Case Report

Intramural hematoma and penetrating atherosclerotic ulcer: a case report and review of literature

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Penetrating atherosclerotic ulcer (PAU) represents part of the "acute aortic syndrome" (AAS) spectrum. It is uncommon but potentially hazardous if delayed or misdiagnosed. It may progress to complications such as intramural hematoma (IMH), aortic dissection (AD) or rupture. It is characterized by the ulceration of the aortic wall as a result of atherosclerotic plaque along the wall of aorta. Here, we describe a case of PAU complicated with IMH. (Rawal Med J 201;40:468-471).

Keywords: Atherosclerotic, Plaque, Atheroma, Aortic dissection, Hematoma

INTRODUCTION

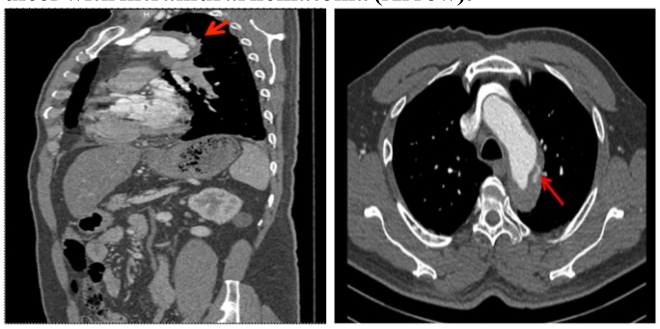
Penetrating atherosclerotic ulcer (PAU) represents part of the "acute aortic syndrome" (AAS) spectrum that is rarer than classical acute aortic dissection (AD). It was first described by Shennan et al. in 1934, and subsequently by Stanson et al. in 1986, as a distinct clinical and pathological entity under AAS.[1,2] It is characterized by the ulceration of the aortic wall as a result of atherosclerotic plaque along the wall of aorta that disrupts the internal elastic lamina.[2] Unlike AD, there is an absence of intimal flap or creation of false lumen. Occasionally, the ulcerated plaque may penetrate and hemorrhage into the medial layer of the aortic wall causing intramural hematoma (IMH). Here, we describe a case of PAU complicated with IMH, which is uncommon but important in early recognition.

CASE PRESENTATION

A 57-year-old Malay man presented to the emergency department, Hospital Enche' Besar Hajjah Khalsom, Kluang with sudden onset of tearing retrosternal chest pain radiating to the back. There was diaphoresis associated with the syncopal attack. Otherwise, there was no neurological deficit. He had hypertension but was not compliant to medication and follow-up. He was a chronic smoker but no significant family history of cardiovascular diseases.

Upon arrival, his blood pressure was 270/150 mmHg with normal pulse rate and saturation of oxygen. Cardiovascular examination showed no pulse deficit and presence of all peripheral pulses. The examination of his respiratory and neurological systems was unremarkable.

Figure 1 and 2. CTA showing penetrating atherosclerotic ulcer with intramural hematoma (Arrow).



Chest radiography revealed widened mediastinum and borderline cardiomegaly with no pleural effusion. Computed tomography angiogram (CTA) (Figure 1 and 2) of the aorta revealed the presence of PAU at the ectatic aortic arch with small IMH.

He was managed medically with analgesic, antihypertensive agents and statin. After one week of hospital stay, he was discharged well with three oral antihypertensive and his blood pressure was maintained less than 140/90 mmHg. Follow-up appointment with cardiothoracic team was given.

DISCUSSION

The incidence of PAU among those with acute aortic syndrome ranges from 5%-10%.[3-6] However, PAUs had also been diagnosed incidentally in asymptomatic patients who underwent radiological

imaging for other purposes. Up to 25 percent of PAUs in the Mayo clinic series were asymptomatic at the time of diagnosis.[3] Thus, the exact prevalence of PAU may be underestimated in the population. As compared to AD, patients with PAU are older (6th to 8th decade of life) with more extensive atherosclerotic disease and higher prevalence of hypertension and smoking history.[3,5,7] These patients have more comorbidity, potentially limiting the therapeutic options. The location of PAU is consistent with the extent of atherosclerosis, with about 80-90% occurring in descending thoracic aorta, and only a handful in the ascending aorta or isolated to arch of aorta.[3,5,6,7] Our patient has uncontrolled hypertension and was a smoker, but is relatively younger compared to patients in these studies. Also, the PAU was in the arch, which is a far less common site.

Asymptomatic presentation of PAU is possible especially if the ulceration is confined to the intimal layer.[3] Nathan et al. reported 80% associated with IMH were symptomatic as compared to 4.5% of PAU without IMH. Likewise, patients with thoracic PAU (22.7%) were more likely to experience acute symptoms compared to those located at abdominal aorta (6.7%).[8] Rupture rate at presentation varies greatly but was reported as high as 38% in some groups.[4] Minority of patients may have peripheral emboli. However, as opposed to AD, pulse deficit, visceral insufficiency and aortic regurgitation are not seen in PAUs, unless it progresses to aortic dissection.[9,10] Consistent with the current data, our patient did not experience such issues at presentation.

The diagnosis of PAU remains mostly a combination of clinical presentation and radiological imaging, as histopathological diagnosis is not possible in patients who do not undergo surgical intervention. Chest radiography may show diffuse or focal enlargement of aorta with the presence of pleural effusion or pericardial effusion signifying evidence of rupture.[11] The advancement of transesophageal echocardiography (TEE), CT and MRI have aided in the recognition of PAU. On contrast CT scan, PAU is identified as a focal medium-filled out pouching in the aortic wall in the absence of intimal flap or false lumen, often with associated thickened aortic wall.[12] It may be surrounded by intramural hematoma beneath the frequently calcified and inwardly displaced intima.[13] MRI and TOE identify PAU as craterlike projections in the aortic wall.[12] On imaging, often there are extensive aortic calcification and atherosclerotic changes, and sometimes one may also find evidence suggestive of rupture, including pleural effusion, pericardial effusion and mediastinal hemorrhage.

For AD, it is generally accepted that Stanford type A disease behaves in a more malignant manner as compared to Stanford type B, thus requiring surgical intervention. Whether this is true for PAU as well remains a matter of debate. PAU may remain stable, resolve or enlarge and progress leading to complications including intramural hematoma, aortic dissection, saccular aneurysm or even rupture. Reports vary considerably on the outcome of PAUs. Rates of complications or progression seemed low in patients who were asymptomatic.[8,14-16] Progression is more likely if significant pain is uncontrolled, pleural effusion is increased, more proximal involvement, or larger PAU. A 20 mm or more in the diameter of PAU has a positive predictive value of 100 percent for progression, whereas 10 mm or more in the maximum depth of PAU has an 80% positive predictive value for progression.[6]

Nathan et al reported 42 percent of PAUs in symptomatic patients progressed (enlarged or developed complications) compared to 17 percent of asymptomatic patients.[8] Series from Mayo clinic seemed to support the less malignant nature of the disease, with majority of the patients remained stable without surgical intervention[3]. In contrast, Yale group reported much more aggressive disease with 40 percent rupture rate in patients with PAU.[9] Under close scrutiny, the Yale group had higher prevalence of ascending aorta involvement (46%) compared to the Mayo clinic group (9%). Thus, the higher occurrence of PAU in ascending aorta in Yale group may be the reason of higher rates of rupture compared to other studies.

As a result of the rarity of PAU and widely differing views on prognosis, clear consensus on treatment is

lacking. For patients presenting with rupture or complications of disease, they will require surgical intervention. However, the dilemma lies in patients without such issues. This is further complicated with higher rates of comorbidity among patients with PAUs, thus, rendering surgical intervention of unacceptable risks in many of them.

Mayo Clinic reported 72 percent of their patients managed with medical treatment had favorable outcome.[3] Thus, pain management and blood pressure reduction form the cornerstone of overall management in this group. Close monitoring of disease progression or complications is important to identify patients who require surgical interventions. Others are more vocal on aggressive surgical treatment based on their experience of high rupture rates and complications if left without operative intervention.[5,6,9] Recent advances in endovascular repair have also added an alternative to surgical treatment in patients with prohibitively high risk for open repair. The outcomes of endovascular repair are reportedly good with low rates of complications and mortality generally.[4,17-21] However, whether endovascular repair will improve the outcome of patients with PAU remains to be determined. Although our patient was young without much comorbidity, he was not subjected to surgical intervention. We opted for medical therapy as his symptoms were controlled well with blood pressure control and analgesia.

In summary, IMH complicating PAU is a rare yet important differential diagnosis for patients presenting with acute central chest pain. Early diagnosis is important, as the disease progression is variable and unpredictable. Late detection may lead to unfavorable outcome, especially in symptomatic patients. The awareness of clinicians in this condition is extremely crucial. Yet, there is still inadequate evidence to suggest the optimal initial treatment for this condition.

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Analysis and interpretation of the data: SY
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