with absence of any appreciable tricuspid or pulmonary insufficiency have been observed. As such, the presence or absence of PH cannot be confirmed, though the lack of remodeling suggests no or only mild PH.

Discussion

Unilateral absence of a pulmonary artery (UAPA) is a rare congenital cardiovascular anomaly that was first described in humans in 1868,[1] and in the veterinary literature in 1986.[2] In 2011, a review of the human medical literature revealed 419 cases worldwide.[3] There is one documented case of UAPA reported in the veterinary literature, in a 2-year-old, Siamese cat.[2] Additionally, this condition has been suggested in two feline cases of pulmonary artery stenosis.[4,5] The images presented here represent veterinary examples of CTA for the diagnosis of UAPA and highlight additional information gained from this modality, including collateral circulation, presence of segmental pulmonary arteries and lung characteristics.

In humans, UAPA is commonly accompanied by concurrent cardiovascular malformations including tetralogy of Fallot, atrial septal defect, coarctation of the aorta, right aortic arch, truncus arteriosus, patent ductus arteriosus and pulmonary atresia.[6,7] When no concurrent heart defects are apparent, the preferred nomenclature is isolated UAPA.[6,8] Left-sided absence is frequently associated with other cardiac malformations, while right-sided absence predominates in isolated cases.[3,6,7] The absent pulmonary artery in isolated UAPA is almost always contralateral to the aortic arch (eg, absence of the RPA with a left-sided aortic arch).[6]

During mammalian embryogenesis, the distal intrapulmonary pulmonary arteries develop from their respective lung buds and join the proximal portion of the right and left sixth aortic arch, which then develop into the left and right extrapulmonary pulmonary arteries. The PT arises from the truncoaortic sac, while the distal right and left sixth aortic arches form the right and left ducti arteriosi. The predominant theory for the development of UAPA describes anomalous involution of the proximal

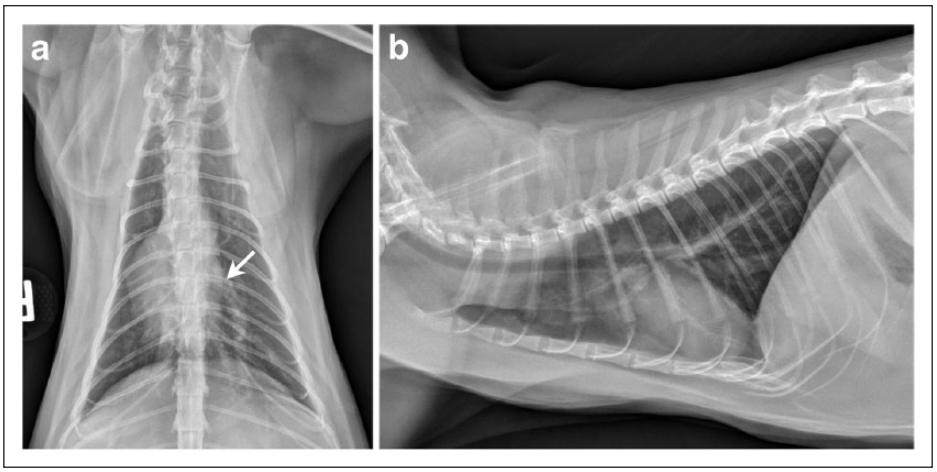

Figure 5 (a) Ventrodorsal and (b) lateral thoracic radiographs from cat 2 with unilateral absence of the right pulmonary artery. Enlargement of the left caudal lobar pulmonary artery (arrow) and vein is apparent with a mediastinal shift to the right and slight interstitial opacity throughout the left lung lobes.

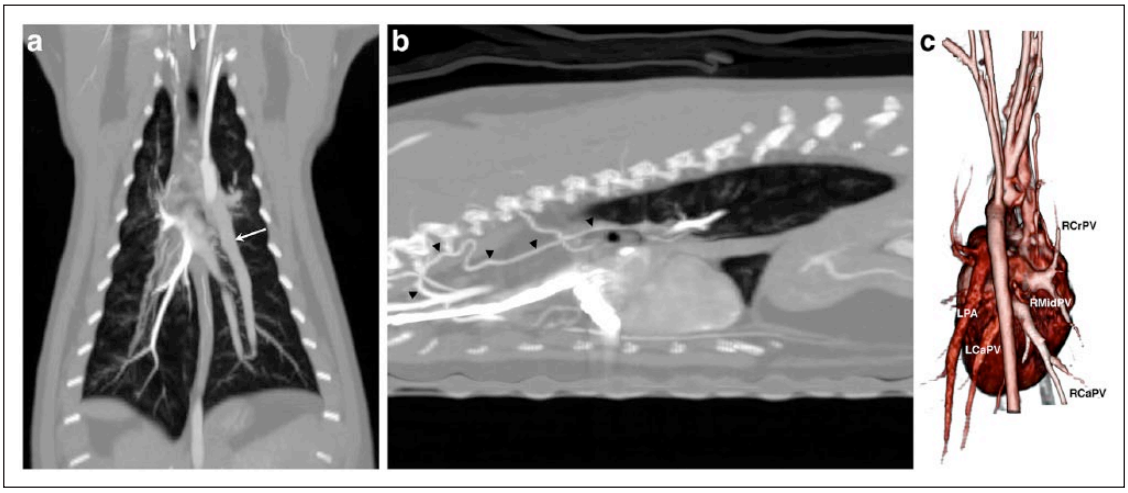

Figure 6 Thoracic CT angiography from cat 2 with unilateral absence of the right pulmonary artery. (a) Dorsal multi-planar reformat showing prominent left caudal lobar pulmonary artery (arrow) and vein, with diminutive right caudal lobar pulmonary artery and vein. (b) A sagittal multi-planar reformat showing the origin and course of the systemic arterial collateral vessel (arrowheads) arising from the right costocervical trunk and coursing caudally to perfuse the right segmental pulmonary arteries. (c) A volume-rendered three-dimensional reformatted image from a dorsal perspective demonstrates the lack of a right pulmonary artery with the pulmonary trunk continuing solely as the left pulmonary artery (LPA). Note that the left–right orientation of Figure 6c is opposite that of Figure 6a. RCrPV = right cranial pulmonary vein; RMidPV = right middle pulmonary vein; RCaPV = right caudal pulmonary vein; LCaPV = left caudal pulmonary vein

sixth aortic arch, while the connection between the intrapulmonary pulmonary arteries and the distal sixth aortic arch persists. The ipsilateral intrapulmonary arteries lose their blood supply once the ipsilateral ductus arteriosus closes. The corresponding pulmonary tissues will become hypoxic and ischemic, prompting growth of anomalous collateral vessels to perfuse the affected lung lobes and predisposing these lungs to infection.[7,9]

Presenting complaints in people with UAPA are variable with recurrent respiratory tract infections, exercise intolerance and mild dyspnea described most frequently; hemoptysis occurs less commonly.[6] Pulmonary hypertension is present in 20–25% of cases.[6] The age at diagnosis in people is variable, with a median of 14 years, ranging from infancy to 58 years.[6] Infants may present in respiratory distress, with cyanosis, growth delay, severe PH and congestive heart failure.[7,10] Adults may be asymptomatic; the disease incidentally diagnosed once thoracic radiographs are performed for other reasons.[6] It has been speculated that the diagnosis can be delayed in

humans owing to non-specific presenting signs or because diagnostics are pursued only once respiratory infection or hemoptysis manifest.[6]

In humans, thoracic radiographs may reveal asymmetric lung lobe size with a deviated mediastinum toward the affected hemithorax and contralateral lung hyperinflation. The affected lung field is often reduced in size, hyperlucent, with absent or decreased pulmonary vascular markings, smaller intercostal spaces and an elevated hemidiaphragm.[9,11] In cat 1, the mediastinal shift away from the affected lung (toward the left) was in contrast to the expected findings of a shift to the right. The reason for this is unclear, but may relate to the infiltrate in the left lung lobes of this cat and compensatory hyperinflation of the better aerated right lung lobes. Alternatively, intralobar pulmonary sequestration can present with emphysematous changes; pulmonary sequestration is defined as a portion of lung that does not connect to the tracheobronchial tree and has a systemic arterial supply.[12] As this cat did have a systemic arterial supply to the right lung, it is possible that a portion of the right lung represented pulmonary sequestration with emphysematous change; however, there was no evidence of pulmonary sequestration or right-sided emphysema noted on post-mortem evaluation. The mediastinal shift had resolved on the follow-up radiographs, but the paradox of emphysematous and hyperinflated left lung lobes on post mortem stands in contrast to the apparent hypoinflation of the left lung on radiographs. An explanation for this discrepancy is challenging, though it is possible that the unilateral emphysema of the left lung resulted in air-trapping and a more prominent size relative to the non-emphysematous right lung at autopsy. The term interstitial pneumonia as noted on this cat's lung histopathology reflects inflammation involving the alveolar or interlobular septa and does not imply an infectious etiology. The radiographs from cat 2 were consistent with human reports showing a shift toward the affected (right) lung. A definitive diagnosis in humans can be obtained by performing CT or MRI, which can reveal mosaic parenchymal changes, patent segmental pulmonary arteries with variable systemic collateral arterial circulation, and bronchiectasis secondary to recurrent respiratory tract infections.[11,13,14]

In humans, the anomalous systemic collateral vessels primarily arise from not only the bronchial arteries, but have also been documented to develop from the intercostal, subdiaphragmatic, subclavian and coronary arteries.[6,15,16] Transthoracic echocardiography is used to confirm the diagnosis, rule out other cardiac or vascular anomalies, and estimate pulmonary arterial pressure.[10] Echocardiography may reveal a dilated unilateral pulmonary artery, absence of the contralateral pulmonary artery, PH and right ventricular hypertrophy.[7,10,14] While angiography remains the historical gold standard for diagnosis in humans, it is reserved for patients undergoing embolization or revascularization surgery.[14]

The previously reported case of UAPA in a cat was similar to the two cases described here; absence of the RPA was documented by non-selective angiography and autopsy evaluation in a 2-year-old Siamese with respiratory distress.[2] In the other two feline reports that relate to this condition, flow was not apparent in the LPA during angiography and constriction at the origin of the RPA was additionally noted.[4,5] While similar to UAPA, it is our opinion that these cases represent pulmonary artery stenosis type IIA,[17] or juxtaductal constrictions, because of the ostial narrowing at the RPA and post-mortem image showing presence of an occluded LPA from the PT to the left lung.[4]

The previously reported Siamese case also demonstrated unilateral interstitial infiltrate in the left hemithorax,[2] as in the two cases presented here. The pulmonary infiltrates are of uncertain etiology; pulmonary edema related to excess blood flow (total cardiac output traversing half the normal pulmonary vasculature) is considered given the response to diuretic therapy. Respiratory infection, as occurs in humans with this condition, is considered less likely given resolution without antibiotic therapy. Angiocardiography has previously characterized the anomaly,[2] though CT appears to be effective for accurate morphologic assessment of the condition, while also allowing improved visualization of the lung parenchyma and systemic collateral vasculature as seen in the two cats described herein.

While there has been no consensus on treatment in humans, therapies are typically reserved for those experiencing massive hemoptysis, recurrent lower respiratory tract infections and PH.[7] Treatment options in humans have included partial or total pneumonectomy, selective embolization of collateral vessels, ductal stenting and systemic-to-pulmonary artery grafting.[7] Pulmonary hypertension in people is treated medically using endothelin receptor antagonists, prostacyclins, phosphodiesterase inhibitors or calcium channel blockers.[6,7] Treatment was attempted for the PH in cat 1, but did not provide sustained palliation. Cat 2 was diagnosed with right-sided UAPA without evidence of PH, though mild PH could not be excluded, and has been successfully managed using oral furosemide and angiotensin-converting enzyme inhibition. Based on these two cases and the human condition, moderate-to-severe PH may be a negative prognostic factor in cats with UAPA.

Conclusions

Right-sided UAPA was diagnosed by CTA in two cats, allowing accurate characterization of the arterial malformation, as well as determination of the collateral blood supply to the affected lung. The condition may be

suspected by thoracic radiographs and echocardiography in cats having respiratory signs, unilateral pulmonary infiltrates and dilated lobar pulmonary vessels; confirmation of the diagnosis is provided by CTA.

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