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Ortner's syndrome secondary to penetrating aortic ulcer. Case report and literature review

Síndrome de Ortner secundario a úlcera penetrante aórtica. Reporte de un caso y revisión de la literatura

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ABSTRACT

Acute aortic syndrome is defined as an acute process in the aortic wall caused by disruption of the medial layer to a varying degree with the risk of aortic rupture and other complications. A penetrating aortic ulcer is included in the acute aortic syndrome and represents the 2-7% of the acute aortic syndrome presentations. With progression, it leads to intramural hemorrhage, the formation of pseudoaneurysm with great risk of rupture. We present a case of a 65-year-old patient with an atypical presentation of penetrating aortic ulcer in the aortic arch associated with intramural hematoma, pseudoaneurysm and Ortner's syndrome that required thoracic endovascular aortic repair, which presented complication of endoleak type IA that was managed with the strategies "wait and see".

Keywords: acute aortic syndromes, Ortner's syndrome, penetrating aortic ulcer.

RESUMEN

El síndrome aórtico agudo se define como un proceso agudo en la pared aórtica causado por la ruptura de la capa medial en mayor o menor grado, con riesgo de rotura aórtica y otras complicaciones. La úlcera aórtica penetrante se incluye en el síndrome aórtico agudo y representa 2-7% de las presentaciones de este síndrome. La úlcera aórtica penetrante puede ocasionar una hemorragia intramural, la formación de pseudoaneurisma, lo que conlleva a un gran riesgo de ruptura. Presentamos el caso de un paciente de 65 años con una presentación atípica de úlcera aórtica penetrante en el arco aórtico asociada a hematoma intramural, pseudoaneurisma y síndrome de Ortner que requirió reparación aórtica endovascular torácica, la cual presentó como complicación una endofuga tipo IA que se manejó con las estrategias "ver y esperar".

Palabras clave: *síndrome aórtico agudo, síndrome de Ortner, úlcera aórtica penetrante.*

INTRODUCTION

Acute aortic syndrome (AAS) is defined as an acute process in the aortic wall caused by disruption of the medial layer to a varying degree with the risk of aortic rupture and other complications.[1-3]

Penetrating aortic ulcers (PAU) were initially described by Shennan in 1934[4] and after his description the PAU is integrated into AAS. Shumacker and King reported the first operative repair of a ruptured descending aorta secondary to a penetrating aortic ulcer in 1959.[5] The clinical and pathologic entity of penetrating aortic ulcers was established, until 1986

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by Stanson.[6] Since that time, the body of literature on this disease has increased significantly.

PAU fits into a spectrum of AAS consisting of classical dissections, intramural hematoma (IMH), limited dissection and iatrogenic/traumatic transection.[7] PAU may be located in the ascending aorta (type A PAU), in the descending thoracic aorta (type B PAU) or abdominal aorta. Simple isolated PAU may be asymptomatic and incidentally detected through imaging or may present with chest, back or abdominal pain.[8,9] Rarer presentations include Ortner's syndrome (recurrent laryngeal nerve palsy due to aortic pathologies),[10] hemopericardium[11] and hemoptysis.[12,13]

The purpose of this report is present an atypical presentation of PAU in aortic arch associated with pseudoaneurysm and Ortner's syndrome that was repaired by means of thoracic endovascular therapy, which in turn, presented endoleak type IA that was managed conservatively.

CASE REPORT

A 65-year-old patient with history of hypertension, chronic obstructive pulmonary disease, non-insulin dependent diabetes, obesity, hyperlipidemia and past medical history of smoking was referred to our emergency room for evaluation of thoracic pain where was implemented the rapid chest pain protocol. The patient referred a sudden pain in the mid-chest without radiation accompanied by dysphonia began three days prior to arrival and had been progressively worsening. He described the pain as vague in sensation, non-radiating, and associated with dysphonia and denied having fevers, shortness of breath. On examination, the patient was hemodynamically stable, with normal heart rate (88 bpm) and rhythm, no cardiac murmurs, respiratory rate of 18, pulmonary examination without abnormalities, pulse present and normal in upper and lower extremities. The blood pressure 142/90 without difference between left and right arm.

His initial workshop showed a normal cardiac enzyme, dimero-D, and the rest of tests unremarkable. The Chest X-ray reported atherosclerotic thoracic aorta that prompted a computed tomography angiography (CTA) of chest was per the "acute aortic syndrome protocol". A chest computerized tomography (CT) scan showed a PAU 1.2 cm distal to the origin of the left common carotid artery and immediately proximal to the left subclavian artery; also, a large pseudoaneurysmal sac measuring 63 mm by 67 mm, was located on the anterior left side of the aortic arch towards the wall chest *(Figure 1)*.

With the finding in the CTA and the high risk for rupture the Heart Team recommended and an urgent intervention, and because the patient's EuroSCORE was 13, and predictive mortality was 41.12%, and the anatomy was adequate the team decide to perform a thoracic endovascular aortic repair. The team approached the aortic arch pseudoaneurysm repair after achieving general anesthesia, through a left femoral artery approach, an endoprosthesis was introduced under fluoroscopic control and controlled hypotension. An endoprosthesis Valiant™ Thoracic Stent Graft Captiva (Medtronic Italia S.p.A.Via Varesina, 162, 20156 Milano) with a diameter of 26 mm and a length of 100 mm, with an oversizing of 20% was chosen. The endoprosthesis was deployed in such a way that the free flow was on the origin of the left common carotid artery *(Figure 2)*. The procedure was carried out without any significant problems, but the final angiogram showed an evident small type IA endoleak that partially refilled the pseudoaneurysm. Patient's pain resolved soon after placement of the stent graft. A CT scan, performed seven days later, confirmed the presence of a small endoleak with slow pseudoaneurysm refilling. The postoperative period was event free, and the patient was discharged on day eight. A follow-up at one month after the procedure, suggested progressive thrombosis of the pseudoaneurysm sac and, at three months, demonstrated occlusion of the pseudoaneurysm with complete resolution of the endoleak. Six months later, an examination done by our division found the patient to be asymptomatic and a routine CT showed the endoprosthesis positioned correctly and resolution of the endoleak and absence of intramural haematoma due to reabsorption *(Figure 3)*.

DISCUSSION

The AAS are a constellation of life-threatening medical conditions, including classic acute aortic dissection, IMH, PAU, which share common pathophysiological pathways,

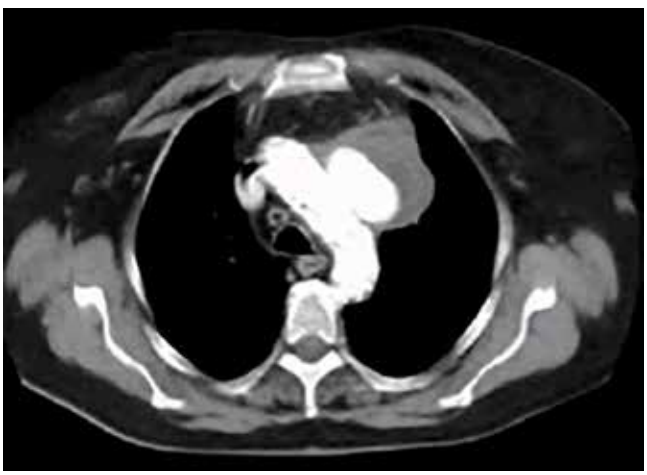

Figure 1: Computer tomography angiography at time of admission showed a penetrating ulcer 1.2 cm distal and pseudoaneurysmal sac measuring 63 × 67 mm, on the anterior left side of the aortic arch towards the wall chest.

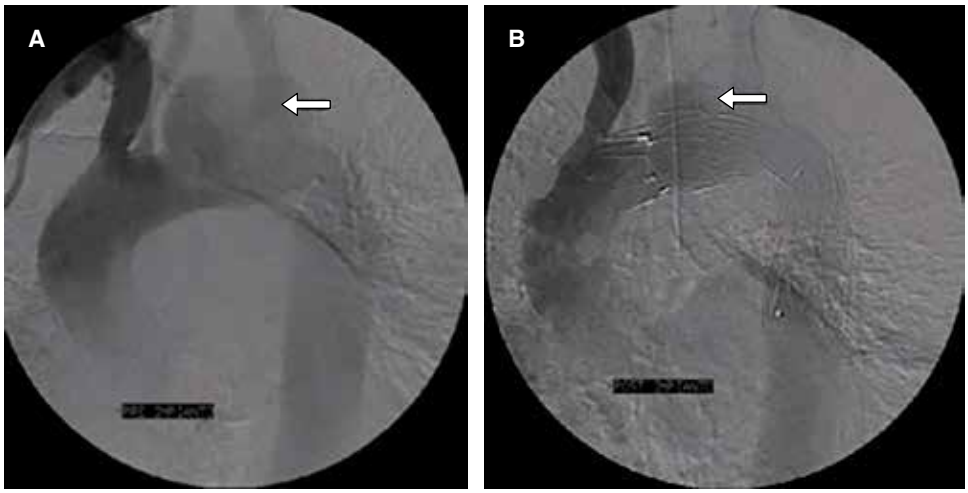

Figure 2:

A) Aortic arch angiogram showing the pseudoaneurysm. **B)** Angiogram post TEVAR deployment demonstrating full coverage of pseudoaneurysm alongside aortic arch visualizing contrast leak inside aneurysm sac.

clinical characteristic, and diagnostic and therapeutic challenges.[14] The incidence of AAS is estimated to be 3.5-6 per 100,000 person-years in the general population and up to 10 per 100,000 person-years in the elderly.[15,16] Isolated PAUs are seen in 2.3 to 7.6% of AAS cases and can be identified in all segments of the aorta however they are most common in the descending thoracic aorta (62%). IMH can coexist with PAU in 45% of cases and may develop into progressive aortic dissection or aneurysm.[17,18] PAU may occur in a solitary location or in multiple segments of the aorta; however, when the ascending aorta is involved, rupture or concomitant IMH are more common.[15,19] In a single-institution review of PAU, the incidence of rupture on presentation was 4.1%, and endovascular or open repair was required in 12.9%. The rupture rate has been reported to be as high as 38% for PAU in an acute presentation, which is considerably higher than that seen for aortic dissection.[20]

Anatomically, there are two commonly used classifications for aortic dissection. The DeBakey system categorizes dissections based on the origin of the intimal tear and the extent of the dissection, and the Stanford system divides dissections according to whether the ascending aorta is involved (type A) or not involved (type B), regardless of the site of origin.[21] Similarly, PAU and IMH can be classified into presence (type A) or absence (type B) of ascending aortic involvement. According with the time course of presentation the AAS is divide into acute (< 14 days), subacute (15-90 days) and chronic (> 90 days) phases.[22] In addition, Svensson sub-classified various types of intimal tears that cause aortic pathologic conditions. Class I-IV intimo-medial defects occur without a known external force, while class V lesions are due to iatrogenic or traumatic causes.[23] PAU is a class II intimo-medial defect and constitutes 2-7% of AAS.

The aorta's microstructure, segmental anatomy, and cardiac impulse hemodynamics determine its function and

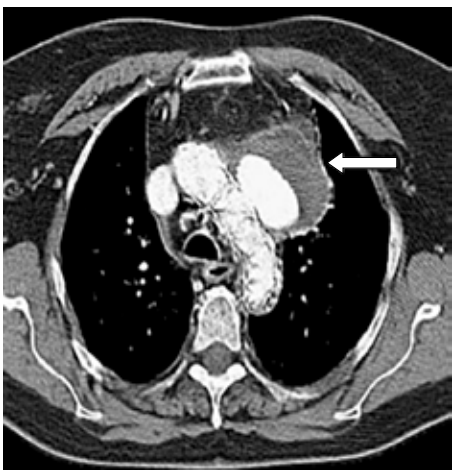

Figure 3:

CTA taken one month after implanting the endoprosthesis with full coverage of PAU, contrast observed inside ulcer corresponding with endo leak type IA.

pathophysiology. The aorta is an elastic artery composed of three inner-to-outer layers –the intima, media, and adventitia– and can be divided into five segments: the root, ascending aorta, arch, descending aorta, and abdominal aorta. Pathologic conditions affecting the aorta are directly influenced by these aortic properties, including its embryologic conotruncal origins.[24] Lesions that involve only the arch or distal aortic segments have a lesser rate of complications in the acute phase but become the dominant concern in the subacute and chronic states.

PAU is an ulcerating lesion most commonly seen in the aortic arch and descending aorta,[25] and affect the intimal layer, with a background of atherosclerotic disease or predisposing factors. It involves an ulcer-like plaque that progressively erodes the intima and burrows through the aortic wall, with remodeling and wall thickening causing a smooth outer bulge. PAU distinguishes itself from common ulcerated atherosclerotic atheroma by burrowing beyond the intima and penetrating the media and its internal elastic lamina, which

separates the two layers. This leads to symptoms and may acutely be associated with a small amount of hemorrhage in the media or contain age-indeterminate thrombus. Given the background of atherosclerotic plaque burden in most affected individuals, the clinical context is important.[25-27] The mechanism of PAU is disruption of the internal elastic lamina than can spread to the media, leading to an IMH, pseudoaneurysm, and if the adventitia is perforated, transmural aortic rupture occurs as the ultimate stage in the natural history.

Although the risks of PAU are well-recognized, their natural history remains poorly defined. The rupture rate has been reported to be as high as 40% (14-40%) for symptomatic presentation.[28,29] In contrast, the natural history of asymptomatic PAU appears more benign, with low rates of rupture and disease progression.[30] The mean age of the patients with PAU are 71.9 ± 6.1 years, more frequent in men (66%), and have the history of smoking (60%), hypertension (89%), hyperlipidemia (62%), and coronary artery disease (42%) are the strongest clinical and laboratory attributes of PAU.[31] The patients are invariably active or past smokers. The same factors hold for IMH. The clinical presentation of PAU is similar to AD except for valvular, cardiac rhythm abnormalities, and ischemic tendencies being seen frequently in the classic aortic dissection. Other atypical findings in the case of PAU are dysphonia, pulse abnormality, signs of a stroke, vascular insufficiency, and end-organ infarction.[32] Pain in the chest, especially radiating to the back, found to be one of the strongest predictors of PAU rupture.[33,34] The radiation of pain may suggest the site of the lesion, anterior chest pain, indicating ascending and pain in back for descending aortic lesions. Intermittent chest pain radiating to the shoulder and back can be another manifestation of the disease, with recurring pain indicating impending rupture.[35] The presence of pleural effusion and a long segment of IMH involvement are frequently seen in symptomatic cases, whereas microembolization events alerted towards the same in asymptomatic cases.[36] Even in the absence of pain, approximately one-third of patients progressed to aneurysm formation over a seven-year follow-up.[25] In many patients, PAU is encountered as an incidental finding devoid of any clinical manifestation whatsoever while investigated for an unrelated condition.

In our case the presentation of hoarseness and dysphonia due to an underlying cardiovascular pathology is a very rare clinical entity known as Ortner's syndrome (OS) or cardiovocal syndrome. The left recurrent laryngeal nerve branches off the left vagus nerve, loops around the ligamentum arteriosum and tracks superiorly between the trachea and the esophagus. In general, injury to left recurrent laryngeal nerve (*i.e.* impingement, stretching, or compression) is more common than injury to the right recurrent laryngeal nerve, likely due to its proximity to the aortopulmonary window and other intrathoracic structures. OS is specific for left recurrent laryngeal nerve injury due to underlying cardiac disease. Although it is commonly associated with severe mitral stenosis (as initially described by Ortner), there are many causes of OS including compression from other vascular (*i.e.* aortic aneurysms, aortic dissections, pulmonary hypertension) or mediastinal (*i.e.* neoplasms) structures.[36-40] Similarly, although the classic symptom associated with OS is dysphonia/hoarseness, there have been several other manifestations of the syndrome described in the literature including aspiration, dysphagia, and shortness of breath.[41]

The diagnosis of penetrating aortic ulcers relies first upon a thorough history and physical examination. The typical patient is elderly with a history of hypertension. These patients can also have a history of coronary artery disease, chronic obstructive pulmonary disease, renal disease, and tobacco use. They typically present with anterior chest or midscapular pain. Similar to aortic dissections, those with anterior chest pain usually have ascending aortic involvement and those with back pain typically have descending aortic involvement. The differential diagnosis with this typical presentation includes acute coronary syndrome, aortic aneurysm, aortic dissection, intramural hematoma, and pulmonary embolism. Physical examination should initially include a review of airway, breathing, and circulation to ensure that the patient is stable. Murmurs indicative of aortic insufficiency typically reflect aortic dissection as opposed to isolated penetrating ulcers, which are focal in nature. Similarly, signs of malperfusion such as neurologic deficits, acute renal insufficiency, visceral vessel compromise, or limb pain with pulse deficit usually occur with dissection as opposed to isolated penetrating aortic ulcers. It is important to note, however, that penetrating aortic ulcers and aortic dissections can occur concomitantly, and therefore, the presence of these signs on physical examination does not exclude a diagnosis of penetrating aortic ulcer. Penetrating aortic ulcers may also be discovered incidentally in asymptomatic patients with imaging performed for other indications.

Radiological imaging is essential to the diagnosis of penetrating aortic ulcers given its similarities to other acute aortic syndromes with respect to clinical presentation. A chest radiograph is the first modality invariably undertaken in a case of chest pain. Patients with IMH and PAU have unremarkable chest radiographs as compared to findings of mediastinal widening with or without pericardial effusion in cases of aortic dissection.[42,43]

CTA is the imaging modality of choice for evaluation of AAS being faster, less invasive, requiring less technical expertise, and ability to reproduce images in any plane with excellent resolution. CTA should be performed after clinical and laboratory evaluation, including cardiac enzymes and D-dimer assay, chest radiograph, and electrocardiogram.[44]

Guidelines regarding indications of CTA (intermediate and high-risk categories), clinical evaluation, and technique to perform CTA in cases of suspected AAS are laid down in 2016.[45] Their main emphasis was to acquire motion artifacts free images, especially of the aortic root with ECG gating. End-systolic versus end-diastolic acquisition depends on the patient's heart rate and the number of the detector array. Recommendations included coverage limited to thoracic aorta, the addition of a non-contrast sequence to detect any associated hematoma, and targeting 250 HU or more attenuation value in the arterial phase. The intimal flap of dissection and associated intramural hematoma is not evident on aortography as seen on CTA. Instead, indirect signs like medial displacement of intimal calcification can be a clue for the same.[46] On CTA, the distinction between true and false lumen can be made reasonably; however, it can be tough in cases where the entire aorta is not included in the scan. The interface between intensely enhancing true and crescentic false lumen can give a beak-like morphology. Acute cases may show outer wall calcification and convex flap morphology towards the true lumen. Transesophageal echocardiography can also be used for diagnosis with a reported high sensitivity and specificity, although its invasive nature and need for a skilled operator are relative disadvantages.[47]

The distinction between PAU and aortic dissection (AD) is vital with the site of the lesion, presence of intramural hematoma, and intimal flap providing a good demarcation improved by dynamic contrast-enhanced imaging.[44] PAU is seen on CTA as contrast filled outpouching or crater-like morphology, ranges in size from few millimeters to 2.5 cm, depth up to 3 cm, are often multiple.[24] There is invariable surrounding IMH and medially displaced calcified intima. A study by Mayo Clinic confirmed this association to the tune of 80 percent.[48] Hyperdensity in PAU on non-contrast study denotes intimal hematoma, an indicator of acute and potentially unstable state warranting prompt intervention.[49,50] The adjacent aortic segment is invariably thickened with some degree of enhancement. PAU usually does not extend beyond the aortic contour, latter being suspicious for rupture, associated hematoma or Subintimal pseudoaneurysm formation.[38] Magnetic resonance imaging (MRI) appearance is akin to the area of flow void showing flow-related enhancement on time-of-flight sequence. Increasing the TE would further enhance the detection of sluggish flow in the ulcer crater. Nonfat sequences were technically less demanding and more accurate than fat saturation sequences and even CTA.

PAU needs to be differentiated from both intramural blood pool and ulcer-like projection, with the former having a wider diameter and intimal atherosclerotic changes and the latter is commonly encountered on follow up imaging in patients with the normal aorta. The disruption of the internal elastic lamina is the key histological finding in cases of PAU, which can be seldom demonstrated. PAU is most commonly seen in descending thoracic aorta followed by abdominal aorta and arch in decreasing order of frequency.

PAU diameter of 20 mm and depth of 10 mm when taken as cutoff, predicted disease progression, suggesting early surgical intervention reasonably. PAU has the worst prognosis in cases of rupture, leading to hemomediastinum and or hemopericardium. Likewise, rupture at initial presentation and maximum aortic diameter predicted the failure of medical treatment.

The occurrence of PAU with IMH generally leads to a progressive disease course with a higher likelihood of catastrophic consequences like aortic rupture and dissection.[7,33] These patients usually belong to an older age group and show involvement of the proximal thoracic aorta. The predictors of disease progression were pain despite expectant treatment, increase in pleural effusion, and disease confined to the proximal thoracic aorta. A higher subset of symptomatic patients explained the same. The presence of pain, hemodynamic instability, suboptimal response to medical treatment, IMH thickness 11 mm or more, periaortic hematoma, and associated PAU beyond a particular dimension are all predictors for rupture.

As with other forms of AAS, medical therapy to optimize blood pressure and heart rate, and reduce aortic wall stress is required to initially treat patients with PAU and IMH. Intravenous beta-blockers and non-dihydropyridine calcium channel blockers are used to keep the blood pressure between 100 to 120 mmHg, and heart rate between 60 and 80 bpm.[49,50]

Adequate pain control is also an important consideration, as uncontrolled pain may result in sympathetic nervous system-mediated heart rate increase.[49] As with AD, patients presenting with complicated type B PAU and IMH should be considered for surgical management. Complicated disease is indicated by persistent or recurrent pain despite adequate control of hypertension, uncontrolled hypertension, aortic expansion on repeat imaging, hemodynamic instability, organ ischemia, maximum aortic diameter > 55 mm and rupture. In addition, surgical repair is indicated for any of the following features: PAU base > 20 mm and depth > 15 mm, IMH with significant periaortic hemorrhage.[49,51-53]

In our case the clinical presentation with symptomatic PAU-IMH associated with pseudoaneurysm and the information that gave us the image studies, there was no doubt that the patient required an emergent surgical therapy. The team decided to realize a TEVAR because of the high mortality predicted by EuroSCORE in our patient and the great results of TEVAR in the last years. When surgery is considered in the PAU with IMH, endovascular techniques are considered first-line therapy.[15,54] Endovascular technical success has been

reported at, or close to, 100% in a number of studies.[31,55-57] In hospital/30-day mortality post TEVAR for PAU is estimated at 4.8%.[31] Overall survival at 1, 5 and 10 years has been reported at 93%,[57,58] 72-84%[31,59] and 60-70%,[57,58] which is reflective of the comorbidities in patients with PAU. The long term aortic-related survival rates have been reported between 96-100%.[58,60]

The complication rate of TEVAR has been estimated to be as high as 38%, and the most common complications include endoleak, upper extremity limb ischemia, cerebrovascular ischemia, spinal cord ischemia, and post-implantation syndrome.[61] Endoleaks are the most common causes of TEVAR reintervention and are defined as functional failures in the deployed endografts, allowing for the persistent flow of blood into the excluded aneurysm sac. They are traditionally divided into five types based on the origin. Type I, or implantation, endoleaks occur at the stent-graft landing sites. These are due to inadequate apposition of stent-graft with arterial wall of the proximal (IA) or distal (IB) attachment sites of the deployed endograft.[62] Due to this direct pressure, there is always risk of rapid aneurysm enlargement and rupture. This can be due to many factors, including preoperative mural thrombus, vessel calcification, branching vessels, graft migration, or incorrect sizing of stent-graft materials. Type I endoleaks have been described as occurring immediately postoperatively so a more practical way to classify this type of endoleak is named them into direct endoleaks.

Type II, or backflow, endoleaks are caused by blood entering the aneurysmal sac in a retrograde fashion via patent branching vessels.[62] Often excluded by the endograft, segmental arteries can have retrograde flow via internal thoracic and intercostal arteries, contributing to endoleak and aneurysmal growth. Type III, or junction, endoleaks are due to extravasation of blood at stent-graft component junction points (IIIa) or due to graft fabric tear, suture breakage, or component fracture (IIIb).[62,63] Type III can also be observed as both early and late onset depending on contributing factors. Type IV, or porosity, and type V endoleaks are extremely rare after TEVAR. Type IV is caused by fabric porosity, which allows for the outflow of plasma into the excluded aneurysm. Type V is due to endotension or aneurysmal expansion without radiological evidence of another source.

Completion angiograms can detect early types I and III. Delayed images can show type II endoleaks. Contrast-enhanced CT is the gold standard for the detection of endoleaks on subsequent visits. Duplex ultrasound can also detect an endoleak. It is economical, free of radiation and contrast. Besides detecting the endoleak, it can also provide information on different types of flow and directions in these endoleaks. Its limitation is operator-dependent.

In a review of 27 studies evaluating TEVAR for aortic dissection, reintervention was required in 15% of cases, with 33% of those due to an endoleak.[64-66] In all indications for TEVAR, endoleaks have an estimated incidence of 3.9-15%. Data are conflicting as to which type of endoleak is most common. Type I and type II endoleaks are considered the most prevalent by several studies. The overall incidence of early and late type I endoleaks is thought to be up to 20%, and with intraoperative incidence at a rate of 3-7%. Once a decision is made to intervene, management of endoleaks varies by type. Type I endoleaks are best handled by extending the proximal and distal portions of the stent graft to include non-diseased portions of the aorta and by using endoanchors, which securely fasten edges.[67] However, the extension of the proximal or distal edges of grafted stents requires consideration of the risk associated with coverage of the left subclavian artery, left carotid artery, or spinal segmental arteries.

In our case the most common complication of the TEVAR appears immediately during the procedure, like early Type IA endoleak. Followed more recent studies the type IA endoleak most imperatively treated intraoperative with simple dilatation of the stent with balloon angioplasty (25 to 30 mm balloon), with the placement of a proximal cuff, Palmaz stent placement, endoanchors, or the embolization and coiling of the aneurysmal sac but these techniques are not always possible for technical and anatomical challenges.[68-71] No one of the treatments proposed was usable for the low refilling flow, that our case presented so we decide to follow the strategy wait and see, and to monitor in the course of time the development. We observed a progressive thrombolization and a complete resolution of the endoleak.

In conclusion, PAUs and IMH are often seen together or in conjunction with pseudoaneurysm or aortic dissection. The clinical presentation of PAU is variable but always must be in differential diagnosis of AAS. When diagnosed in the symptomatic patient, these complex aortic pathologies represent a potentially life-threatening medical condition. Prompt identification, medical management, and patients' selection for intervention are critical components of care, along with long-term surveillance. The TEVAR is considered first-line therapy in the patients with Type B AAS, which have an excellent result but it is not free of complications. Endoleaks are the most common complications following TEVAR and the management remains one of the inherent challenges to endovascular treatment. Proper planning and appropriate selection of stent-graft can prevent most of these endoleaks.

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