

Right Phrenic Nerve Palsy: A Rare Presentation of Thoracic Aortic Aneurysm

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Abstract

Phrenic nerve palsy causing hemidiaphragm paralysis is a very uncommon feature of thoracic aortic aneurysm. In one case, a 45-year-old man complained of chronic chest pain, dysphagia, and hoarseness of voice; posteroanterior view chest radiograph revealed lobular enlargement of the superior mediastinum and elevated right hemidiaphragm. Contrast-enhanced computed tomography (CT) of the thorax revealed a giant partially thrombosed aneurysm originating from the ascending aorta and extending into the aortic arch, causing a widening of the aortapulmonary window and a compression of the thoracic esophagus. Right hemidiaphragm elevation was explained by the gross mass effect of the aneurysm on the right hilum, causing right phrenic nerve palsy. The patient was to be operated on for surgical correction of the aneurysm, but died before surgery due to spontaneous rupture.

Keywords: dysphagia, hoarseness, thoracic aortic aneurysm, palsy, phrenic nerve

Introduction

Thoracic aortic aneurysms are usually asymptomatic (1), with the most commonly reported symptom being chest pain. Right hemidiaphragm paralysis, due to right phrenic nerve palsy, is a very rare feature of thoracic aortic aneurysm. To the best of our knowledge, only one case of thoracic aortic aneurysm causing phrenic nerve palsy is reported in the literature (2). Here, we present a case of ascending thoracic aortic aneurysm in a patient presented to us complaining of chest pain, dysphagia, and hoarseness of voice, and featuring elevated right hemidiaphragm observed in posteroanterior view chest radiograph.

Case report

A 45-year-old man presented symptoms of chronic chest pain for five months, as well as dysphagia and hoarseness of voice for the last two months. There was no history or occurrence of fever, cough, haemoptysis, dyspnea or weight loss. On physical examination his vitals were within

the normal limit. There was no lymphadenopathy present, and an examination of respiratory system, cardiovascular system and abdomen did not reveal any abnormality. The patient was a smoker but there were no other predisposing risk factors found upon examination for aortic aneurysm.

On laboratory examination, his complete blood count, renal function tests, liver functions tests, and lipid profile were within the normal limit. A posteroanterior view chest radiograph revealed lobular enlargement of the superior mediastinum with silhouetting of the ascending aorta and aortic arch, along with elevated right hemidiaphragm (Figure 1). Ultrasonography of the abdomen was normal. Contrast-enhanced computed tomography of the thorax revealed a giant partially thrombosed aneurysm originating from the ascending aorta and extending into the aortic arch with no extension into the descending aorta. The size of aneurysm was 5.3 × 12.2 × 13.5 cm (Antero-Posterior × Transverse × Cranio-Caudal). Thrombus was eccentrically located

within the lumen on right anterolateral aspect, with a low thrombus-to-lumen ratio of about 0.33. The aneurysm was causing a widening of the aorta-pulmonary window with resultant left recurrent laryngeal nerve palsy. The aneurysm was also causing compression of the thoracic esophagus, which manifested as dysphagia (Figure 2). There was also evidence of mass effect on the right hilum, with compression of the superior vena cava, and along the expected location of right phrenic nerve (Figure 3). The right phrenic nerve is closely associated with the superior vena cava, as it passes anterior to the root of the right hilum, as shown in the cross sectional CT diagram (Figure 4). Direct visualisation of the nerve is at times difficult on a CT. There was no evidence of any high-attenuation hematoma in the adjacent mediastinum, pleural cavity, pericardium, or luminal structures such as the airway or esophagus, indicating a lack of rupture. The intact aortic wall without any discontinuity also suggested a lack of rupture. However, a large aneurysm size, a low thrombus-to-lumen ratio, and draping of the posterior aspect of aneurysm over the vertebrae, known as the “draped aorta sign,” were suggestive of instability or impending rupture. On flexible bronchoscopy examination, there was left vocal cord paralysis. The patient was diagnosed with a case of ascending thoracic aortic aneurysm causing compression of the esophagus, the left recurrent laryngeal nerve, and the right phrenic nerve. The patient was referred to cardiothoracic-vascular surgery unit for surgical repair of the aortic aneurysm, but unfortunately died due to spontaneous rupture of aneurysm before surgery.

Discussion

An aortic aneurysm is a permanent localised dilatation whose diameter is at least 1.5 times greater than the normal diameter of that portion of the aorta (3). The aortic aneurysm is classified as either saccular or fusiform, based on its shape. The most common cause of aneurysm is atherosclerosis. It is also associated with hypertension, smoking and advanced age, usually in the 6th and 7th decades of life. It can also occur due to weakening of the aortic wall associated with connective tissue disorders such as the Marfan and Ehler-Danlos syndromes. Inflammation of the aorta and injury from falls or motor vehicle accidents are other potential contributing factors. In our patient, only a history of smoking was present, with no other predisposing factors. There was no family history of aortic aneurysm.

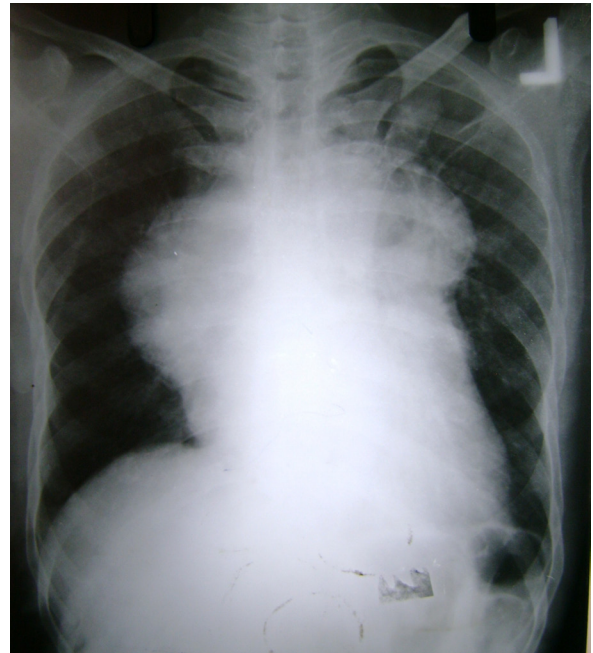


Figure 1: Posteroanterior view chest radiograph showing lobular enlargement of superior mediastinum with silhouetting of ascending aorta and aortic arch. Also noted was elevated right hemidiaphragm consistent with right phrenic nerve palsy.

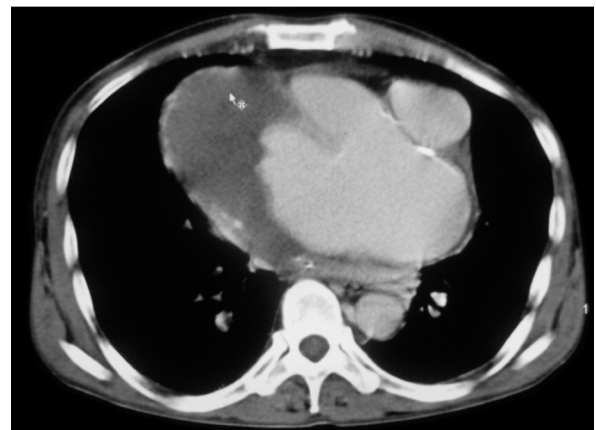


Figure 2: Contrast-enhanced axial computed tomography at the level just below the aortic arch shows giant partially thrombosed aneurysm originating from ascending aorta and extending into arch of aorta, causing widening of aorta-pulmonary window, and which is responsible for left recurrent laryngeal nerve palsy. Aneurysm is also compressing thoracic esophagus.

Thoracic aneurysms are usually asymptomatic and are often identified incidentally on chest radiographs as mediastinal widening (1). Chest pain, due to compression or erosion of surrounding structures, is the most common symptom. Acute, sudden onset of severe pain in the back or abdomen may represent rupture and

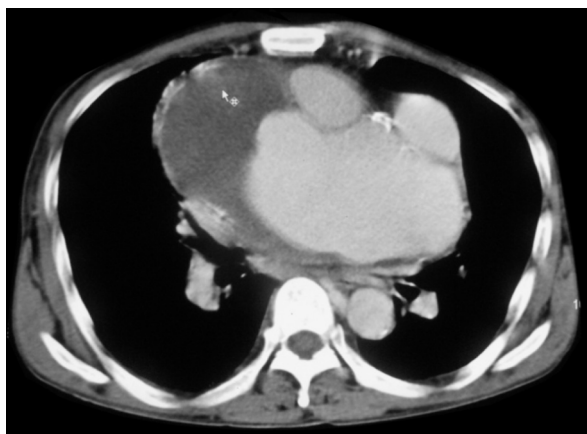


Figure 3: Contrast-enhanced axial computed tomography at the level of hila shows aneurysm causing mass effect on right hilum with compression of superior vena cava and the expected location of right phrenic nerve. Right phrenic nerve is closely associated with the superior vena cava as it passes anterior to the root of right hilum. Direct visualisation of nerve is at times difficult on CT.

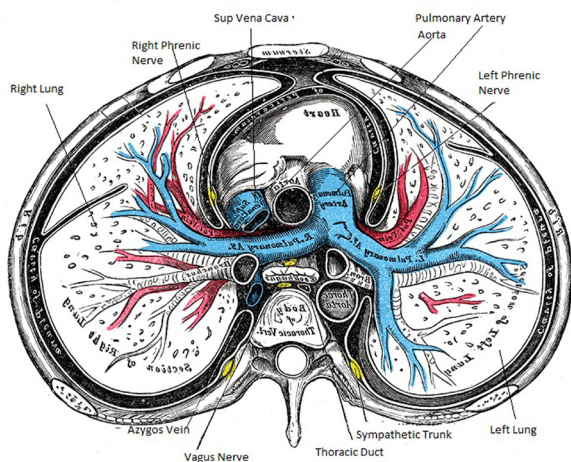


Figure 4: Cross-sectional CT diagram illustrating relationship of right phrenic nerve and ascending aorta.

is a life-threatening medical emergency. Thoracic aortic aneurysm may present atypical symptoms such as dysphagia due to compression of the esophagus; hoarseness due to vocal cord paralysis or compression of the recurrent laryngeal nerve; superior vena caval syndrome due to compression of the superior vena cava; cough, dyspnea or both due to tracheal compression; hemoptysis due to rupture of the aneurysm into a bronchus; and shock due to rupture of the aneurysm. Thoracic or lumbar vertebral body erosion may cause back pain, spinal instability, and neurologic deficits from spinal cord compression; this is particularly the case with mycotic aneurysms.

Phrenic nerve palsy is a very unusual feature and, to the best of our knowledge, there is only one reported case in literature of thoracic aortic aneurysm coupled with phrenic nerve palsy (2). In a case reported by Rabago et al., a patient with a descending thoracic aortic aneurysm presented left hemidiaphragmatic paralysis consistent with left phrenic nerve palsy (2). Common causes of phrenic nerve palsy include malignancy such as bronchogenic carcinoma, as well as mediastinal and neck tumors. Phrenic nerve palsy can also occur due to a penetrating injury or due to iatrogenic causes arising, for example, during cardiac surgery and central venous catheterisation. Many cases of phrenic nerve palsy are idiopathic.

In our case, there is ascending thoracic aortic aneurysm extending into the aortic arch, as well as right hemidiaphragm elevation suggestive of right phrenic nerve palsy, due to the pressure effect of the aneurysm on the right hilum. Dysphagia and hoarseness of voice were also present in our patient, which are uncommon symptoms of thoracic aortic aneurysm. Dysphagia due to aortic aneurysm is termed as dysphagia aortica, while hoarseness due to the left recurrent laryngeal nerve palsy, caused by a specific cardiovascular pathology such as an aortic aneurysm, is called cardio-vocal syndrome or Ortner's Syndrome.

CT is the modality of choice for identifying rupture and impending rupture of thoracic aortic aneurysms. Prompt diagnosis is imperative, as the patient may exsanguinate to death in a very short span of time. Most ruptures are manifested as a high-attenuation hematoma on non-contrast scans extending into the mediastinum, pleural cavity, pericardium, or adjacent luminal structures such as the airway or esophagus. Discontinuity of the aortic wall or a focal gap in otherwise continuous circumferential wall calcifications, along with active extravasation of contrast material on contrast-enhanced scans, may point to the location of a rupture. Contained

or impending ruptures are more difficult to identify. A peripheral crescent-shaped area of hyperattenuation in the mural thrombus of an aneurysm may represent an impending rupture. There is usually a delay of several hours between the initial intramural hemorrhage and frank extravasation. Other features suggestive of instability, acute contained or impending rupture include large aneurysm size, a low thrombus-to-lumen ratio, and draping of the posterior aspect of an aneurysm over the vertebrae, also referred to as “the draped aorta sign”. It is believed that a thick circumferential thrombus protects against rupture. As the aneurysm size increases, there is a decrease in the thrombus-to-lumen ratio and thus, a greater predisposition to rupture.

Surgical correction of thoracic aortic aneurysms is advised on the basis of its size or growth rate and symptoms. Elective repair of ascending aneurysms should be performed if the size is greater than 5.5 cm, or greater than 6.5 cm in the case of descending aneurysms, in the absence of any familial disorders (4). Patients with aneurysms as well as a history of familial disorders such as Marfan syndrome should be surgically corrected earlier, particularly if the ascending aorta is more than 5.0 cm, or the descending aorta more than 6.0 cm. A rapid increase in size is also an indicator for surgical correction. An increase in size of 1 cm or more per year is an indication of the need for elective surgical repair (5). Symptomatic patients should undergo surgical resection regardless of size because of the risk of rupture, as in our case. Acutely symptomatic patients require emergent operation. Emergent operation is also indicated in cases of acute rupture.

Surgical correction can be done by open surgical repair in which the aneurysm is removed and replaced with a tube graft. A new modality for surgery is endovascular stent graft aneurysm repair (6).

Conclusion

This case reports a rare involvement of right phrenic nerve palsy and also highlights the awareness of some radiological characteristics of impending rupture, for which urgent measures are mandatory.

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Conflict of Interest

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Conception and design: JA

Drafting of the article and final approval of the article: JA, MAS

Critical revision of the article for the important intellectual content: NAK, MAA

Provision of study materials or patient: NAK

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