



Rare Cause of Vascular Myelopathy Mimicking Gullian Barre Syndrome

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Abstract

Spontaneous intra abdominal aortic dissection is a rare cause of vascular myelopathy associated with high morbidity and mortality. A high index of clinical suspicion is required to diagnose as early as possible with minimum investigations available so as provide immediate management. Though endovascular management is considered option of treatment for all dissections we managed our patient conservatively and are under continuous surveillance at present.

Keywords: Aortic dissection; Vascular myelopathy; Endovascular; Surveillance

Introduction

Aortic dissection is a life-threatening condition caused by a tear in the intimal layer of the aorta or bleeding within the aortic wall, resulting in the separation (dissection) of the layers of the aortic wall. It is most common in 65-75 years of age, with an incidence of 35 cases per 100,000 people per year in this population. Other risk factors include hypertension, dyslipidemia and genetic disorders that involve the connective tissue, such as Marfan syndrome. Proper diagnostic confirmation and adequate treatment are crucial in managing affected patients. As aortic dissection poses a special diagnostic challenge for physicians because of its relative rarity and because symptoms of aortic dissection are likely to mimic other more common conditions. For same reasons, the correct diagnosis is missed and so delayed initial presentation as was seen in our case. Early diagnosis is important as morbidity and mortality are related to late implementation of treatment. Specific features that relate to management decisions are important, such as the presence of rupture, the extent of the dissection, the involvement of branch vessels and end-organ ischemia.

Case Presentation

A 52 year old lady diagnosed outside as Guillain barre syndrome was referred to our tertiary institute for further management in emergency. On re-evaluation she had history of acute onset proximal lower limb weakness with mild back pain and bladder incontinence. Patient denied any prior history of fever, trauma or systemic symptoms or any other comorbid illness. On examination her blood pressure was 170/110 mmHg, radial pulse was 92/min and other upper limb pulses were normal. Lower limb pulses-bilateral dorsalis pedis, posterior tibial, popliteal were absent. Femoral pulse bilateral was low volume. Cardiovascular and abdominal examination was normal. Central nervous system examination revealed is flexic hypotonic bilateral lower limbs with mute plantar response bilateral and loss of all sensations below L1 level. Investigations including hemogram, renal and liver function tests, serum electrolytes, vasculitis profile were normal. Only imaging which we did was Computed Tomography Angiography (CTA) of abdominal and lower limb vessels which revealed dissection extending from upper abdominal to infra-renal abdominal aorta and then also present at bifurcation of common iliac artery (Figure 1). Patient was managed conservatively with beta blockers and anticoagulants, as patient refused surgical management and supportive measures for myelopathy were initiated. At present patient is on our follow up to look for any further progression of disease and advised imaging after 3 months.

Discussion

Aortic dissection occurs from an intimal tear which separates the layers of the aortic wall, allowing blood to flow between the layers. Unlike ascending aorta, abdominal aortic dissections are rare events and diagnosis is often missed in one third of patients. Abdominal aortic wall is relatively

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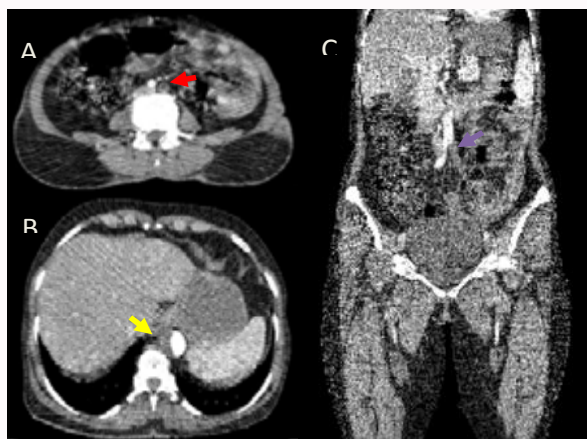


Figure 1: CT angiogram of the abdominal aorta shows evidence of eccentric hypodensity (yellow arrow in 1A) along the aortic lumen likely S/O dissection which is extending caudally across the bifurcation causing occlusion of the left common iliac artery (red arrow in 1B). Coronal reformat (1C) of the same shows occluded left common iliac artery (yellow arrow).

avascular and less rich in elastic fibers than the thoracic segment and in addition has a stiff wall making it prone to degenerative changes such as atherosclerosis and aneurysmal dilation, but provide some protection against dissection [1]. Trimarchi et al. [2], in his large series of acute aortic dissections found abdominal dissection only in 1.3%. In addition studies in form of case reports and small patient series report incidences 1.1% to 4% due to its low incidence. Jonker et al. [3], in his lucid review on abdominal dissections found hypertension as risk factor in 51% of patients and abrupt onset of symptoms in 74% of patients with abdominal pain being the most common symptom seen in 47% of patients which was not seen in our patient [2]. In addition myelopathy was present only in 3% of patients which was the only predominant symptom seen in our patient which is rare. Though Computed Tomography (CT) angiography is considered first imaging modality of choice to detect the dissection only in 9% cases dissection was diagnosed [3]. In our case CT angiography revealed the diagnosis so that immediate management could be started. Spontaneous dissections are more commonly seen predominantly in patients more than 60 years of age and have chronic presentations. Our patient age was 52 years and presented acutely which is uncommon. Majority of dissections are seen between renal artery and inferior mesenteric artery and also in common iliac artery (50%) as seen in our case. To make a diagnosis is not as easy as it seems to be, high index of clinical suspicion, thorough history and clinical examination is warranted so that prompt diagnosis can be made as soon as possible and management initiated.

Though treatment strategies remain unclear but management results are better compared to more common thoracic dissections. In Jonker et al. [3] review 27% patients were managed medically and dissections in distal common iliac artery were better managed with open repair. Since our patient refused surgical management, had uncomplicated dissection needs continuous surveillance for development of any fatal complications (9% patients) like rupture [4,5]. So impending signs of complications need to be explained to the patient. As abdominal dissections are rare and only some cases and series have been reported. The mechanism of neuronal injury resulting in myelopathy is reduction of spinal cord perfusion pressure and hypoxia from impaired collateral supply to spinal arteries fed from the vertebral, internal mammary, intercostal, lumbar and hypogastric (internal iliac) arteries [6,7]. We believe our case is also unique as aortic dissection presenting with myelopathy is rarely described, thus can help in adding knowledge in literature for conservative management of abdominal dissection in addition to delineating proper history and clinical examination in such cases. This case demonstrates that in case of uncomplicated dissection which was misdiagnosed as GBS. Conservative management can be considered with careful continuous surveillance.

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