CASE REPORT

Aortic Aneurysm: A Rare Cause of Ortner's Syndrome

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Abstract

A young man presented with hoarseness of voice and was found to have left vocal cord paralysis and a large opacity on chest X-ray in the left upper zone. CT angiography showed a giant aneurysm of the aortic arch involving the left subclavian artery. Using a dual perfusion system, with the femoral bypass circuit taking care of the spinal protection and the aortic bypass circuit providing the cerebral protection, the aneurysm was excised and a 16 mm Dacron® graft was anastomosed to the aortic arch and the left subclavian artery was anastomosed to the interposition graft. He had a smooth post-operative course and his hoarseness subsided in next 6 months.

Key Words: *Hoarseness. Aortic arch aneurysm. Recurrent laryngeal nerve. Ortner's syndrome. Subclavian aneurysm. Vocal cord paralysis. Cardio-vocal syndrome.*

INTRODUCTION

In 1897, an Austrian Physician, Nobert Ortner described a rare cause of hoarseness of voice in patients with severe mitral stenosis secondary to rheumatic heart disease. This rare condition is known as Ortner's syndrome.[1] Ortner explained the hoarseness of voice due to left recurrent laryngeal nerve palsy as a result of an enlarged left atrium (a sequel of severe mitral stenosis) compressing the nerve against the aortic arch in the aorto-pulmonary window.[2]

Over the years, this rare syndrome has been associated with cardiovascular diseases that have been described as causing left recurrent laryngeal nerve (RLN) palsy in the aorto-pulmonary window thus causing Ortner's syndrome which include enlargement of left atrium due to mitral regurgitation and atrial myxomas, severe pulmonary hypertension due to congenital heart diseases, iatrogenic (due to cardiac surgery) and thoracic aortic aneurysms. Thus, this syndrome is also known as cardio-vocal syndrome.[3]

An aneurysm having a diameter more than 10 cm is defined as a giant aneurysm. We present a rare case of cardio-vocal hoarseness or Ortner's syndrome secondary to a giant aortic arch aneurysm.

CASE REPORT

A 28 years old soldier presented to the otolaryngology outpatient with hoarseness of voice, sore throat and nasal blockade. He was treated with antibiotics and anti-inflammatory drugs and in about 10 days his symptoms subsided except for the hoarseness. On further evaluation, left vocal cord paralysis was seen on indirect laryngoscopy. Chest X-ray showed a large opacity in the left upper zone. A contrast enhanced CT scan of the chest revealed a large saccular aneurysm of the arch of aorta, involving the origin of left subclavian artery and proximal descending thoracic aorta and causing passive collapse of the superior lobe of the left lung. The patient was referred to the department for further investigation and definitive management.

His general physical examination was unremarkable not having any stigmata of Marfan's or Ehler Danlos syndrome. On chest auscultation, a systolic flow murmur was audible and there were decreased breath sounds in the left upper zone. Multi-slice CT showed a giant aneurysm of the aortic arch starting proximal to and involving the origin of the left subclavian artery and the proximal part of the descending thoracic aorta. The aneurysm was 13.5 cm in diameter, and 21 cm in length. A three dimensional reconstruction was also done to properly elucidate its relations to the surrounding structures (Figure 1). His trans-thoracic and trans-oesophageal echocardiography revealed normal cardiac dimensions and an ejection fraction of 50%. Other synchronous aneurysmal lesions of the rest of aorta were ruled out via ultrasonography and echocardiography. Pre-operative laboratory investigations were normal and after pre-anaesthetic evaluation, repair of the aneurysm was planned.

Expertise and equipment for establishing left heart bypass were not available, therefore, the cardio-pulmonary bypass was established by cannulating the left femoral artery and the femoral vein i.e. the femoro-femoral bypass before thoracotomy, to decompress the aneurysm. The aneurysm was approached via left lateral thoracotomy and after separating it from the lung proximal and distal control of the aneurysm was taken

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Received: December 07, 2011; Accepted: March 14, 2013.

(Figure 2). This was followed by aortic cannulation and the venous cannulation was done in the main pulmonary artery, thus, establishing a dual perfusion system as shown in the flow chart, with the femoral bypass circuit taking care of the spinal protection and the aortic bypass

Figure 1: Three dimensional reconstruction of the pre-operative multi-slice CT scan showing the aneurysm.

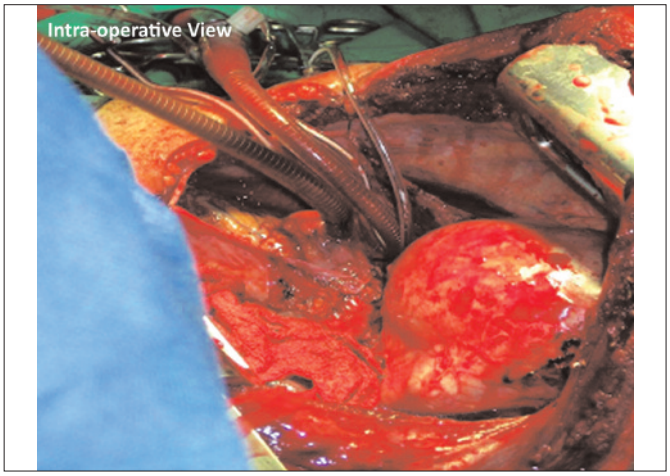

Figure 2: The giant aortic arch aneurysm is visible after left lateral thoracotomy and establishing cardiopulmonary bypass.

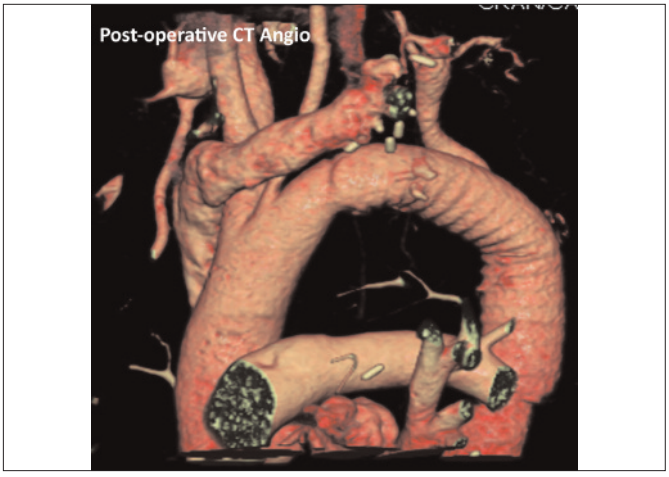

Figure 3: Three dimensional reconstruction of the postoperative multi-slice CT scan showing the Dacron® graft.

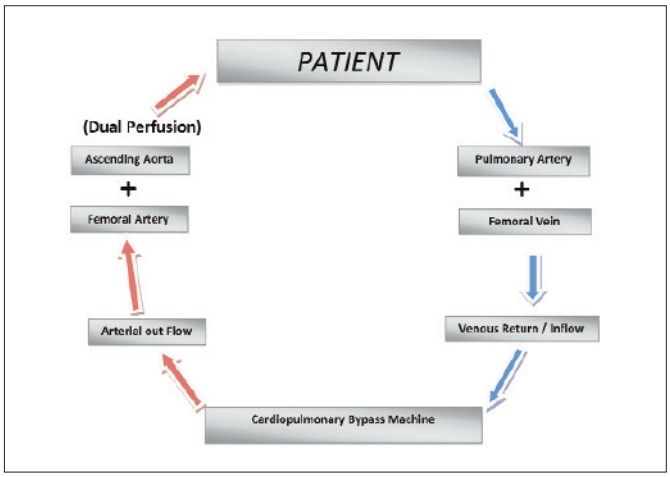

Figure 4: Flow chart of the cardiopulmonary bypass circuit (flow chart explaining the cardiopulmonary bypass circuit used in the surgery of our patient).

circuit providing the cerebral protection. Once the cardiopulmonary bypass (CPB) was fully initiated, the aneurysm was excised and a 16 mm Dacron® graft was anastomosed to the aortic arch just after the origin of left common carotid artery and distally to the descending aorta. Left subclavian artery was anastomosed to the interposition graft (Figure 3).

Weaning from CPB was smooth and the postoperative ICU stay was uneventful. Ten days in the postoperative period, his hoarseness started to subside with marked improvement in his phonation and he was discharged uneventfully on the 10th postoperative day. Six months postoperatively the hoarseness was virtually non-existent.

DISCUSSION

Understanding the pathophysiology of cardio-vocal syndrome mandates a review of the anatomy of RLN. In the superior mediastinum, RLN arises from the vagus nerve, the right RLN after crossing the first part of the right subclavian artery curls back to follow a course between the trachea and esophagus. However, on the left side, the RLN arises on the antero-lateral aspect of the aortic arch and hooks back in a space between the aortic arch and pulmonary artery known as the aorto-pulmonary window crossing the ligamentum arteriosum and posteriorly ascending in the tracheo-esophageal groove.[2,4]

Cadaveric studies of the anatomic course and relations of the left RLN have concluded that in Ortner's syndrome the nerve is compressed between the aorta, ligamentum arteriosum and the left pulmonary artery, as the distance between the aorta and the pulmonary artery in the aorto-pulmonary window is only 4 mm.[3,4] Although this syndrome initially was described for cardiovocal hoarseness due to an enlarged left atrium by Ortner but over the years it has been used by many authors to describe cardio-vocal hoarseness due to many causes

like enlargement of left atrium due to mitral regurgitation and atrial myxomas, severe pulmonary hypertension due to congenital heart diseases, iatrogenic (due to cardiac surgery) and thoracic aortic aneurysms.[3]

The incidence of thoracic aortic aneurysms (TAA) in males has been reported to be 16.3 per 100,000 person/year and in females it is upto 9.1 per 100,000 person/ year.[5] In patients with hoarseness of voice 11% of the cases have idiopathic or rare causes of hoarseness of voice which include cardio-vocal hoarseness.[4] From an Otolaryngologist's point of view cardio-vocal hoarseness is an uncommon diagnosis in patients attending their outpatient department.[1] Cardiovascular causes are considered only when the more common causes are excluded.[4]

In TAAs, most commonly involved site of the thoracic aorta is the ascending thoracic aorta with or without aortic root involvement followed by the aneurysms of the descending thoracic aorta. In only 10% of the cases of TAAs the aortic arch is involved.[6]

Patients having thoracic aortic aneurysms are usually asymptomatic.[6,7] Upto 5% of patients with TAAs develop cardio-vocal hoarseness due to the left recurrent laryngeal nerve palsy.[7] Hoarseness is subtle and initially may occur intermittently but progressively worsens and the patient may develop complete aphonia.[2]

Computed tomography (CT) angiography is an ideal test for detecting TAA, for delineating its anatomy, determining its size, defining the aortic branches, the extent involvement and the affect on adjacent structures. Nowadays, 3-dimensional reconstruction of the TAA plays an integral role in deciding and planning the appropriate surgical procedure.[6]

The AHA guidelines for TAAs dictate that an aneurysm more than or equal to 5.5 cm at the time of diagnosis, an aneurysm with an expansion rate of more than 0.5 cm per year and onset of symptoms (which includes hoarseness) are the indications for surgical intervention.[6] The size of the thoracic aortic aneurysm is a key parameter for predicting the risks of complications like dissection, rupture and death. When the critical size of 6 cm is reached 31% patients suffer a dissection or rupture. The annual risk of these grave complications is 14.1% in patients with a TAA of 6 cm or more compared to 5.3% in patients with TAA in the range of 4 - 4.9 cm.[8]

According to the Laplace's law, tension is equal to the product of pressure and radius, therefore, with an increasing radius, the wall tension also increases. Thus, the larger the aneurysm, the greater the risk of rupture.[9]

A symptomatic patient with a TAA should be operated regardless of the size.[6,8] The onset of, hoarseness represents an imminent catastrophe in the form of aneurysm rupture. Although rare, cardio-vocal syndrome secondary to a TAA may be the sole clinical sign preceding its rupture.[10]

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