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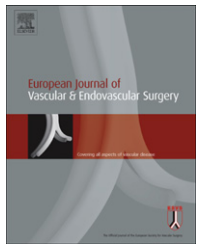

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Intramural Hematoma and Penetrating Ulcers: Indications to Endovascular Treatment

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Abstract Intramural hematoma (IMH) of the aorta and penetrating aortic ulcer (PAU) are important variant forms of classic double-barrel aortic dissection in patients presenting with acute aortic syndrome. Recent insights provided by modern high-resolution imaging are currently challenging previous pathophysiologic concepts underlying IMH and PAU, suggesting a close relationship of both entities. Thoracic endovascular aortic repair (TEVAR) offers a less invasive approach to the treatment of affected patients with very encouraging early to midterm results. This review discusses current indication for TEVAR in IMH and PAU patients in the view of an improved understanding of these diseases.

Introduction

Diseases of the thoracic aorta contribute significantly to the high overall mortality from cardiovascular disease.[1,2] In recent years, advances in modern high-resolution imaging have provided valuable insights into the pathophysiology of aortic diseases and have identified important subforms of classical double-barrel aortic dissection, resulting in a better understanding of acute aortic diseases.[3,4] Acute aortic syndrome (AAS), which contrasts nicely with acute coronary syndrome, describes patients who present with somewhat uniform sudden chest/back pain symptoms of sharp/tearing character (''aortic pain''), which are caused by various acute aortic diseases.[4] The subsumption of such patients under the term ''acute aortic syndrome'' allows for a more uniform, standardized approach to diagnosis and management of acute aortic diseases.[4]

Acute aortic syndrome encompasses classic aortic dissection with true and false-lumen (class 1 dissection according to Svensson[3]) and less common variant forms or potential precursors of classic dissection such as intramural hematoma of the aortic wall (IMH, class 2 dissection), subtle dissection (class 3 dissection), which is found intra-operatively in Marfan patients, penetrating aortic ulcer (PAU, class 4 dissection), and iatrogenic or traumatic transsection of the aorta (class 5 dissection). In some

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patients, AAS may also be caused by symptomatic aortic aneurysm.[2,5]

The aim of this review is to summarise the currently available literature with respect to indication to endovascular treatment in patients presenting with IMH or PAU.

Intramural hematoma

Intramural hematoma accounts for approximately 6–10% of all acute aortic syndromes.[6,7] In Asian studies, the incidence of IMH among AAS patients was significantly higher, approximating 30–40% of patients.[5,8] First described by Krukenberg in 1920,[9] IMH is commonly defined as bleeding into the outer layers of the aortic media, presumably caused by apoplectic rupture of aortic vasa vasorum, which are prone to rupture due to alterations from chronic arterial hypertension.[1,4] By definition, IMH lacks a detectable intimal tear/disruption, and has therefore no communication with the true aortic lumen, but is confined within the aortic wall (''dissection without intimal tear''). On imaging IMH is identified by crescent-shaped or concentric thickening of the aortic wall, which in its early stage shows hyper-intensity on T2 imaging by MRI or hyper-density in non contrast-enhanced computed tomography (CT). Thus, IMH can even be detected on a native CT scan. Luminal displacement of intimal calcification may be helpful in differentiating IMH from mural thrombus. In addition, transesophageal echocardiography provides important information for diagnosing IMH.[6] Intramural hematoma may affect the entire thoracic aorta and usually – similar to classic dissection – involves a longer segment of the aorta.

Symptoms of IMH may be very similar to those of classic dissection, and patients can not be reliably distinguished by clinical presentation alone.[4] However, the risk of malperfusion is lower with IMH and thus symptoms of organ ischemia may be absent in IMH patients.

So far, the natural history of IMH is not fully understood. Published data are somewhat conflicting with some reporting a rather favorable outcome of IMH as compared to classic dissection, whereas others have reported similar mortality and complication rates to double-barrel dissection.[5] Complications of IMH are quite common and include progression towards overt false-lumen dissection in 28–47% of cases, early aneurysm formation or (contained) rupture in 20-45% of patients.[7] Patients may, however, show spontaneous reabsorption of IMH under medical treatment, although regression is less common. Predictors of progression include recurrent or persisting pain, and presence of penetrating aortic ulcer, while younger age, aortic diameter $<$4.0–4.5 cm, and hematoma thickness $<$1.0 cm portend a better prognosis.[4,5]

Previous studies have shown that the location of the IMH is of paramount importance for patient's prognosis. In analogy to the Stanford classification of classic aortic dissection, involvement of the ascending aorta (type A-IMH) demands urgent surgical repair in most of the patients due to the risk of rupture or progression to frank dissection. It should, however, be noted that in particular groups from Japan and Korea have documented a more benign course of type A-IMH with non-operative management, recommending primarily strict blood pressure control, prolonged bed rest (4 weeks), and close imaging surveillance.[7,8] It has been speculated that the relatively low mortality rate in Asian IMH patients may be explained by a genetic factor or the fact that more limited IMHs were included in these series.[7] Intramural hematoma confined to the aortic arch or the descending thoracic aorta (type B-IMH) may primarily be safely treated non-surgically with close imaging follow-up.

Penetrating aortic ulcer

Penetrating aortic ulcer of the aorta has first been described by Shennan in 1934,[10] but has only recently been acknowledged as a distinct pathologic variant of classic false-lumen aortic dissection.[11] Previous investigators have estimated that 2.3–7.6% of acute aortic syndromes are caused by PAU.[12] Penetrating atherosclerotic ulcer is defined by an ulceration of an aortic atherosclerotic plaque penetrating through the internal elastic lamina into the aortic media.[11]

Penetrating aortic ulcer occurs most often in patients with extensive atherosclerotic disease.[11] Therefore, usually patients in their 7th life decade and older are affected by PAU. Similar to IMH, patients may not be distinguished from classic dissection by clinical presentation alone, since symptoms may be very similar. Penetrating aortic ulcer has a characteristic appearance on angiography, reminiscent of duodenal ulcer. Nowadays, angiography is only rarely performed to diagnose PAU, whereas non-invasive imaging modalities, which in addition provide important information on the tissue surrounding the aorta, are used more frequently. On imaging, PAU typically appears as one or more focal, contrast-material filled, craterlike outpouchings of the lumen, with a thickened aortic wall and inward displacement of calcified intima by concomitant intramural haematoma.[13] Penetrating ulcers have been described along the whole length of the aorta but most commonly involve the mid and distal descending thoracic aorta. There is a strong association of PAU with concomitant abdominal aortic aneurysms. Penetrating ulcers of the ascending aorta (type A PAU) are rare, but dreadful requiring urgent surgical repair in most patients.[4,13]

Complications of PAU include development of (localized) intramural hematoma due to arrosion of aortic vasa vasorum by the ulcer, (pseudo)aneurysm formation, progression to overt aortic dissection, or rupture in up to 40% of patients.[4,14–16]

Again similar to IMH, the natural history of PAU is not fully understood yet. There are conflicting data in the literature with respect to disease behaviour.[14–16] While some authors reported PAU to be malignant,[11,14–16] others reported a lower incidence of life-threatening complications.[17,18] It appears that symptomatic patients presenting with AAS caused by PAU have a worse prognosis than those who are asymptomatic and PAU is found incidentally on axial imaging.

Recent insights into IMH and PAU

The rigorous classic definition of IMH as being a ''dissection without a tear'' has long been doubted by several