An Unusual Cause of Chest Pain: Extensive “Type B” Intramural Hematoma with a Descending Aortic Pseudoaneurysm

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Introduction

Intramural hematoma of the aorta (IMH) represents approximately 10-30% of cases of acute aortic syndromes [1]. The disease process is a variant of aortic dissection where a collection of blood accumulates within the aortic media without the presence of an intimal flap. The typical presentation involves acute chest or back pain, typically in elderly male patients with history of hypertension and atherosclerosis. IMH is classified according to the Stanford classification system, with a reported incidence of type A IMH in 57% of the cases versus 43% of type B lesions [2]. Aortic pseudoaneurysm or contained rupture complicates 20-45% of IMH cases and in 28-47% of patients; there can be progression to overt aortic dissection [3]. Here we describe a case of IMH complicated by an aortic pseudoaneurysm.

Case

A 62 year old man with a history of hypertension and active cocaine use presented with intermittent dull left sided chest pain radiating to his lower back. He was markedly hypertensive on presentation with a blood pressure of 190/110mmHg. A 12-lead electrocardiogram showed normal sinus rhythm with T-wave inversions in leads V4-V6. A troponin-I level was within normal limits at 0.020 ng/ml (N-0.000-0.045). An initial chest radiograph showed ectasia of the aortic arch and descending thoracic aorta. A transthoracic echocardiogram was performed, showing severely reduced left ventricular systolic function with a left ventricle ejection fraction of 25-30% with severe global hypo kinesis. In addition, the aortic root was noted to be dilated, measuring 4.2 cm at the sinuses of Valsalva. On suprasternal views, there was a suggestion of a possible descending aortic aneurysm and possible mural thrombus.

Given these findings, a CT angiogram of the aorta was performed. This revealed an extensive type B intramural hematoma originating distal to the left subclavian artery and extending to involve the entire thoracic aorta up to the level of the diaphragm. Additionally, there was an irregular saccular structure in the mid-portion of the descending aorta measuring 5 cm in the cranio-caudal plane that appeared most consistent with a pseudo aneurysm (Figure 1). A subsequent coronary angiogram showed severe two vessel disease of the obtuse marginal and posterior left ventricular branches. Cardio-thoracic surgery was consulted and the patient subsequently underwent a carotid subclavian bypass along with thoracic endovascular aneurysm repair (TEVAR). The repair included a three-piece aortic stent up to the level of the celiac vessels (Figure 2).

Discussion

Contrast-enhanced CT imaging is the preferred imaging modality used to detect acute aortic syndromes and is useful in differentiating between an aortic dissection, intramural aortic hematoma and penetrating atherosclerotic ulcer. A smooth non-enhancing crescentic region of aortic wall thickening within the intima without a spiraling intimal flap is considered diagnostic of an intramural hematoma [4]. A normal aortic diameter in the acute phase is the strongest prognostic measure for regression of IMH whereas the absence of echo lucent

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areas and atherosclerotic ulcerated plaques are associated with evolution to aneurysms [1]. Saccular pseudoaneurysms can enlarge at an approximate rate of 1.2 cm per year and thus have a high risk of rupture [5]. Type A IMH involving the ascending aorta is typically managed with emergent surgery given high mortality rate and better survival rates [2]. Type B IMH is usually managed conservatively with anti-hypertensives, but persistent pain, hemodynamic instability and complicated features like saccular or fusiform aneurysms, ulcers, dilated descending aorta, peri-aortic hemorrhage and pleural effusions in the acute phase are considerations for surgical or endovascular treatment as in our patient [6,7]. TEVAR is an option for elderly patients with comorbidities and has a variable rate of neurological complications [7,8]. It does carry a re-intervention risk but with good patient selection in experienced centers, it has moderate post-op outcomes with lower mortality rates [7,8].

The approach of TEVAR requires a proximal and distal landing pass of at least 2 cm. In patients where achievement of a proximal seal necessitates coverage of the left subclavian artery, revascularization with extrathoracic debranching with carotid subclavian bypass is necessary [9]. Carotid sub-clavian bypass in the era of TEVAR has proved to have good technical success with low mortality risk [10]. The post hospitalization mortality rates of IMH at 1 year were found to be around 26% in one study, warranting the need for follow up surveillance imaging after discharge [3].

References