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REVIEW ARTICLE

The red connection: a review of aortic and arterial fistulae with an emphasis on CT findings

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Abstract Fistulae between the aorta and adjacent structures are a rare, emergent, and potentially life-threatening process. Most commonly, aortic fistulae arise secondarily as a complication of prior aortic surgery with fistulization to adjacent structures. Rarely, a primary fistula may arise from the aorta in the setting of a pre-existing aneurysm or from a mass, inflammation, or infection. Although the incidence of aortic fistulae remains low, the frequency continues to increase as aortic surgical interventions and post-surgical follow-up with imaging become more common. Computed tomography (CT) is the modality of choice in evaluating the patient with suspected aortic fistula because of its accessibility and short scan time. In addition, CT allows for more clear depiction of para-aortic or intra-aortic gas than ultrasound or magnetic resonance (MR). This gas may be the first clue of a fistula. Given the high mortality associated with aortic fistulae, familiarity with the imaging findings of the spectrum of aortic fistulae is essential knowledge in the emergency setting. This review will discuss the imaging appearance of aortic and arterial fistulae to the bronchi, esophagus, gastrointestinal tract, ureters, and veins on CT.

Keywords Aorta · Fistula · Aortoenteric fistula · Aortobronchial fistula · Aortoesophageal fistula · Arterioureteral fistula · Aortic aneurysm

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Introduction

The long course and high intraluminal pressure of the aorta allow pathologic processes of the aorta and its major branches to potentially fistulize to adjacent structures [1, 2]. Aortic fistulae may be primary or secondary. Primary aortic fistulae most often arise due to aneurysmal disease, although other causes including inflammation, infection, foreign body, or tumor involving neighboring structures may lead to fistula formation. Secondary fistulae are those that arise as a complication of aortic surgery and are, overall, much more common than primary aortic fistulae. Although aortic fistulae may be clinically occult, patients can present with hematemesis, melena, hemoptysis, hematuria, abdominal pain, or chest pain; the specific presenting symptoms are dependent on the structure involved with the fistula (Table 1). These fistulae can be life threatening given the potential for exsanguination. Therefore, it is essential that the emergency radiologist be familiar with these conditions as imaging plays a key role in diagnosis and facilitates prompt treatment. Furthermore, imaging is frequently performed for a non-specific clinical presentation where there is little or no pre-test suspicion of an aortic or arterial fistula and the radiologist may be the initial physician to raise awareness of this diagnosis. As active contrast extravasation from the aorta to the fistulized structure is an exceptional feature usually seen with ongoing exsanguination, this review will focus on the more common, but also more subtle, secondary imaging features of aortic fistulae.

Imaging

Computed tomography (CT) is the preferred imaging modality for diagnosing aortic fistulae given its image acquisition speed and availability in the emergency setting. In addition, CT allows for detection of subtle foci of gas which can be a

Table 1 Typical clinical features and CT findings for the various aortic and arterial fistulae

		Aortobronchial	Aortoesophageal	Aortoenteric	Aortovenous	Arterioureteral
Clinical features		Hemoptysis	Hematemesis Dysphagia Chest pain	GI bleeding	Shock Congestive heart failure Pelvic, leg venous congestion Continuous abdominal bruit	Abdominal/flank pain Gross hematuria Hypotension and shock History of radiation or abdominal surgery
CT Findings	Common	Loss of fat plane between aorta and bronchus Irregular outpouching of aorta	Loss of fat plane between aorta and esophagus Irregular outpouching of aorta	Loss of fat plane between aorta and bowel wall Disruption of aortic wall or pseudoaneurysm	Loss of fat plane between aorta and adjacent vein	Loss of fat plane between ureter and common iliac artery Contour irregularity of artery at level of ureter
	More specific	Ground glass opacities around the affected bronchus Tree in bud nodules secondary to aspirated blood Debris within the affected bronchus Pneumothorax	Active extravasation of contrast into esophageal lumen Clot or contrast within the distal esophagus/stomach	Extravasation into bowel or periaortic space Perigraft gas >4 weeks after repair of ruptured AAA Perigraft fluid, edema >3 months after elective AAA repair Focal bowel wall thickening in loops filled with gas or fluid	Extravasation of contrast into vein Dilated distal vein filled with arterial phase contrast Surrounding hematoma	Hydroureter Hydronephrosis

key finding in suggesting an aortic fistula. A CT protocol specific for aortic fistulae is difficult to implement as clinical suspicion may not be high prior to imaging. Nevertheless, multiphase CT angiography is preferred when feasible. Oral contrast should be avoided as it may obscure subtle contrast extravasation into the bowel lumen. Obtaining a non-contrast series can be helpful to differentiate contrast extravasation from high-attenuation material in the bowel lumen, surgical material, or vascular calcifications. Post-contrast images should be obtained in the arterial phase for optimal opacification of the aorta [3]. Delayed images can be helpful for detecting flow in the fistulous tract or accumulation of contrast within connecting structures [4]. At our institution, CT evaluation of the aorta consists of unenhanced scanning with 5-mm slice thickness followed by arterial-phase CT performed with bolus tracking and 1-mm slice thickness. When there is specific suspicion of an aortic fistula or the patient has an aortic graft, 80-s delayed imaging is also obtained [5].

Magnetic resonance (MR) is an alternative modality, although given the acuity of this indication, MR availability, exam length, and the requirement for patient cooperation and repeated breath-holds limit the use of MRI in evaluating aortic fistulae. In a study from 1987 where MR was performed for follow-up of surgically treated aortic pathology, indirect signs of an aortoenteric fistula were present in one of two cases [6]. If modern MR equipment is used for evaluation of a patient with a suspected aortic fistula, the diagnosis will most likely be evident on pre- and post-contrast dynamic T1-weighted imaging using principles similar to multiphase CT imaging [7, 8]. If available, time-resolved magnetic resonance angiography may also be helpful for showing flow between the aorta and adjacent structures [8].

Other imaging modalities that can be used to detect and characterize aortic fistulae include catheter angiography and fluoroscopic studies with water-soluble contrast [9]. However, these modalities are not generally as sensitive, nor as practical as CT when evaluating for aortic fistulae in the emergency setting. Ultrasound is rarely indicated for the diagnosis of aortoenteric fistulae given user variability. Additionally, visualization of the aorta can be difficult due to bowel gas or in patients with large body habitus. Although not indicated to evaluate for fistulae, nuclear medicine scintigraphy using Indium-111-labeled white blood cells can assist in the detection of perigraft infection.

Aortobronchial fistulae

An aortobronchial fistula represents a communication between the aorta and the airway, typically between the membranous wall of the left mainstem bronchus and the descending aorta given the close proximity of these two structures. The patient can present with hemoptysis with a range of severity from low to high volume that may be life threatening.

Special attention to a history of thoracic aortic disease or prior surgery may be a clue to an underlying aortobronchial fistula and prompt a clinician to request a CT. Overall, CT has shown to be efficacious in the detection of the cause of hemoptysis [10]. Bypassing CT to go straight to bronchoscopy may localize bleeding, but fail to diagnose the cause [11]. Risk factors for the development of an aortobronchial fistula include thoracic aortic aneurysm, treated or untreated, and prior cardiac or other thoracic surgery [12]. In addition, mediastinal inflammatory processes as well as primary pulmonary, bronchial, or metastatic neoplasms may be causative [13, 14].

In the setting of an aortobronchial fistula, the chest radiograph can suggest aortic enlargement or can demonstrate airspace disease adjacent to the aorta. On CT, findings of an aortobronchial fistula include an irregular aortic outpouching and loss of the normal fat plane between the aorta and airway (Fig. 1). Debris, typically high in attenuation reflecting hemorrhage, or rarely active extravasation of contrast, may be seen within the airway. Prior to the development of a frank communication, fat stranding and inflammation in the intervening space separating the aorta and bronchus may be an early finding. Pulmonary findings of ground glass and tree-in-bud nodules represent the downstream aspirated hemorrhage. Pneumothorax may also be present if the fistula is more complex and involves a communication with the pleural cavity.

It is important to recognize that patients with an aortobronchial fistula often have intermittent bleeding, and the frank communication between the aorta and bronchus is often not identified at the time of imaging. Therefore, utilization of signs such as direct abutment of the aorta and bronchus, surrounding inflammation, or evidence of associated aspirated blood products can be used to suggest the diagnosis, even if the communication is not directly seen. A high index of suspicion is paramount, as even with optimal imaging, aortobronchial fistulae are occasionally discovered only at surgery or worse, at autopsy where it is discovered greater than 30 % of the time [15]. Treatment of aortobronchial fistula is increasingly being performed with endovascular aortic stent graft insertion [16].

Aortoenteric fistulae

Aortoenteric fistulae classically present with abdominal pain, gastrointestinal bleeding, and a pulsatile abdominal mass, although this is an uncommon presentation. Primary aortoenteric fistulae arise most commonly from aortic aneurysms but may be seen with infection, malignancy, foreign bodies, radiation, or inflammatory processes [17]. Secondary aortoenteric fistulae, arising after abdominal aortic aneurysm (AAA) repair, are approximately ten times more common than primary fistulae and are more commonly associated with open repairs. The most common location of secondary fistula formation is at the superior mesenteric artery angle with a

Fig. 1 Sagittal CT image demonstrating effacement of the plane between the left mainstem bronchus and thoracic aorta (*arrow*) with a focal contour deformity of the aortic wall. Of note, this is not a ductus diverticulum as it arises distal to the left subclavian artery origin. This is a case of aortobronchial fistula

communication forming between the aorta and the transverse duodenum. Secondary fistulae may be related to chronic low-grade infection of the aortic graft and repetitive pressure on the intestine from aortic pulsations [18, 19]. In patients undergoing routine post-operative imaging for monitoring of AAA repair, the diagnosis may be suggested prior to development of clinical symptoms [20].

CT findings of aortoenteric fistulae include perigraft gas persisting greater than 4 weeks post-operatively, aortic wall disruption, gas within the aortic lumen, or contrast extravasation into the bowel lumen [3, 18, 21]. If the study is performed with oral contrast, leak of contrast from the bowel into the periaortic space would also suggest the presence of an aortoenteric fistula [22]. Findings that may be seen in both perigraft infection and aortoenteric fistulae include loss of the fat plane between the aorta and bowel (Fig. 2), focal bowel wall thickening, bowel tethering to the aorta, perigraft fluid or soft tissue thickening greater than 3 months after repair, or aortic pseudoaneurysm [23]. Downstream intraluminal clot may be identified as a secondary sign.

Aortoenteric fistulae remain a lethal condition with mortality rates up to 44 % even with prompt open repair [24]. The treatment is usually resection of the infected/fistulized graft along with extra-anatomic bypass; without treatment, mortality is essentially 100 % [25]. Many patients with an aortoenteric fistula present with an episode of self-limited hemorrhage known as a “sentinel” or “herald” bleed. These self-limited bleeds serve as a clinical warning sign of more life-threatening hemorrhage and afford time for imaging, localization, and surgical planning.

Aortoesophageal fistulae

Aortoesophageal fistulae, a subset of aortoenteric fistulae, have unique clinical and imaging features. Aortoesophageal fistulae can present with upper gastrointestinal bleeding and

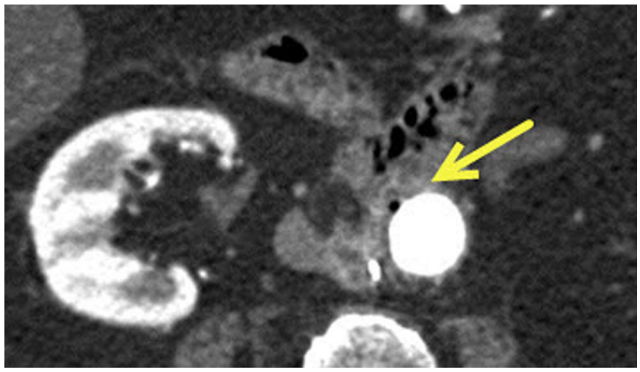

Fig. 2 Transverse CT image showing effacement of the fat plane between the aorta and duodenum with air in the aortic wall (*arrow*), a classic appearance of an aortoenteric fistula

occasionally frank hematemesis. The presence of bright red arterial blood may be helpful clinically to distinguish an aortoesophageal fistula from esophageal variceal or gastric bleeding, both of which tend to be darker. Although the classic Chiari triad of a sentinel bleed, chest pain, and an asymptomatic interval followed by massive fatal hematemesis was initially described for aortoesophageal fistulae secondary to foreign body ingestions, other etiologies may display some of these clinical features [2, 26]. Diagnosis before the final exsanguinating event may be achieved with imaging. Unlike most other aortic fistulae, aortoesophageal fistulae are more commonly primary with thoracic aortic aneurysms being the leading etiology followed by esophageal foreign bodies and esophageal malignancy [2]. The descending aorta is the most common location of fistulization with the esophagus given the proximity of the two structures.

CT findings of an aortoesophageal fistula include an irregular outpouching of the aorta and effacement of the fat plane between the aorta and esophagus particularly at a site of an aortic aneurysm [27]. Other signs include gas in or around the aorta and rarely extension of oral or arterial contrast between the two structures (Fig. 3). Anterior displacement of the esophagus by an aortic aneurysm on barium esophagram has been described [2, 28] as well. Downstream high-attenuation material or contrast in the distal esophagus or stomach representing clot or rarely extravasated contrast may also be seen [29]. An unenhanced scan not only allows detection of confounding calcification but also allows for improved detection of foreign bodies which may prove an invaluable clue to the diagnosis [30]. Delayed enhanced CT will not only improve sensitivity for extravasation but will also improve sensitivity for detection of esophageal malignancy as a possible etiology [31].

The treatment of aortoesophageal fistulae involves a combination of endovascular and surgical techniques, and mortality is high [32]. When pre-test suspicion is high, one should consider CT tailored toward the diagnosis of an aortic fistula to facilitate diagnosis and expedite the treatment of this deadly condition.

Fig. 3 Sagittal CT image demonstrating a direct communication between the retrograde filling false lumen of a descending thoracic aortic dissection and the esophagus with extravasated contrast entering the esophageal lumen (*arrow*). An endovascular stent is seen in the true lumen. This is a surgically proven case of aortoesophageal fistula

Aortovenous fistulae

Aortovenous fistulae most commonly arise as a result of rupture of an aortic aneurysm into an adjacent venous structure. Other causes, including trauma, are also possible. The most common location is the inferior vena cava followed by the iliac veins and left renal vein. When the left renal vein is involved, it is typically a retroaortic left renal vein [33]. Clinically, the patient may present with congestive heart failure due to high-output left to right shunting, a pulsatile abdominal mass, abdominal or back pain, shock, pelvic and leg venous congestion, or arterial insufficiency [34]. If the left renal vein is the site of fistula, acute kidney injury may result. A suggestive physical exam finding is the continuous abdominal bruit.

CT imaging findings of aortovenous fistulae are loss of the fat plane between the aorta and suspected vein, dilation of the outflow vein, early venous filling with contrast, and perivascular hematoma (Fig. 4). In patients with aneurysms, close evaluation of the margins of the aneurysm where it is adjacent to other vascular structures is warranted. In the pelvis, where there is a contralateral comparison, asymmetric vascular contrast opacification may be a helpful finding. Ultrasound is a useful diagnostic test in this context and may show an arterialized venous waveform and low-resistance arterial waveform just proximal to or at the site of the fistula. In addition, tissue vibration and turbulent vascular flow may be seen.

Treatments include open repair and endovascular methods in patients with conducive iliac and femoral artery anatomy. Aortovenous fistulae may be only detected at the time of

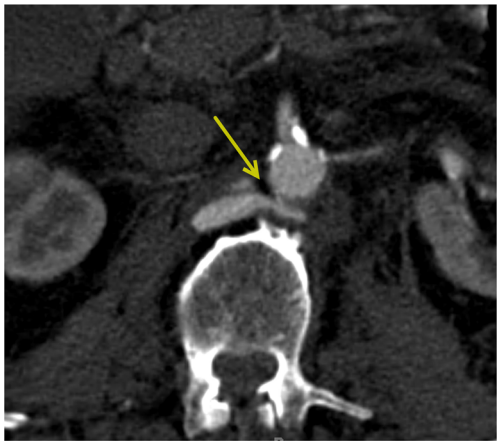

Fig. 4 Transverse CT image showing loss of intervening fat plane and a direct communication (*arrow*) between the descending abdominal aorta and a retroperitoneal vein due to an aortovenous fistula

surgery and, if not recognized, often lead to worse outcomes [34]. With the trend toward endovascular repair methods for aneurysm treatment, endovascular repair with stent graft insertion is becoming more common [35].

Arterioureteral fistulae

Arterioureteral fistulae are abnormal communications between arterial structures and a ureter and differ from the other entities discussed as the process leading to fistulization more commonly arises secondarily from the ureter rather than primarily from aneurysmal vascular disease. Arterioureteral fistulae arise most frequently at the common iliac arteries but may arise anywhere along the course of the ureters [36]. Patients with a history of extensive pelvic surgery or radiation presenting with flank pain, gross hematuria, and shock should raise suspicion for an arterioureteral fistula. In patients with ureteral stents, an arterioureteral fistula may be unmasked during stent exchange [37]. As the diagnosis may be intraprocedural, many patients are evaluated and treated in the intervention suite or operating room before further diagnostic imaging is performed.

In an unsuspected arterioureteral fistula, CT may be employed for evaluation of hematuria as the hematuria may be intermittent. In a review of published cases, CT was helpful in less than half of cases although it was similar in utility to ureteroscopy and ureterography. Angiography was the most helpful pre-operative test [38]. When performed, routine portal venous phase contrast-enhanced CT is most frequently done; excretory phase CT may also be helpful. The CT findings include loss of the fat plane between the ureter and an adjacent artery and a focal contour abnormality of the artery at the level of the ureter (Fig. 5) [39]. Hydronephrosis, hydroureter, and clot in the renal pelvis are secondary signs which should raise suspicion of an arterioureteral fistula, but are not specific and seen far more commonly in alternate

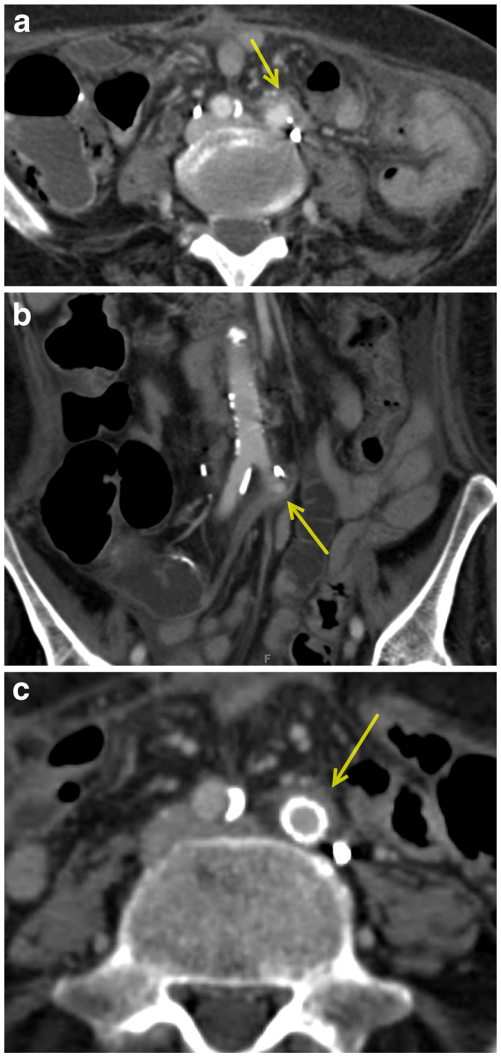

Fig. 5 A patient with an ileal conduit and bright red hematuria underwent CT for further evaluation. **a** Transverse CT image showing a mildly dilated left ureter crossing the left common iliac artery (*arrow*) with effacement of the intervening fat plane. **b** Oblique coronal CT image showing an outpouching of the left common iliac artery where it is crossed by the ureter (*arrow*) which continues into the ileal conduit. **c** Transverse CT image after intervention showing a covered stent in the left common iliac artery and resolution of the arterial outpouching and ureteral dilation

diagnoses. Excretory phase CT may demonstrate ureteral irregularity or ureteral filling defects. A direct communication or extravasation on CT may conceivably be seen although to our knowledge have not been reported.

Treatment for arterioureteral fistulae is challenging, and a multidisciplinary approach involving interventional radiology, vascular surgery, and urology is generally needed. A plan for treatment of both the arterial and ureteral components of the fistula is required. A covered arterial stent graft is frequently used to occlude the fistula without directly treating the ureter [39]. Given the spectrum of native anatomy, prior surgery, or radiation, one must consider a wide range of techniques from embolization to ligation and extra-anatomic

Fig. 6 Transverse CT image demonstrating air in the aneurysm sac (*arrow*) greater than 3 months after a stent graft repair; this graft infection was surgically proved and should not be mistaken for an aortoenteric fistula given the lack of adjacent organ involvement

bypass of the involved artery [40]. An important consideration for ureteral treatment is the need to permanently remove an indwelling stent which may mandate other methods of urinary diversion such as percutaneous nephrostomy tubes [38].

Potential mimics

Aortic and arterial fistulization are rare processes; potential mimics will be frequently encountered. As the use of elective surgical repair of abdominal and thoracic aortic aneurysms increases, perigraft infection, a diagnosis with overlapping imaging findings, is more common than a fistula [21]. It should also be noted that in the immediate post-operative period, perigraft soft tissue thickening and perigraft gas are expected findings and can persist for 3 months and 4 weeks, respectively (Fig. 6) [21]. Recognition of these normal post-operative findings is important as it will prevent incorrectly suggesting that a fistula or infection is present in the early post-operative phase. For the radiologist, distinguishing between aortic fistulae and perigraft infection is difficult. Rather than making a definitive distinction

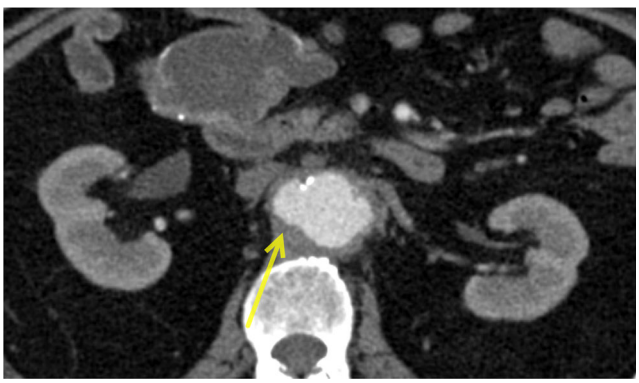

Fig. 7 Transverse CT image showing an outpouching of the right lateral aspect of the abdominal aorta with inflammatory thickening of the aortic wall (*arrow*); this is a surgically proven mycotic aneurysm and should not be confused for a fistula as the inflammation is centered in the aortic wall

Fig. 8 Transverse CT image showing enhancing soft tissue surrounding the right side of the aorta (*arrow*) with tethering of the duodenum and loss of the intervening fat plane; this is biopsy-proven retroperitoneal fibrosis which should be suggested given mesenteric root inflammation. Aortoenteric fistula would be unlikely given this appearance especially in the absence of prior surgery

between these two entities, including both of them in the differential diagnosis should be the goal in imaging as treatments for both are similar. Symptomatology and laboratory features (i.e., anemia, leukocytosis, elevated inflammatory markers) are also useful in distinguishing between these two diagnoses on a clinical level.

Other clinical entities unrelated to recent aortic grafting may mimic aortic fistulae. When the center of the disease process is the aorta or its wall without ancillary findings in adjacent organs, one should consider an infectious or inflammatory aortitis as the most likely diagnosis (Fig. 7). The most common cause of inflammatory aortitis are the large vessel vasculitides Takayasu arteritis and giant cell arteritis; infectious aortitis may occur in the setting of bacteremia from many causes. Esophageal, mediastinal, and retroperitoneal inflammatory conditions may also potentially overlap with aortic fistulae clinically and radiographically [41]. For instance, retroperitoneal fibrosis, which can affect the aorta, duodenum, and other retroperitoneal structures, can obliterate the normal retroperitoneal fat planes and lead to soft tissue thickening around the aorta without aortic fistula formation (Fig. 8). Similarly, Erdheim-Chester disease, an infiltrative histiocytic condition, or lymphoma may both cause a rind of periaortic soft tissue, and imaging features of other organ involvement or lymphadenopathy may alert the radiologist to the alternate diagnosis.

Conclusion

Aortic or arterial fistulization with surrounding thoracic and abdominal structures is a rare but life-threatening condition

that warrants awareness of the CT findings and their significance. Classic clinical presentations are uncommon, although knowledge of symptom patterns may raise awareness before emergent diagnostic imaging is performed. Familiarity with secondary signs of aortic fistulae on CT imaging and potential mimics allows the radiologist to act promptly and appropriately in a condition where expedient intervention is critical to patient survival.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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