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CASE REPORT

Combined Recurrent Laryngeal and Phrenic Nerve Paralysis due to Aortic Arch Aneurysm

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ABSTRACT

Idiopathic left recurrent laryngeal nerve palsy is a relatively common entity. However, very occasionally, a sinister mediastinal cause is diagnosed on investigation. Even more rare is the situation where the same lesion also causes a phrenic nerve palsy.

We present such an interesting case where an aortic arch aneurysm, in an elderly male with renal disease, led to both recurrent laryngeal and phrenic nerve palsies. The clinical features, management options and related literature are discussed.

Keywords: Recurrent laryngeal nerve palsy, Phrenic nerve, Aortic aneurysm.

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INTRODUCTION

Unilateral vocal fold paralysis (UVFP) is a common disorder seen in clinical laryngology with an unknown incidence and prevalence in general population.[1] Vocal fold paralysis is not a disease entity in itself, but a sign of an underlying disorder of the vagus nerve or its recurrent laryngeal branch. It could be an isolated palsy or may be in combination with other symptoms depending on the cause and site of the lesion. Thoracic aortic artery aneurysm presenting as hoarseness due to vocal fold paralysis, though rare, is well described in literature. Phrenic nerve palsy is an extremely unusual feature associated with thoracic aneurysm.

We present here a rare case of combined recurrent laryngeal nerve and phrenic palsy, resulting from a thoracic aortic artery aneurysm. The clinical features and management options are discussed.

CASE REPORT

A 65 years old male patient, chronic smoker, was referred from department of Nephrology to ENT Outpatient Department with history of hoarseness of 1 month duration. He also gave history of dyspnea on exertion, orthopnea and intermittent claudicating pain over both lower limbs of 1 month. He had no dysphagia, aspiration and neck or chest discomfort. His past medical history included polycystic renal disease and renal failure on treatment at the department of nephrology at our institute. He was hypertensive since 5 years and diabetic (type 2 DM) since 10 years, and on medications for these. On examination, he had a normal decubitus. The laryngeal examination showed left vocal cord fixity in a neutral position with phonatory gap. There was no other cranial nerve involvement clinically. A clinical diagnosis of left recurrent laryngeal nerve palsy was made and was evaluated to find out the cause. An opinion from physician was sought. A chest radiograph revealed a mediastinal widening with elevated left hemi-diaphragm (Fig. 1). Sputum AFB staining and cytology for malignant cells were negative. A computerized tomogram (CT scan) of thorax was taken. This showed an 8.4 × 3.5 cm saccular aneurysm involving the arch of aorta in the aortopulmonary window region, with a 6.2 cm lumen and 3.5 cm crescentic thrombus noted in the periphery. The hemidiaphragmatic paralysis on the left side was also noted (Figs 2A and B). The patient was sent to the Cardiothoracic department for an opinion on further management. He was advised surgery, which was declined by the patient. He was subsequently lost to follow-up.

DISCUSSION

An aortic aneurysm is a permanent localized dilatation resulting in an increase in diameter of at least one and a half times the normal diameter of that portion of the aorta. Chest pain, due to compression or erosion of surrounding structures, is the most common symptom.[2] Phrenic nerve palsy is an unusual feature and, to the best of our knowledge, there are only two such reported cases in world literature. Of these, one was due to aneurysm of the ascending aorta with complaints of chest pain, dysphagia and hoarseness.[2] The other was the first ever reported case, caused by descending thoracic aorta aneurysm, presenting with chronic dysphagia and back pain.[3] Khan et al described that about 10% of aortic aneurysms were painless and could present with symptoms secondary to the complications of the dissection.[4] Hoarseness as the presenting feature of thoracic aortic aneurysm is a rare entity, occurring in approximately 5% of cases.[5] The case reported here presented with hoarseness and orthopnea and the aneurysm affected the arch of aorta.

The most common cause for aortic aneurysm is atherosclerosis. The risk factors for the development of aneurysm are hypertension, old age, smoking, chronic obstructive pulmonary disease and iatrogenic trauma. Connective tissue disorders having a high predilection for

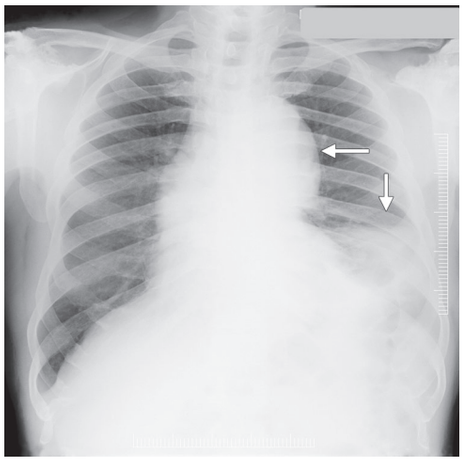
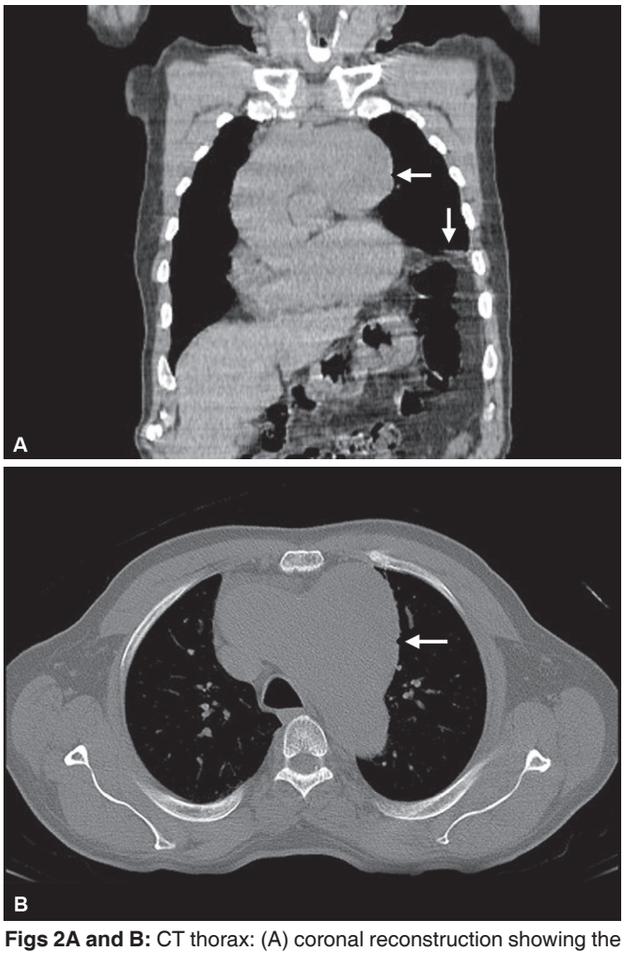


Fig. 1: Chest radiograph showing a mediastinal widening (left arrow) with elevated left hemidiaphragm (down arrow)



Figs 2A and B: CT thorax: (A) coronal reconstruction showing the saccular aneurysm of arch of aorta (left arrow) and elevated left hemidiaphragm (down arrow), (B) axial cut

thoracic aneurysm are Marfan's syndrome and Ehler Danlos syndrome. The incidence of thoracic aortic dissection has been reported to be seven times more common in patients with polycystic kidney disease than in the general population at autopsy.[6,7] But, there are only few reported cases of such an association and majority of them, involve the abdominal aorta.[8] The opinion that polycystin and extracellular matrix abnormalities may primarily predispose individuals with polycystic kidney disease to thoracic aortic dissection is supported by various studies.[7-9] In the present case report, the patient had multiple risk factors. He was an elderly hypertensive who was a smoker with polycystic kidney disease.

Asymptomatic aneurysms are detected usually on routine chest radiography. The aneurysm produces a widening of the mediastinal silhouette, enlargement of the aortic knob or displacement of the trachea from midline. However, smaller aneurysms may not be apparent on chest X-ray. A computer tomogram with intravenous contrast is an accurate diagnostic tool in the evaluation of thoracic aneurysmal disease. Helical computer tomogram of the thoracic aorta provides a minimally invasive and rapid assessment of the aortic lumen and branch vessels, the aortic wall and the surrounding tissues.[5] Our patient initially underwent a chest radiograph. Based on its findings, a compressive lesion was suspected and a high resolution computer tomogram was done. Contrast enhancement was not done as the patient had an abnormal renal function test.

Aortic arch aneurysms are a therapeutic challenge, as they are usually associated with a high surgical risk and their natural history is poorly understood. The main aim of treating aortic aneurysms is to prevent rupture and dissection, which are the most serious complications. There are currently three therapeutic options available: surgical, endovascular and medical. For the thoracic aorta, the accepted threshold for surgical intervention (in the absence of aortic regurgitation) is 55 mm, except when associated with Marfan's syndrome or bicuspid aortic valve, for which the figure is 50 mm, or less in some cases. For the descending aorta, the surgical threshold is 60 to 65 mm.[10] The patient presented here ideally required a surgical management but he declined as the surgical risks were high.

CONCLUSION

We present this case to highlight a very rare combination of three uncommon medical entities–aortic arch aneurysm causing combined phrenic and recurrent laryngeal nerve paralyses. A high index of suspicion is required for the proper diagnosis, especially for those patients at high-risk for an aneurysm. Early intervention is essential to avoid fatal complications. An extensive medline search revealed that, a case of arch of aorta aneurysm presenting with symptoms of combined recurrent laryngeal and phrenic nerve palsy in a patient with polycystic kidney disease has not been reported till date.

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