

Review Article

Penetrating Atherosclerotic Ulcer of the Thoracic Aorta: Diagnosis and Treatment

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Penetrating atherosclerotic ulcer (PAU) of the thoracic aorta was first described by Shennan in 1934.¹ PAU is part of acute aortic syndrome, so care must be taken concerning its diagnosis and treatment. Atherosclerotic plaques may ulcerate and erode through the internal elastic lamina into the aortic media, resulting in a PAU.^{2,3} Besides typical aortic dissection, the causes of acute aortic syndromes include aortic intra-mural hematoma (IMH) and PAU of the aorta.⁴ Although there is considerable resemblance in the initial clinical presence of these disorders, their pathogenesis and diagnostic characteristics differ, but all can rapidly bring on life-threatening complications. The background of a penetrating ulcer of the aorta is not well known.⁵ The main symptom is a severe, acute chest pain radiating to the inter-scapular area, similar to classical acute aortic dissection of the thoracic aorta. Patients usually have co-morbidities that make them poor surgical candidates and are treated with transluminal (transfemoral) placement of endovascular stent grafts.⁶ Surgical cases are those demonstrating haematoma expansion, impending rupture, or inability to control blood pressure.

Pathogenesis

In penetrating ulcer of the aorta, an atherosclerotic plaque ulcerates and disrupts the internal elastic lamina.^{7,8} When this

occurs, the media is exposed to pulsatile blood flow. This causes bleeding into the wall leading to intra-medial haematoma.⁹ The plaque may precipitate a localised intra-medial dissection associated with a variable amount of haematoma within the aortic wall, which can spread into the adventitia, forming a pseudoaneurysm, or may also cause rupture. The rate of rupture in PAU is 42%, higher than in IMH (35%) or in aortic dissection (3.6-7%).¹⁰ Ulceration of an aortic atheroma occurs frequently in patients with advanced atherosclerosis. However, a range of congenital abnormalities –such as Marfan’s syndrome, Ehlers-Danlos syndrome,^{11,12} and annulo-aortic ectasia, bicuspid aortic valve, and familial aortic dissection– are conditions that predispose to acute aortic syndromes.¹¹⁻¹³ PAU generally affects older patients with atherosclerosis involving the aorta and multiple co-morbidities, such as hypertension, diabetes mellitus, heart failure, chronic obstructive pulmonary disease, and chronic renal insufficiency.⁵

Clinical and imaging diagnosis of PAU

Frequently, PAU is asymptomatic and is diagnosed rather incidentally. In some cases symptoms are present but cannot determine PAU. The initial clinical appearance of PAU is similar to that of classic aortic dissection. PAU is diagnosed on computer tomography (CT) scan and

aortography as a contrast-filled, pouch-like protrusion of the aorta in the absence of a dissection flap or a false lumen (Figure 1); extensive aortic calcification and degenerative lesions are often present in other sites apart from the ulceration.^{14,15} Transoesophageal echocardiography demonstrates a crater-like ulcer with jagged edges, usually with the presence of extensive aortic atheroma; there is no intimal flap or false lumen. Aortic dissection usually begins with an intimal tear at the point of greatest hydraulic stress and blood courses immediately along the outer third of the media. The imaging studies useful in diagnosing typical aortic dissection demonstrate two channels separated by an intimal flap on CT scans, echocardiography, and angiography. On the other hand, IMH is a localised separation of the layers of the aortic wall by partially or totally clotted blood in the absence of an intimal tear, presumably caused by rupture of the *vasa vasorum* in the media.¹¹ Because there is no intimal discontinuity, the space does not communicate directly with the aortic lumen and, unlike the false lumen of classic aortic dissection, does not generally show enhancement with contrast administration on CT scanning and angiography.¹⁶ On transoesophageal echo examination, IMH displays an aortic wall haematoma in the absence of an intimal flap or PAU.¹⁷

Historical records of PAU

Little is known about the history of PAU. This entity has also been considered by most authors to have a poorer prognosis than classic aortic dissection.^{2,3,14,17-19} However, Harris et al²⁰ reported that disease progres-

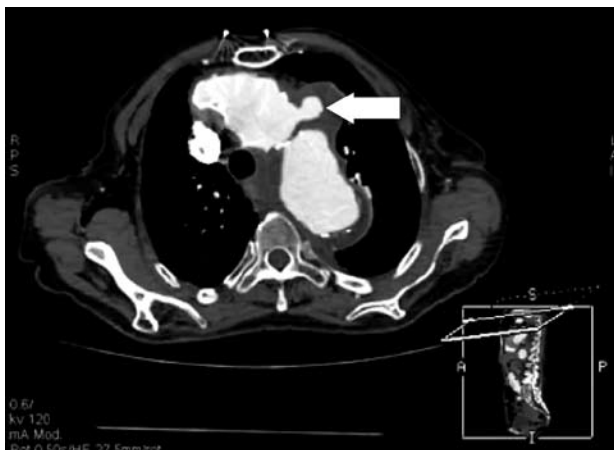


Figure 1. Chest computed tomographic scan showing the ulcer, which appears as a projection beyond the confines of the lumen (arrow), of the ascending aorta.

sion is slow, with a low prevalence of acute rupture or other life-threatening complications. For example, in the study by Stanson et al,² 13 out of 16 patients presented symptoms of back or chest pain, whereas in the study by Harris et al,²⁰ only four out of 18 patients showed these symptoms. Furthermore, if many incidental cases of PAU are included, the survival rate for affected patients may increase. According to the literature, pain is believed to be one of the most important factors determining the indication for surgical intervention.² In a retrospective study of patients with an initial diagnosis of aortic dissection, Coady et al¹⁰ found PAU in 19 (9%) out of 212 patients. In their series, the PAU patients were significantly older than those with type-A aortic dissection. Patients with PAU and IMH also tended to be older than those with type-B dissection.¹⁰ Diffuse atherosclerosis, plaque calcification, and hypertension were invariably present, apart from a history of smoking and high cholesterol level. The most common site of PAU was the descending thoracic aorta.^{10,11} In 10.5% of patients, ulcers were located in the ascending aorta. The acute rupture rate for PAU was 42.1%, as opposed to 35.3% for IMH (7.5% for type-A dissections and 4.1% for type-B dissections, $p=0.0001$). All ruptures occurred upon onset or during initial hospitalisation. Concomitant aneurysms of the abdominal aorta were found in 42.1% of patients in their series.¹⁰

Treatment

The ideal way of treating this potentially lethal disease is not well documented.^{3,5,10,11,14} PAU involving the ascending aorta is rare; however, the ulcer usually ruptures and is commonly lethal. Thus, early-urgent or emergent operative intervention is clearly recommended. Patients with PAU involving the descending aorta can initially be treated conservatively by following an aggressive medical therapy and also with close observation, similarly to descending aortic dissections. However, PAU and IMH are more serious and lethal conditions than classic descending aortic dissection.¹⁴ Surgery becomes necessary when there are signs of expansion of the IMH, rupture into the pericardial or pleural cavity, uncontrolled chest pain and haemodynamic instability. In the absence of clinical deterioration, the patient may be treated in a conservative way, with heart rate and blood pressure control. Because of the fact that PAU usually affects elderly patients who also have severe atherosclerosis and other comorbid conditions, patients with PAU in

the descending aorta are often not ideal candidates for surgical intervention. The factors described above should be taken into consideration before the decision is taken regarding the most appropriate type of treatment. Given the substantial morbidity associated with conventional surgery, endovascular repair is an attractive and challenging alternative method in these high-risk patients.^{6,11,21}

Successful treatment of aortic dissecting aneurysms and penetrating ulcer of the thoracic aorta using stent-grafts has recently been described.^{21,22} This new method offers the possibility of treating patients who are not eligible for conventional surgical procedures. However, there is limited experience with stent-grafting for penetrating ulcer of the aorta. Limiting factors for successful stent-graft repair are the larger size of the delivery systems, stent migration, rupture of the aortic wall by stent penetration, and the lack of appropriately sized devices.^{21,23,24}

Most authors suggest that surgical intervention with grafting of the affected area is the treatment of choice because of possible malignant involvement.^{2,3,14,18,19} In particular, persistent or recurrent pain, haemodynamic instability, and a rapidly expanding aortic diameter have been considered indications for surgical treatment.² On the other hand, some authors believe that immediate surgical treatment is not always required because the disease may have a benign course.^{15,20} In the study by Harris et al,²⁰ few patients developed aortic dissection or aortic rupture during follow up. These authors emphasised that most patients with PAU are put at high risk if surgical intervention is applied, because of their advanced age and poor general health. There are no large series in the international literature that stress the use of stent-grafts. Murgo et al²⁵ emphasised that surgical repair of the descending thoracic aorta is frequently complicated by respiratory disease, renal insufficiency, or spinal ischaemia, and recommended the transluminal placement of endovascular stent-grafts for PAU. All their cases were symptomatic and complicated by rupture, false aneurysm, or retrograde dissection. Endovascular stent-graft deployment was successful in all patients. Common complications, such as perigraft leak and transient spinal ischaemia, may occur. The 30-day survival rate was 100%, but one patient died from pneumonia with cardiac failure 34 days after the procedure.²⁵⁻²⁷

Transluminal placement of endovascular stent-grafts for the treatment of penetrating ulcers of the descending thoracic aorta appears to be a possible al-

ternative to typical intervention.^{6,21,22,24,25} After treatment, follow up by CT is essential to detect possible complications of the disease. Because rupture cannot be predicted from the early imaging findings, we have concluded that penetrating ulcers of the descending thoracic aorta should initially be treated aggressively and that pertinent clinical and radiological data should be monitored carefully during the first month after the onset of symptoms.^{26,27}

In the field of endovascular surgery many reports have been published; a single-centre report refers to the successful treatment of many aortic pathologies in emergent or urgent setting, without serious complications.²⁸ A misdiagnosed Takayasu's arteritis mimicking aortic intramural haematoma was treated through open classical surgery. Transoesophageal echocardiography was suggestive of an IMH of the distal ascending aorta and transverse arch, data confirmed by CT. The histological examination revealed Takayasu's arteritis.²⁹ This is an example of deficiency in the precise diagnosis in the case of endovascular surgery. According to some authors, in asymptomatic cases, there are two possible ways of handling the situation: the first being conservative treatment and the second endovascular repair (Figure 2). In our opinion, in the era of endovascular surgery, the stent-graft is a useful and safe instrument in the hands of the cardiovascular and thoracic surgeon and could be used with the patient's consent.

Conclusion

In conclusion, PAU of the aorta is an acute condition for which methods of appropriate diagnosis and ideal management should be codified. A high degree of clinical suspicion is necessary for a proper diagnosis. PAU affects the elderly, who usually have co-morbidities and a record of severe atherosclerosis, hypertension, and hyperlipidaemia, with lung and/or renal dysfunction or peripheral vascular disease. The symptomatology is similar to that of a thoracic aortic dissection, i.e. acute chest, retrosternal or interscapular pain. Differentiation of PAU from other causes of aortic disease, such as aortic dissection, aortic aneurysm, and spontaneous aortic rupture, is difficult or even impossible in many cases. Since critical cases of PAU cannot be identified on the basis of initial imaging findings, careful follow up is needed in affected patients, particularly during the first month after onset. Deciding between the conservative, surgical or endovascular mode of treatment is a common

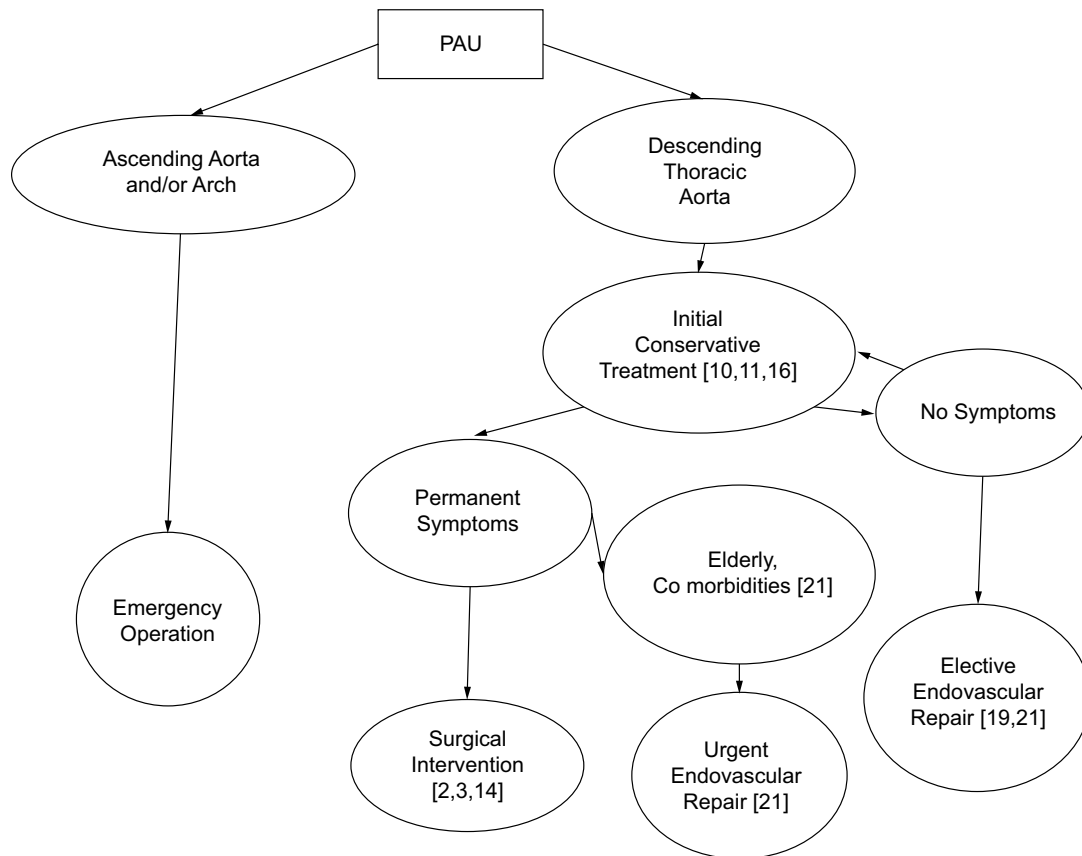


Figure 2. Algorithm proposed by the authors for the management of penetrating atherosclerotic ulcer (PAU) of the thoracic aorta.

dilemma. The presence of co-morbidities and the side of PAU are the most important determinants of decision.

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